

# Tumors of the Sacrum

Diagnosis and Treatment of  
Benign and Malignant Tumors

Pietro Ruggieri · Andrea Angelini  
Daniel Vanel · Piero Picci  
*Editors*

---

## Tumors of the Sacrum

---

Pietro Ruggieri • Andrea Angelini  
Daniel Vanel • Piero Picci  
Editors

# Tumors of the Sacrum

Diagnosis and Treatment of Benign and  
Malignant Tumors

 Springer

*Editors*

Pietro Ruggieri  
Department of Orthopedics and  
Orthopedic Oncology  
University of Padova  
PD, Italy

Andrea Angelini  
Department of Orthopedics and  
Orthopedic Oncology  
University of Padova  
PD, Italy

Daniel Vanel  
Department of Pathology  
Istituto Ortopedico Rizzoli  
Bologna, Italy

Piero Picci  
Laboratory of Experimental Oncology,  
Musculoskeletal Oncology,  
Istituto Ortopedico Rizzoli  
Bologna, Italy

ISBN 978-3-319-51200-6      ISBN 978-3-319-51202-0 (eBook)  
DOI 10.1007/978-3-319-51202-0

Library of Congress Control Number: 2017940037

© Springer International Publishing AG 2017

This work is subject to copyright. All rights are reserved by the Publisher, whether the whole or part of the material is concerned, specifically the rights of translation, reprinting, reuse of illustrations, recitation, broadcasting, reproduction on microfilms or in any other physical way, and transmission or information storage and retrieval, electronic adaptation, computer software, or by similar or dissimilar methodology now known or hereafter developed.

The use of general descriptive names, registered names, trademarks, service marks, etc. in this publication does not imply, even in the absence of a specific statement, that such names are exempt from the relevant protective laws and regulations and therefore free for general use.

The publisher, the authors and the editors are safe to assume that the advice and information in this book are believed to be true and accurate at the date of publication. Neither the publisher nor the authors or the editors give a warranty, express or implied, with respect to the material contained herein or for any errors or omissions that may have been made. The publisher remains neutral with regard to jurisdictional claims in published maps and institutional affiliations.

Printed on acid-free paper

This Springer imprint is published by Springer Nature  
The registered company is Springer International Publishing AG  
The registered company address is: Gewerbestrasse 11, 6330 Cham, Switzerland

---

# Contents

## Part I General Aspects

- 1 Epidemiology of Bone Lesions of the Sacrum** . . . . . 3  
Piero Picci
- 2 Overview on Bone Sacral Tumors** . . . . . 9  
Alexandra Gangi and Ricardo Gonzalez
- 3 Clinical and Neurological Manifestations of Sacral Tumors** . . . . . 21  
Alexandra Gangi and Ricardo Gonzalez
- 4 Imaging of Sacral Tumors and Tumor Simulators:  
Experience of the Mayo Clinic** . . . . . 25  
Laurel A. Littrell and Doris E. Wenger
- 5 Imaging of Sacral Tumors: Experience of the Rizzoli Institute** . . . . . 65  
Alessandra Bartoloni, Alberto Bazzocchi, and Daniel Vanel
- 6 Biopsy and Staging of Sacral Tumors** . . . . . 83  
John E. Mullinax and Ricardo J. Gonzalez
- 7 Histopathology of Sacral Tumors and Pseudotumors** . . . . . 93  
Marilyn M. Bui, Yi Ding, Evita Henderson Jackson,  
and Angelo Paolo Dei Tos

## Part II Benign Lesions

- 8 Giant Cell Tumor of the Sacrum** . . . . . 123  
Andreas F. Mavrogenis, Georgios N. Panagopoulos,  
Andrea Angelini, and Pietro Ruggieri
- 9 Osteblastoma of the Sacrum** . . . . . 137  
Andrea Angelini and Pietro Ruggieri
- 10 Osteoid Osteoma of the Sacrum** . . . . . 147  
Andrea Angelini and Pietro Ruggieri

<b>11</b>	<b>Aneurysmal Bone Cyst of the Sacrum</b> . . . . .	153
	Andrea Angelini, Giuseppe Rossi, Andreas F. Mavrogenis, and Pietro Ruggieri	
<b>12</b>	<b>Schwannoma of the Sacrum.</b> . . . .	163
	Andreas F. Mavrogenis, Georgios N. Panagopoulos, Andrea Angelini, and Pietro Ruggieri	
<b>13</b>	<b>Benign Cartilaginous Tumors of the Sacrum.</b> . . . .	171
	Andrea Angelini and Pietro Ruggieri	
<b>Part III Malignant Lesions</b>		
<b>14</b>	<b>Metastases of the Sacrum.</b> . . . .	181
	Andreas F. Mavrogenis, Georgios N. Panagopoulos, Andrea Angelini, and Pietro Ruggieri	
<b>15</b>	<b>Chordoma of the Sacrum.</b> . . . .	195
	Andrea Angelini and Pietro Ruggieri	
<b>16</b>	<b>Osteosarcoma of the Sacrum</b> . . . . .	213
	Andrea Angelini, Douglas G. Letson, and Pietro Ruggieri	
<b>17</b>	<b>Ewing's Sarcoma of the Sacrum</b> . . . . .	221
	Andrea Angelini, Douglas G. Letson, and Pietro Ruggieri	
<b>18</b>	<b>Lymphoma and Myeloma of the Sacrum</b> . . . . .	227
	Andreas F. Mavrogenis, Georgios N. Panagopoulos, Andrea Angelini, Pier Luigi Zinzani, and Pietro Ruggieri	
<b>19</b>	<b>Chondrosarcoma of the Sacrum</b> . . . . .	237
	Andrea Angelini, Andreas F. Mavrogenis, and Pietro Ruggieri	
<b>Part IV Treatments</b>		
<b>20</b>	<b>Anatomy and Surgical Approaches to the Sacrum</b> . . . . .	247
	Sean Accardo and Ricardo Gonzalez	
<b>21</b>	<b>Tumors of the Sacrum: Diagnosis, Management, and Surgical Techniques.</b> . . . .	255
	Eric T. Newman, Francis J. Hornicek, and Joseph H. Schwab	
<b>22</b>	<b>Computer Navigation in the Sacrum</b> . . . . .	275
	David M. Joyce	
<b>23</b>	<b>Sacral Biomechanics and Reconstruction.</b> . . . .	321
	Matthew T. Houdek, Peter S. Rose, Steven L. Moran, Michael J. Yaszemski, and Franklin H. Sim	

---

<b>24</b>	<b>Soft Tissue Reconstruction Following Sacrectomy</b> . . . . .	333
	Matthew T. Houdek and Steven L. Moran	
<b>25</b>	<b>Embolization for Sacral Tumors</b> . . . . .	341
	Andreas F. Mavrogenis, Vasilios Igoumenou, Andrea Angelini, Giuseppe Rossi, and Pietro Ruggieri	
<b>26</b>	<b>Palliative Treatments for the Sacrum</b> . . . . .	353
	Andreas F. Mavrogenis, Georgios N. Panagopoulos, Andrea Angelini, Giuseppe Rossi, Alberto Bazzocchi, and Pietro Ruggieri	
<b>27</b>	<b>Radiation Therapy for Primary Malignant Sacral Tumors</b> . . . . .	365
	Joseph H. Schwab and Francis J. Hornicek	
<b>28</b>	<b>Tumors of the Sacrum: The Role of Chemotherapy</b> . . . . .	373
	Stefano Ferrari	

---

## List of Contributors and Author Bios

---

### List of Contributors

**Sean Accardo, M.D.** Department of Sarcoma, H. Lee Moffitt Cancer Center, Tampa, FL, USA

**Andrea Angelini, M.D., Ph.D.** Department of Orthopedics and Orthopedic Oncology, University of Padova, Padova, Italy

**Alessandra Bartoloni, M.D.** Diagnostic and Interventional Radiology, Istituto Ortopedico Rizzoli, Bologna, Italy

**Alberto Bazzocchi, M.D.** Diagnostic and Interventional Radiology, Istituto Ortopedico Rizzoli, Bologna, Italy

**Marilyn M. Bui, M.D., Ph.D.** Department of Anatomic Pathology, H. Lee Moffitt Cancer Center, Tampa, FL, USA

Department of Sarcoma, H. Lee Moffitt Cancer Center, Tampa, FL, USA

**Angelo Paolo Dei Tos, M.D.** Department of Anatomic Pathology, General Hospital of Treviso, Treviso, Italy

**Yi Ding, M.D.** Department of Pathology, Beijing Jishuitan Hospital, Beijing, China

**Stefano Ferrari, M.D.** Department of Chemotherapy, Istituto Ortopedico Rizzoli, Bologna, Italy

**Alexandra Gangi, M.D.** Department of Surgical Oncology, H. Lee Moffitt Cancer Center, Tampa, FL, USA

**Ricardo Gonzalez, M.D.** Department of Surgical Oncology, H. Lee Moffitt Cancer Center, Tampa, FL, USA

**Evita Henderson-Jackson, M.D.** Department of Anatomic Pathology, H. Lee Moffitt Cancer Center, Tampa, FL, USA

Department of Sarcoma, H. Lee Moffitt Cancer Center, Tampa, FL, USA



**Francis J. Hornicek, M.D., Ph.D.** Section of Orthopaedic Oncology, Department of Orthopaedic Surgery, Massachusetts General Hospital, Boston, MA, USA

**Matthew T. Houdek, M.D.** Department of Orthopedic Surgery, Mayo Clinic, Rochester, MN, USA

**Vasilios Igoumenou, M.D.** First Department of Orthopaedics, National and Kapodistrian University of Athens, Athens, Greece

**David M. Joyce, M.D.** Department of Sarcoma, H. Lee Moffitt Cancer Center, Tampa, FL, USA

**Douglas G. Letson, M.D.** Department of Surgery, University of South Florida, H. Lee Moffitt Cancer Center, Tampa, FL, USA

**Laurel A. Littrell, M.D.** Department of Radiology, Mayo Clinic, Rochester, MN, USA

**Andreas F. Mavrogenis, M.D., Ph.D.** First Department of Orthopaedics, National and Kapodistrian University of Athens, Athens, Greece

**Steven L. Moran, M.D.** Division of Plastic and Reconstructive Surgery, Mayo Clinic, Rochester, MN, USA

**John E. Mullinax, M.D.** Department of Surgical Oncology, H. Lee Moffitt Cancer Center, Tampa, FL, USA

**Erik T. Newman, M.D.** Section of Orthopaedic Oncology, Department of Orthopaedic Surgery, Massachusetts General Hospital, Boston, MA, USA

**Georgios N. Panagopoulos, M.D.** First Department of Orthopaedics, National and Kapodistrian University of Athens, Athens, Greece

**Piero Picci, M.D.** Laboratory of Experimental Oncology, Musculoskeletal Oncology, Istituto Ortopedico Rizzoli, Bologna, Italy

**Peter S. Rose, M.D.** Department of Orthopedic Surgery, Mayo Clinic, Rochester, MN, USA

**Pietro Ruggieri, M.D., Ph.D.** Department of Orthopedics and Orthopedic Oncology, University of Padova, Padova, Italy

**Joseph H. Schwab, M.D., M.S.** Section of Orthopaedic Oncology, Department of Orthopaedic Surgery, Massachusetts General Hospital, Boston, MA, USA

**Franklin H. Sim, M.D.** Department of Orthopedic Surgery, Mayo Clinic, Rochester, MN, USA

**Daniel Vanel, M.D.** Department of Pathology, Istituto Ortopedico Rizzoli, Bologna, Italy

**Doris E. Wenger, M.D.** Department of Radiology, Mayo Clinic, Rochester, MN, USA

**Michael J. Yaszemski, M.D., Ph.D.** Department of Orthopedic Surgery and Biomedical Engineering, Mayo Clinic, Rochester, MN, USA

**Pier Luigi Zinzani, M.D., Ph.D.** Institute of Hematology “L. e A. Seràgnoli,” University of Bologna, Bologna, Italy

---

## Author Bios

**Andrea Angelini** is an orthopedic surgeon at the Department of Orthopedics and Orthopedic Oncology, University of Padova.

Born in Savignano sul Rubicone, Italy, in 1983, he obtained his medical degree (cum laude) from the University of Bologna in 2008. He obtained board certification in orthopedic surgery at the University of Bologna and Istituto Ortopedico Rizzoli in 2014, followed by a Ph.D. in oncology and experimental pathology from the University of Bologna in 2016. His main field of interest is surgery for musculoskeletal tumors: he collaborated for a book (editor of one book), chapters (contributor of four published book chapters), and numerous scientific papers (62 published papers available on PubMed, about 600 citations, H-index 13, IF 85). He has held courses and talks at international meetings (n. 60 presentations), and he is coauthor of more than 200 meeting abstracts. He was awarded for his scientific activity by EFORT, SIOT, EMSOS, and ISOLS. He lives in Bologna and is married with one child.

**Piero Picci** is director of the Laboratory of Experimental Oncology of Istituto Ortopedico Rizzoli in Bologna, Italy. He obtained his degree in medicine and surgery from the University of Bologna (1979) and completed his board certification in oncology at the same university (1983).

He is coordinator of the EU project PROTHETS (Prognostic and Therapeutic Targets in Ewing Family of Tumors) and research line leader (Ewing sarcoma) in the EU project EUROBONET. He is a participant in four other EU projects.

He is author or coauthor of 446 papers on international journals (433 on Medline, H-index 70), 56 papers on national journals, 71 book chapters, 518 abstracts from international congresses, and 99 abstracts from national congresses.

His memberships include being founder and chairman of the Italian Sarcoma Group (1997); founder, board member, treasurer, vice-president, and president of EMSOS; board member of CTOS; and member of ISS, ASCO, and AIOM.

**Pietro Ruggieri** is chairman of the Department of Orthopedics and Orthopedic Oncology at the University of Padova, and Director of the Scientific Center for Research of Musculoskeletal Tumors “Mario Mercuri”.

Born in Taranto (Italy) in 1958, he graduated in medicine at the University of Bologna in 1982, completed board of orthopedics in 1987 and obtained his Ph.D. in oncology from the University of Bologna in 1989. He was a fellow at the University of Florida, Gainesville, under the direction of Dr. Enneking and Dr. Springfield in

1987 and was a fellow of Dr. Frank Sim at Mayo Clinic in 1991. He is author of more than 800 scientific papers on national/international journals (over 290 of these on PubMed), with an impact factor of over 533 and a citation H-index of 41. He has been a speaker or lecturer in more than 500 international congresses. He has been president of ISOLS. He is board member of EMSOS, co-director of the subspecialty of tumors for SICOT, and scientific coordinator for EFORT. Since October 2016, he is a member of the MSTS Membership Committee. His main fields of research are musculoskeletal oncology, reconstructive surgery, and prostheses in musculoskeletal oncology.

**Daniel Vanel** is in charge of research and teaching in musculoskeletal tumors at Istituto Ortopedico Rizzoli, Bologna, Italy.

He is former chairman of radiology of Institut Gustave Roussy, France.

He is former president of the European Society of Musculoskeletal Radiology and International Cancer Imaging Society.

He was awarded the Gold Medal of the International Skeletal Society.

He authored 276 articles in Medline.

---

## Part I

# General Aspects

Piero Picci

These data come from the Archives of musculoskeletal tumor and pseudotumoral lesions of the Istituto Ortopedico Rizzoli in Bologna. We do not report incidence data, but frequency data registered at a referral center for musculoskeletal lesions. From September 1900 to December 2014, the archive comprises 28,477 cases, of which 790 (2.77%) were lesion of the sacrum. To better understand the specificity of sacrum lesions, data will be compared to the figures of bone lesions affecting the whole skeleton.

---

## 1.1 Diagnosis

Conventional bone lesion classification usually subdivides these in “pseudotumoral,” “benign,” and “malignant,” considering the last separately between primary and part of a systemic disease (i.e., carcinoma metastasis, lymphoma, myeloma).

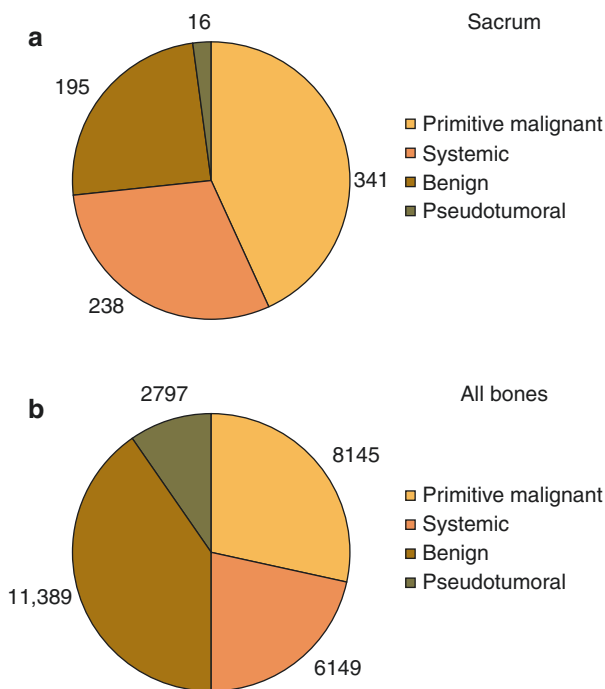
Distribution between these macro-entities differs between the sacrum and all other bone sites. In the sacrum, about three fourths of the cases are malignant, while in the other sites, only one half are malignant. Primary malignant tumors are more frequent in the sacrum (341 cases, 43.2%) followed by systemic lesions (238 cases, 30.1%), benign lesions (195 cases, 24.7%), and pseudotumoral lesions (16 cases, 2.0%). In the whole skeleton, benign lesions are more frequent (11,386 cases, 40.0%), followed by primary tumors (8145 cases, 28.6%), systemic lesions (6149 cases, 21.6%), and pseudotumoral lesions (2797 cases, 9.8%) (Fig. 1.1). Table 1.1 reports all sacrum lesions.

---

P. Picci, M.D.

Laboratory of Experimental Oncology, Musculoskeletal Oncology, Istituto Ortopedico Rizzoli, Bologna, Italy

e-mail: [piero.picci@ior.it](mailto:piero.picci@ior.it)



**Fig. 1.1** Distribution of bone lesions affecting (a) the sacrum and (b) the entire skeleton, in the Rizzoli experience. Lesions have been classified as pseudotumoral, benign, primitive malignant and malignant as part of a systemic disease

Frequency of the different entities is totally different from the general distribution in the whole skeleton. From this comparison many important differences are evident, apart from the obvious high frequency of chordomas and intraosseous schwannoma, the latter originating from the sacral roots. There is an important increase in the percentage of systemic lesions as bone metastasis from carcinoma (+25%), lymphoma (+64%), and myeloma (+130%). Among primary malignant tumors, there is an increase of Ewing sarcoma (+54%) and angiosarcoma (+50%), but there is an important decrease in the frequency of osteosarcomas (−54%) and chondrosarcomas (−43%). Within the benign tumors, there is an increase in frequency of giant cell tumor (+40%) and osteoblastoma (+145%), the latter compensated by a decrease in the frequency of osteoid osteoma (−56%).

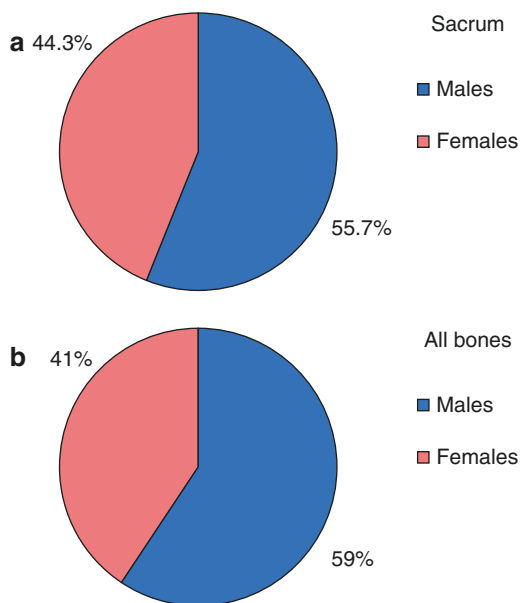
The frequency of aneurysmal bone cyst and angioma of the bone is not dissimilar from other sites. To be noted is the higher frequency (+120%) of Paget disease in the sacrum. It is important to report that three of the eight secondary osteosarcomas in the sacrum developed on Paget disease. Table 1.2 reports the incidence of the 15 most frequent entities in the sacrum compared to the frequency in the whole skeleton.

**Table 1.1** Lesions of the sacrum

<i>Pseudotumoral lesions</i>	16
Paget disease	9
Histiocytosis X	6
Solitary bone cyst	1
<i>Benign lesions</i>	195
Giant cell tumor	58
Aneurysmal bone cyst	27
Osteoid osteoma	25
Intraosseous schwannoma	25
Osteblastoma	21
Angioma of bone	9
Fibrous dysplasia	5
Notochordal benign tumor	5
Benign not otherwise specified	5
Solitary osteochondroma	4
Ependymoma	4
Teratoma	4
Chondroblastoma	1
Chondromyxoid fibroma	1
Intraosseous lipoma	1
<i>Primary malignant tumors</i>	341
Chordoma	167
Ewing sarcoma	63
Osteosarcomas	46
Classic	35
Secondary	8
Low-grade central	2
Telangiectatic	1
Chondrosarcomas	29
Central	14
Peripheral	6
Clear cell	3
Mesenchymal	3
Dedifferentiated	3
Sarcoma not otherwise specified	10
Angiosarcoma	7
Intraosseous solitary fibrous tumor	6
Undifferentiated pleomorphic sarcoma (UPS)	4
Intraosseous malignant schwannoma	3
Intraosseous leiomyosarcoma	3
Intraosseous synovial sarcoma	2
Intraosseous myoepithelioma	1
<i>Systemic tumors</i>	238
Carcinoma metastasis	153
Myeloma	49
Lymphoma	36

**Table 1.2** Comparison of frequency in the sacrum and in the whole skeleton for the 15 most frequent entities

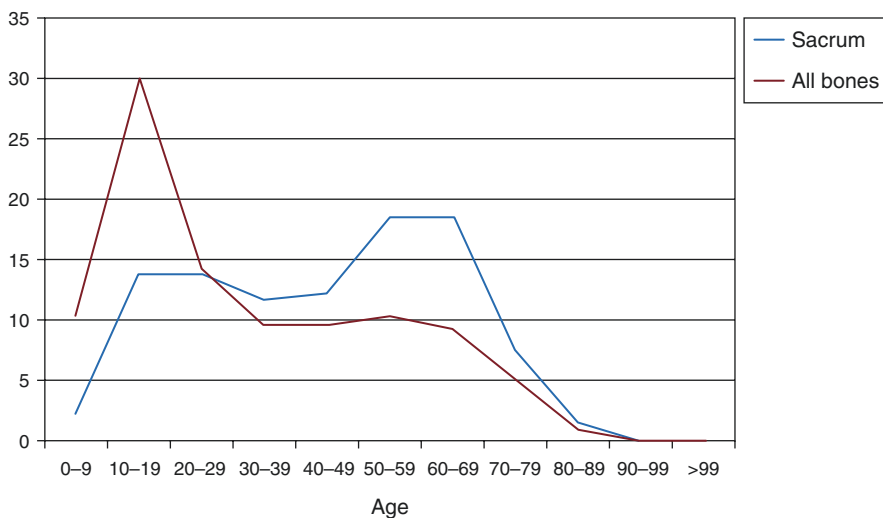
	Sacrum		Whole skeleton		Delta
	N°	%	N°	%	%
Chordoma	167	21.1	123	0.4	5175
Metastasis from carcinoma	153	19.4	4305	15.5	25
Ewing sarcoma	63	8.0	1437	5.2	54
Giant cell tumor	58	7.3	1451	5.2	40
Myeloma	49	6.2	745	2.7	130
Osteosarcomas	46	5.8	3507	12.7	-54
Lymphomas	36	4.6	767	2.8	64
Chondrosarcomas	29	3.7	1813	6.5	-43
Aneurysmal bone cyst	27	3.4	1093	3.9	-13
Osteoid osteoma	25	3.2	1992	7.2	-56
Intraosseous schwannoma	25	3.2	17	0.1	3100
Osteblastoma	21	2.7	311	1.1	145
Paget disease	9	1.1	140	0.5	120
Angioma of bone	9	1.1	279	1.0	10
Angiosarcoma	7	0.9	180	0.6	50

**Fig. 1.2** Gender distribution of patients with lesions affecting (a) the sacrum and (b) the entire skeleton, in the Rizzoli experience

## 1.2 Gender

A slight prevalence is evident in females with sacral lesions (44.3%) in comparison to other sites (41.0%) (Fig. 1.2).





**Fig. 1.3** Incidence by age

### 1.3 Age

The analysis of age shows major differences in the sacrum, compared to other bone sites.

In the sacrum, with a range of 0–89 years, the mean is 44 and the median 47, while in all other sites with a similar age range (from 0 to 103), the mean is 32 and the median is 25.

It is evident that sacral lesions develop in much older patients compared to the other bone sites. Figure 1.3 reports the incidence by decades of the two groups.

**Conflict-of-Interest Statement** No benefits have been or will be received from a commercial party related directly or indirectly to the subject matter of this article.

Alexandra Gangi and Ricardo Gonzalez

---

## 2.1 Introduction

Primary sacral tumors are rare, accounting for approximately 5–7% of all spinal tumors [1]. Metastases are the most common malignant tumors of the sacrum and can be derived from lung, breast, kidney, prostate, head and neck, gastrointestinal, or skin (melanoma) cancers [2, 3, 62]. Primary benign and malignant tumors of the sacrum may arise from bone or neural elements or the bone marrow in cases of hematological malignancies. Approximately 10% of all benign tumors or pseudotumors have been known to involve the sacrum. These can include giant cell tumors (60% of cases), aneurysmal bone cysts (4%), and osteoblastomas. Of malignant bone tumors, 6–8% involve the sacrum and include chordoma (50%), lymphoma (9%) and multiple myeloma (9%), Ewing’s sarcoma in children (8%), chondrosarcoma in adults, and osteosarcoma [4].

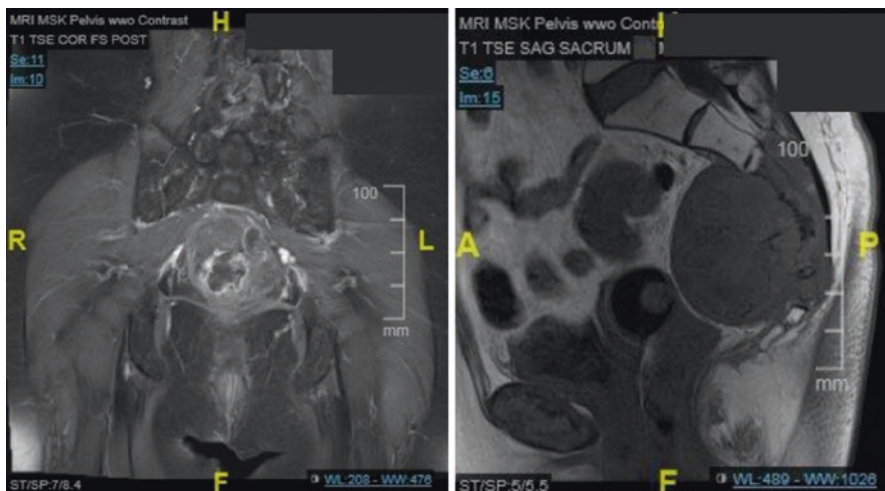
---

## 2.2 Clinical Presentation

Sacral tumors are generally diagnosed late, and the clinical pattern depends on the anatomic location of the lesion within the sacrum and involvement of specific anatomic structures [5–13]. The topic will be discussed in greater detail in the specific chapter.

---

A. Gangi, M.D. • R. Gonzalez, M.D. (✉)  
Sarcoma Department, H. Lee Moffitt Cancer Center,  
12902 Magnolia Drive, Tampa, FL 33602, USA  
e-mail: [Alexandra.Gangi@moffitt.org](mailto:Alexandra.Gangi@moffitt.org); [Ricardo.Gonzalez@moffitt.org](mailto:Ricardo.Gonzalez@moffitt.org)



**Fig. 2.1** Coronal and sagittal views of 8.5 × 6.5 cm sacral chordoma involving S3-C1

### 2.3 Imaging

Imaging is a useful adjunct in the diagnosis of sacral tumors. Although the sacrum can be frequently obscured by overlying stool or bowel gas, plain radiographs can be helpful with initial diagnosis. Nonetheless, for more thorough evaluation and better defined spatial understanding, additional imaging such as computed tomography (CT) scan or magnetic resonance imaging (MRI) are required [14, 15, 59].

In general, computed tomography is superior in showing bony details and calcifications and allows for better visualization of adjacent viscera. Lumbar CT scans usually ordered for sciatica or cruralgia must include S1 and S2 in the examination so that sacral lesions are not missed. CT-guided biopsy is particularly useful in the sacrum. If CT is substituted for MRI and there is a presacral soft tissue mass, administration of both rectal and intravenous contrast should be considered to better evaluate involvement of the pelvic structures (Fig. 2.1). When possible, however, MRI is the imaging modality of choice to specify the diagnosis, tumor extent into the sacral canal, neurovascular involvement, and aid in preoperative planning [14, 15]. In some lesions that are hypervascular, such as renal cell carcinoma, leiomyosarcoma, giant cell tumors, and hemangiopericytomas, preoperative angiography and embolization should be considered [15, 16]. This allows for reduced tumor vascularity and safer resection in select patients [15–17].

### 2.4 Biopsy

Given that the differential diagnosis of sacral tumors is extensive, a biopsy should be performed in almost all cases. A transrectal or transvaginal biopsy should generally not be performed because it violates the containing membranes of presacral

fascia and periosteum and could lead to seeding of the rectum or vagina with tumor cells. The preferred biopsy method is image-guided core biopsy, if it can be performed safely.

---

## 2.5 Benign Sacral Tumors

Most lesions of the sacrum are benign. Common benign sacral tumors in children are sacrococcygeal teratomas (the most common), lipomas, dermoids, epidermoid cysts, and bone islands or enostoses [18, 19]. Congenital abnormalities such as spina bifida occulta, tethered cord, hairy nevi, dermal sinus tracts, and dimples are associated with tumors of the sacrum in children [6, 20].

Sacrococcygeal teratomas are rare congenital tumors that arise from pluripotent cells. Although approximately 70% are benign, there is a tendency toward malignant transformation [18]. Approximately 20% of sacrococcygeal teratomas are identified prenatally; 70% are identified at birth, and the remaining 10% are identified within the first year of life. In adults, sacrococcygeal teratomas are rare and more commonly benign. On radiographs, the tumors are seen as protruding soft tissue masses with amorphous, punctuate, or spiculated calcifications. CT and MRI usually show a heterogeneous mixture of solid and cystic components [23]. Most sacrococcygeal teratoma resections are performed via a posterior approach, but occasionally a combined abdominal-sacral approach is required. In some patients, extent of resection warrants a temporary and rarely permanent colostomy [24].

While, in children, most sacral tumors tend to be benign, the frequency of benign lesions in adults is significantly lower. The most common benign sacral tumors in adults are giant cell tumors (13% of all sacral tumors), aneurysmal bone cysts, osteoblastomas, schwannomas, osteoid osteomas, skeletal osteochondromas, chondromyxoid fibromas, nerve sheath, and meningeal tumors of the sacrum [24–28, 58, 63].

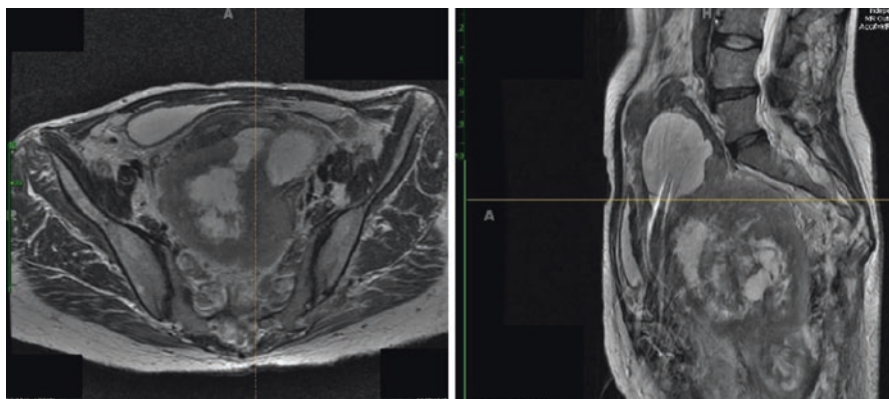
The sacrum is the third most common location for giant cell tumors which tend to affect patients in their second and fourth decades of life. Giant cell tumors also tend to be more common in females [23, 28, 29]. Sacral giant cell tumors usually develop in an eccentric position, but commonly extend to involve both sides of the midline. Additionally, they tend to have the propensity to cross the sacroiliac joints and intervertebral disks, which is unusual for many other spinal lesions and is a useful distinguishing feature of giant cell tumors [23]. Although generally classified as a benign tumor, 5–10% of giant cell tumors have been reported to be malignant. Malignancy can be characterized based on mitotic activity, 1/mm<sup>2</sup> or less is highly unlikely to be malignant, and histology and sarcomatous features within the primary specimen can indicate an increased likelihood of malignant degeneration. Additionally, patients may develop lung metastases and recurrence which demonstrate malignancy initially missed in primary tumor pathology evaluation. For these patients prognosis is poor and 5-year tumor-free survival is <50% [30]. The standard treatment for giant cell tumors is wide excision or aggressive curettage followed by adjuvant phenol, hydrogen peroxide, liquid nitrogen or argon beam therapy, embolization, and bone grafting or cementation. Cryosurgery and radiation therapy are also possible options [11, 30–34, 64]. It is important to attempt complete resection, as recurrence rates have

been noted to be as high as 50% if complete resection is not achieved [10, 21, 35]. In appropriately selected patients, sacrectomy is an optional procedure which can render the patient free of disease and improve risk of recurrence [31, 32].

The second most common benign tumor in adults is an osteblastoma. Typically, osteoblastomas affect young adults, with a male/female ratio of 2:1. Approximately 40% of these lesions occur in the spine with approximately 17% arising in the sacrum specifically [3]. Osteoblastomas should be excised. The lesions recur in 10–15% of cases, but the rate approaches 50% in the more aggressive pattern. Malignant transformation of osteoblastoma to osteosarcoma with metastases has also been reported [14].

There are additionally a handful of rarely occurring tumors of the sacrum, osteoid osteomas, cavernous hemangiomas, and chondromyxoid fibromas. Osteoid osteomas of the sacrum represent <2% of sacral tumors [4, 22, 58]. En bloc resection and radiofrequency ablation are both viable options and render low rates of recurrence [2, 33, 34]. Cavernous hemangiomas are the most common benign tumors of the spine, but only exceptionally involve the sacrum [36]. Chondromyxoid fibroma is a rare benign tumor of the sacrum [36]. Differential diagnosis should include chondrosarcoma, chordoma, and giant cell tumor. Surgical excision of the affected area or curettage and bone grafting are the treatments of choice for chondromyxoid fibroma. Radiation therapy should only be considered for the rare surgically inaccessible tumor [36]. Nonetheless, all of these lesions should be considered on the differential diagnosis when considering tumor subtypes.

Nerve sheath tumors may arise from the sacral nerve roots and include schwannomas and neurofibromas (Fig. 2.2). The most common nerve sheath benign sacral tumors are the giant sacral schwannomas; the mean diameter of these tumors is approximately 10.5 cm. Cyst formation, hemorrhage, and necrosis are relatively common in giant sacral schwannomas; unlike neurofibromas, schwannomas tend to be encapsulated. En bloc resection is the treatment of choice. Although difficult because of their size and the presence of critical sacral nerve roots, most can be resected completely, and recurrence is rare [37, 38].

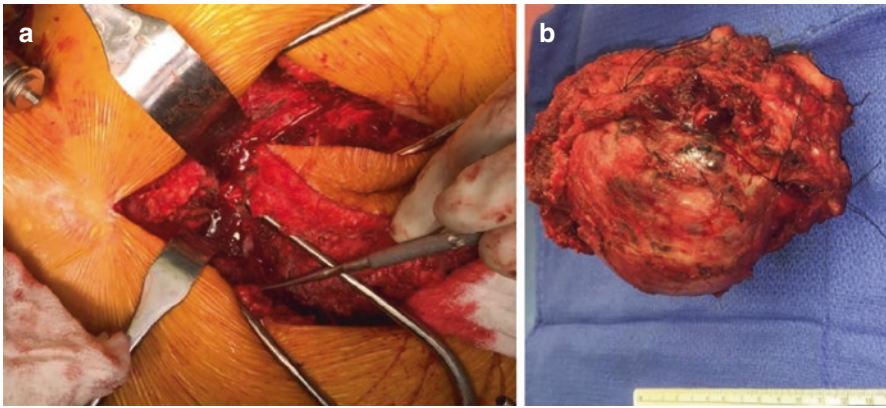


**Fig. 2.2** Plexiform neurofibroma involving the sacrum in a patient with history of neurofibromatosis type 1

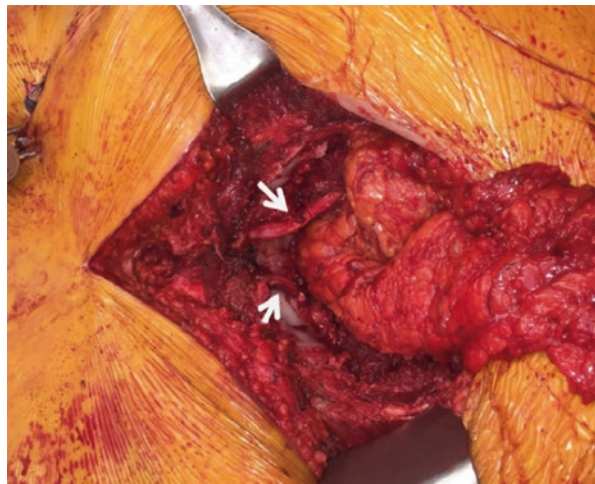
## 2.6 Malignant Sacral Tumors

The most common malignant tumors of the sacrum include chordomas, multiple myelomas, Ewing's sarcomas, and primitive neuroectodermal tumors (PNET). Primary lymphomas, osteosarcomas, chondrosarcomas, angiosarcomas, fibrosarcomas, carcinoma, and amyloid tumors of the sacrum are also malignant, but are quite rare.

Chordomas are the most common primary malignant tumor of the sacrum and the most common tumor of any type involving the sacrum [29, 38]. The majority of sacral chordomas occur in the sacrococcygeal region in patients who are 40 years of age or older and occur almost twice as frequently in men compared to women [39]. Chordomas are slow growing and often displace but generally do not invade the rectum and/or the bladder (Figs. 2.3 and 2.4). Metastases are not common, and if



**Fig. 2.3** (a) Prone approach to resection of sacral chordoma with attempted preservation of nerve roots. (b) Resected specimen



**Fig. 2.4** Post-resection of chordoma with intact nerve roots (*arrows*)

metastatic disease is encountered, it is usually a late event [39]. On imaging, there is frequently a well-circumscribed osteolysis without an osteosclerotic rim, and a solid tumor with cystic areas is seen in approximately 50% of cases [3, 14, 15, 23]. Dedifferentiated chordoma is a rare variant that is clinicopathologically analogous to dedifferentiated chondrosarcoma. The sarcomatous component of dedifferentiated chordomas generally demonstrates more aggressive biological behavior and has a higher propensity to metastasize [40]. Primary treatment for chordomas is wide resection, and patient prognosis is dependent upon the completeness of resection and the violation of the tumor margins at the initial surgery. It is imperative to obtain an R0 resection to prevent recurrence; therefore, sacrifice of sacral nerve roots at the time of initial surgery may be necessary and is not uncommon. Total sacrectomy for chordomas involving the S1 nerve root have been reported [32, 41–43]. Local recurrence is the most important predictor of mortality in patients with chordomas and is related to the extent of initial resection. Local recurrence of sacral chordomas results in high morbidity rates and is associated with an approximately 20-fold increased risk of tumor-related death [8, 9, 11, 13, 39]. If the lesion is incompletely resected, adjuvant radiation therapy is another option; however, its efficacy is debatable [7, 11, 13, 44]. Results with brachytherapy techniques for recurrent sacral chordoma have been reported in small numbers of patients with varying success rates [45]. Chemotherapy has been of little value in the management of chordomas [11, 46]. Metastases, which can be found in the liver, lung, and regional lymph nodes, eventually develop in 5–43% of patients [44, 47].

Multiple myeloma is the second most common primary malignant neoplasm of the sacrum. Its incidence peaks in the sixth and seventh decade of life and is more common in males. The earlier solitary form, plasmacytoma, affects younger patients when compared with multiple myeloma. Lesions tend to be larger than those of multiple myeloma and tend to be osteolytic and expansile. These lesions also have poorly defined margins and are frequently associated with a soft tissue mass. Plasmacytomas generally progress to multiple myeloma in 10–15 years [14, 23, 48].

Lymphomas are the third most common primary malignant tumors of the sacrum but represent less than 5% of malignant bone tumors [30]. They predominantly affect men in their fifth to sixth decades of life. Lymphomas can cause aggressive bony destruction, although they tend to extend to the soft tissue leaving the underlying bones intact [14, 23, 49]. Three imaging signs, although nonspecific, are suggestive of lymphomas. These include the intensity and extent of uptake on bone scan (reveals a hot spot), the massive bone marrow invasion on MRI (poorly defined margins with a wide zone of transition) despite normal radiographic findings, and the large soft tissue mass with no visible cortical lesion on CT [50]. This highlights the importance of pursuing investigations (particularly bone scintigraphy and MRI) in patients with persistent pain despite their having no detectable abnormality on conventional radiography [15].

Ewing's sarcoma and PNET represent the fourth most common primary malignant tumors of the spine [26, 61]. Within the spine, the sacrum is the most common site of involvement. The age range for Ewing's sarcoma is 5–30 years, with 75% occurring in the first two decades of life. The male/female ratio is 3:1. Imaging

findings tend to show paraspinous soft tissue masses and extradural space involvement [22, 51]. Some cases of sacral Ewing's sarcomas may present as a predominant soft tissue mass, extending to pelvic structures or to the spinal canal, with limited osteolysis [15]. Immunohistochemical studies are needed to distinguish Ewing's sarcoma from PNET, with the latter being characterized by neural differentiation [22, 23, 51, 60]. Primary treatment for Ewing's sarcoma and PNET is chemotherapy and radiation therapy; however, many patients require decompressive surgery and stabilization secondary to symptomatology. Unfortunately, these lesions are associated with the worst prognosis when they occur in the sacrococcygeal region, with low likelihood of local control (60%) and poor long-term survival [19].

There are a number of more rare malignant sacral tumors. Osteosarcomas account for 4% of primary malignant tumors of the sacrum. Many of the osteosarcomas of the sacrum are secondary to Paget's disease [30]. Sacral chondrosarcomas, fibrosarcomas, and angiosarcomas are unusual [52]. A 2% incidence of primary and secondary chondrosarcomas of the sacrum has been reported [4]. Most of sacral fibrosarcomas arise from a pre-existing lesion, usually previously irradiated bone, Paget's disease, or fibrous dysplasia [3].

Another rare malignant lesion is a malignant peripheral nerve sheath tumors (MPNST) (neurofibrosarcomas or malignant schwannomas). These tumors are associated with neurofibromatosis type 1 as they usually arise from pre-existing neurofibromas. Additionally, they have a tendency to recur locally and spread hematogenously, and despite aggressive surgery and adjuvant therapy, the prognosis for patients with MPNST is poor [23].

---

## 2.7 Surgical Treatment of Sacral Tumors

For a majority of the aforementioned benign and malignant tumors, complete tumor resection with negative resection is the mainstay of therapy. The surgical goals should be to remove the tumor completely with clear margins while maximizing postoperative function. For malignant lesions, a radical surgical approach such as partial or total sacrectomy, with sacrifice of sacral roots, is often warranted to achieve total resection with clear margins [8]. Various sacrectomies have been described depending on the tumor location, extent, and histology, and decision regarding partial or total sacrectomy for en bloc resection can be made after radiological evaluation and appropriate tissue diagnosis.

Total sacrectomy is indicated when a malignant or aggressive benign lesion involves the proximal sacrum [41, 42]. Partial sacrectomy which includes transverse, sagittal, or a combination of both can be considered for sacral tumors that lie entirely to one side of the sacrum. According to the transverse axis and sacroiliac joint involvement, sacral tumors are considered to be either high midline lesions (above S3 without lateral invasion of a sacroiliac joint), high lateral lesions (above S3 with sacroiliac joint invasion), and low midline lesions (below S3). Lateral lesions with sacroiliac joint involvement should be treated by sagittal sacrectomy, while high or low midline tumors without sacroiliac joint involvement should be



treated by transverse sacrectomy [15]. These technically demanding procedures require multidisciplinary (neurosurgery, surgical and orthopedic oncology, and plastic surgery) involvement and should only be undertaken at institutions with experience in treating such patients.

---

## 2.8 Radiation Therapy

In those cases where primary complete resection of sacral tumors is difficult because of proximity to neural and vascular structures, radiation therapy may be useful. For sacral metastases, radiation therapy may be the initial treatment of choice, whereas in some cases of primary sacral tumors, conventional radiation therapy may be used in conjunction with surgery as adjuvant treatment (for palliation, prevention of pathological fractures, or to slow progression of or reverse neurologic compromise) [53]. When considering radiation therapy for such patients, it is important to remain cognizant of surrounding structures and to limit radiation doses as appropriate.

---

## 2.9 Embolization

Embolization is a useful adjuvant therapy in the management of sacral tumors. Typically, Gelfoam, alcohol embolizing emulsions, coils, ethanol, and microfibrillar collagen are used for embolization [49, 50, 54–57]. If a vascular sacral lesion is suspected based on presentation and imaging, then preoperative angiography should be performed to characterize the vascular anatomy and to determine if the lesion would be amenable to embolization. Of note, sacral tumors may have significant collateral circulation, and tumor neovascular recruitment may result in the formation of an extensive collateral vascular network [55]. It is recommended that embolization should be performed as close as possible to the time of surgery. Typically, timing of embolization is critical and should be planned carefully in conjunction with surgical resection [55–57]. Also, it is important to note that ischemic neuropathy is a potential complication of any pelvic embolization that can result in motor and sensory deficits in the pelvis and lower extremities. Therefore, care must be taken to identify and avoid embolization of the neurovascular anatomy. Rectal ischemia can result from superior hemorrhoidal artery embolization. Any embolization of sacral tumors may result in injury to nontargeted tissue including muscle infarction, injury to the skin, or injury to the colon or other organs [53].

---

### Conclusion

Primary benign and malignant tumors of the sacrum are rare lesions that account for fewer than 7% of all intraspinal primary tumors. Metastatic lesions, multiple myeloma, and lymphoma are far more common than primary sacral tumors. Patients with sacral tumors present with nonspecific symptoms, including pain, palpable mass, and neurologic deficits. Additionally, the management of tumors of the sacrum is challenging. Radical resection through

partial or complete sacrectomy can prolong the overall survival of patients with primary malignant or aggressive benign tumors; however, it is necessary to establish immediate stability through spinopelvic reconstruction for early ambulation and preservation of the quality of life. While modern radiation therapy and stereotactic radiosurgery have the potential to reduce complications and embolization can be used as an adjunct to surgery, thorough operative planning by a multidisciplinary team is critical to the success of treatment of such lesions.

**Conflict-of-Interest Statement** No benefits have been or will be received from a commercial party related directed or indirectly to the subject matter of this article.

---

## References

1. Feldenzer JA, McGauley JL, McGillicuddy JE. Sacral and presacral tumors: problems in diagnosis and management. *Neurosurgery*. 1989;25:884–91.
2. Diel J, Ortiz O, Losada RA, Price DB, Hayt MW, Katz DS. The sacrum: pathologic spectrum, multimodality imaging, and subspecialty approach. *Radiographics*. 2001;21(1):83–104.
3. Llauger J, Palmer J, Amores S, Bague S, Camins A. Primary tumors of the sacrum: diagnostic imaging. *AJR Am J Roentgenol*. 2000;174(2):417–24.
4. Unni KK. Dahlin's bone tumors: general aspects and data on 11,087 cases. 5th ed. Philadelphia: Lippincott-Raven; 1997.
5. Payer M. Neurological manifestation of sacral tumors. *Neurosurg Focus*. 2003;15(2):E1.
6. Deutsch H, Mummaneni PV, Haid RW, Rodts GE, Ondra SL. Benign sacral tumors. *Neurosurg Focus*. 2003;15(2):E14.
7. Chandawarkar RY. Sacrococcygeal chordoma: review of 50 consecutive patients. *World J Surg*. 1996;20(6):717–9.
8. Cheng EY, Ozerdemoglu RA, Transfeldt EE, Thompson Jr RC. Lumbosacral chordoma. Prognostic factors and treatment. *Spine*. 1999;24(16):1639–45.
9. Yonemoto T, Tatzaki S, Takenouchi T, Ishii T, Satoh T, Moriya H. The surgical management of sacrococcygeal chordoma. *Cancer*. 1999;85(4):878–83.
10. Lin PP, Guzel VB, Moura MF, et al. Long-term follow-up of patients with giant cell tumor of the sacrum treated with selective arterial embolization. *Cancer*. 2002;95(6):1317–25.
11. York JE, Kaczaraj A, Abi-Said D, et al. Sacral chordoma: 40-year experience at a major cancer center. *Neurosurgery*. 1999;44(1):74–80.
12. Althausen PL, Schneider PD, Bold RJ, Gupta MC, Goodnight Jr JE, Khatri VP. Multimodality management of a giant cell tumor arising in the proximal sacrum: case report. *Spine*. 2002;27(15):E361–5.
13. Bergh P, Kindblom LG, Gunterberg B, et al. Prognostic factors in chordoma of the sacrum and mobile spine: a study of 39 patients. *Cancer*. 2000;88(9):2122–34.
14. Manaster BJ, Graham T. Imaging of sacral tumors. *Neurosurg Focus*. 2003;15(2):E2.
15. Nair S, Gobin YP, Leng LZ, Marcus JD, Blisky M, Laufer I, Patsalides A. Preoperative embolization of hypervascular thoracic, lumbar, and sacral spinal column tumors: technique and outcomes from a single center. *Interv Neuroradiol*. 2013;19(3):377–85.
16. Pikis S, Itshayek E, Barzilay Y, Hasharoni A, Kaplan L, Gomori M, Cohen JE. Preoperative embolization of hypervascular spinal tumors: current practice and center experience. *Neuro Res*. 2014;36(6):502–9.
17. Gerber S, Ollivier L, Leclère J, et al. Imaging of sacral tumours. *Skelet Radiol*. 2008;37(4):277–89.

18. Ng EW, Porcu P, Loehrer Sr PJ. Sacrococcygeal teratoma in adults: case reports and a review of the literature. *Cancer*. 1999;86(7):1198–202.
19. Lam CH, Nagib MG. Nonteratomatous tumors in the pediatric sacral region. *Spine*. 2002;27(11):E284–7.
20. O'Neill OR, Piatt Jr JH, Mitchell P, Roman-Goldstein S. Agenesis and dysgenesis of the sacrum: neurosurgical implications. *Pediatr Neurosurg*. 1995;22(1):20–8.
21. Turcotte RE, Sim FH, Unni KK. Giant cell tumor of the sacrum. *Clin Orthop Relat Res*. 1993;291:215–21.
22. Papagelopoulos PJ, Choudhury SN, Frassica FJ, Bond JR, Unni KK, Sim FH. Treatment of aneurysmal bone cysts of the pelvis and sacrum. *J Bone Joint Surg Am*. 2001;83(11):1674–81.
23. Peh WC, Koh WL, Kwek JW, Htoo MM, Tan PH. Imaging of painful solitary lesions of the sacrum. *Australas Radiol*. 2007;51(6):507–15.
24. Wakhlu A, Misra S, Tandon RK, Wakhlu AK. Sacrococcygeal teratoma. *Pediatr Surg Int*. 2002;18(5–6):384–7.
25. Boretz RS, Lonner BS. Atypical presentation of an osteoid osteoma in a child. *Am J Orthop*. 2002;31(6):347–8.
26. Popuri R, Davies AM. MR imaging features of giant presacral schwannomas: a report of four cases. *Eur Radiol*. 2002;12(9):2365–9.
27. Pogoda P, Linhart W, Priemel M, Rueger JM, Amling M. Aneurysmal bone cysts of the sacrum. Clinical report and review of the literature. *Arch Orthop Trauma Surg*. 2003;123(5):247–51.
28. Campanacci M, Baldini N, Boriani S, Sudanese A. Giant-cell tumor of bone. *J Bone Joint Surg Am*. 1987;69(1):106–14.
29. Disler DG, Miklic D. Imaging findings in tumors of the sacrum. *AJR Am J Roentgenol*. 1999;173(6):1699–706.
30. Gong L, Liu W, Sun X, Sajdik C, Tian X, Niu X, Huang X. Histological and clinical characteristics of malignant giant cell tumor of the bone. *Virchows Arch*. 2012;460(3):327–34.
31. Ozaki T, Liljenqvist U, Halm H, Hillmann A, Gosheger G, Winkelmann W. Giant cell tumor of the spine. *Clin Orthop Relat Res*. 2002;401:194–201.
32. Sar C, Eralp L. Surgical treatment of primary tumors of the sacrum. *Arch Orthop Trauma Surg*. 2002;122(3):148–55.
33. Wuisman P, Lieshout O, Sugihara S, van Dijk M. Total sacrectomy and reconstruction: oncologic and functional outcome. *Clin Orthop Relat Res*. 2000;381:192–203.
34. Lackman RD, Khoury LD, Esmail A, Donthineni-Rao R. The treatment of sacral giant-cell tumours by serial arterial embolisation. *J Bone Joint Surg Br*. 2002;84(6):873–7.
35. Randall RL. Giant cell tumor of the sacrum. *Neurosurg Focus*. 2003;15(2):E13.
36. Lath R, Rajshekhar V, Chacko G. Sacral haemangioma as a cause of coccydynia. *Neuroradiology*. 1998;40(8):524–6.
37. Klimo Jr P, Schmidt RH, Schmidt MH. Nerve sheath tumors involving the sacrum. Case report and classification scheme. *Neurosurg Focus*. 2003;15(2):E12.
38. Abernathy CD, Onofrio BM, Scheithauer B, Pairolo PC, Shives TC. Surgical management of giant sacral schwannomas. *J Neurosurg*. 1986;65(3):286–95.
39. Fourny DR, Gokaslan ZL. Current management of sacral chordoma. *Neurosurg Focus*. 2003;15(2):E9.
40. Fleming GF, Heimann PS, Stephens JK, et al. Dedifferentiated chordoma. Response to aggressive chemotherapy in two cases. *Cancer*. 1993;72(3):714–8.
41. Guo Y, Yadav R. Improving function after total sacrectomy by using a lumbar-sacral corset. *Am J Phys Med Rehabil*. 2002;81(1):72–6.
42. Jackson RJ, Gokaslan ZL. Spinal-pelvic fixation in patients with lumbosacral neoplasms. *J Neurosurg*. 2000;92(1 Suppl):61–70.
43. Tomita K, Tsuchiya H. Total sacrectomy and reconstruction for huge sacral tumors. *Spine*. 1990;15(11):1223–7.
44. Bjornsson J, Wold LE, Ebersold MJ, Laws ER. Chordoma of the mobile spine. A clinicopathologic analysis of 40 patients. *Cancer*. 1993;71(3):735–40.

45. Kumar PP, Good RR, Skultety FM, Leibrock LG. Local control of recurrent clival and sacral chordoma after interstitial irradiation with iodine-125: new techniques for treatment of recurrent or unresectable chordomas. *Neurosurgery*. 1988;22(3):479–83.
46. Azzarelli A, Quagliuolo V, Cerasoli S, et al. Chordoma: natural history and treatment results in 33 cases. *J Surg Oncol*. 1988;37(3):185–91.
47. Papagelopoulos PJ, Mavrogenis AF, Galanis EC, Savvidou OD, Boscainos PJ, Katonis PG, Sim FH. Chordoma of the spine: clinicopathological features, diagnosis, and treatment. *Orthopedics*. 2004;27(12):1256–63.
48. Lanzieri CF, Sacher M, Solodnik P, Hermann G, Cohen BA, Rabinowitz JG. Unusual patterns of solitary sacral plasmacytoma. *AJNR Am J Neuroradiol*. 1987;8(3):566–7.
49. Chiras J, Cognard C, Rose M, et al. Percutaneous injection of an alcoholic embolizing emulsion as an alternative preoperative embolization for spine tumor. *AJNR Am J Neuroradiol*. 1993;14(5):1113–7.
50. Shimada A, Sugimoto KJ, Wakabayashi M, Imai H, Seikguchi Y, Nakamura N, Sawada T, Ota Y, Komatsu N, Noguchi M. Primary sacral non-germinal center type diffuse large B-cell lymphoma with MYC translocation: a case report and review of the literature. *Int J Clin Exp Pathol*. 2013;6(9):1919–28.
51. Grubb MR, Currier BL, Pritchard DJ, Ebersold MJ. Primary Ewing's sarcoma of the spine. *Spine*. 1994;19(3):309–13.
52. Shives TC, McLeod RA, Unni KK, Schray MF. Chondrosarcoma of the spine. *J Bone Joint Surg Am*. 1989;71(8):1158–65.
53. Gibbs IC, Chang SD. Radiosurgery and radiotherapy for sacral tumors. *Neurosurg Focus*. 2003;15(2):E8.
54. Gottfried ON, Schmidt MH, Stevens EA. Embolization of sacral tumors. *Neurosurg Focus*. 2003;15(2):E4.
55. Yakes WFJ, Carrasco CH, Luethke JM. Embolization of lumbosacral lesions. In: Doty JR, Rengachary SS, editors. *Surgical disorders of the sacrum*. New York: Thieme; 1994. p. 294–308.
56. Hess T, Kramann B, Schmidt E, Rupp S. Use of preoperative vascular embolisation in spinal metastasis resection. *Arch Orthop Trauma Surg*. 1997;116(5):279–82.
57. Smith TP, Gray L, Weinstein JN, Richardson WJ, Payne CS. Preoperative transarterial embolization of spinal column neoplasms. *J Vasc Interv Radiol*. 1995;6(6):863–9.
58. Biagini R, Orsini U, Demitri S, et al. Osteoid osteoma and osteoblastoma of the sacrum. *Orthopedics*. 2001;24(11):1061–4.
59. Knoeller SM, Uhl M, Gahr N, Adler CP, Herget GW. Differential diagnosis of primary malignant bone tumors in the spine and sacrum. The radiological and clinical spectrum: minireview. *Neoplasma*. 2008;55(1):16–22.
60. Fiandaca MS, Ross WK, Pearl GS, Bakay RA. Carcinoid tumor in a presacral teratoma associated with an anterior sacral meningocele: case report and review of the literature. *Neurosurgery*. 1988;22(3):581–8.
61. Schnee CL, Hurst RW, Curtis MT, Friedman ED. Carcinoid tumor of the sacrum: case report. *Neurosurgery*. 1994;35(6):1163–7.
62. Capanna R, Briccoli A, Campanacci L. Benign and malignant tumors of the sacrum. In: Frymore J, editor. *The adult spine: principles and practice*. Philadelphia: Lippincott-Raven; 1997. p. 2367–405.
63. Papagelopoulos PJ, Currier BL, Shaughnessy WJ, et al. Aneurysmal bone cyst of the spine. Management and outcome. *Spine*. 1998;23(5):621–8.
64. Feigenberg SJ, Marcus Jr RB, Zlotecki RA, Scarborough MT, Berrey BH, Enneking WF. Radiation therapy for giant cell tumors of bone. *Clin Orthop Relat Res*. 2003;411:207–16.

Alexandra Gangi and Ricardo Gonzalez

---

## 3.1 Clinical Presentation

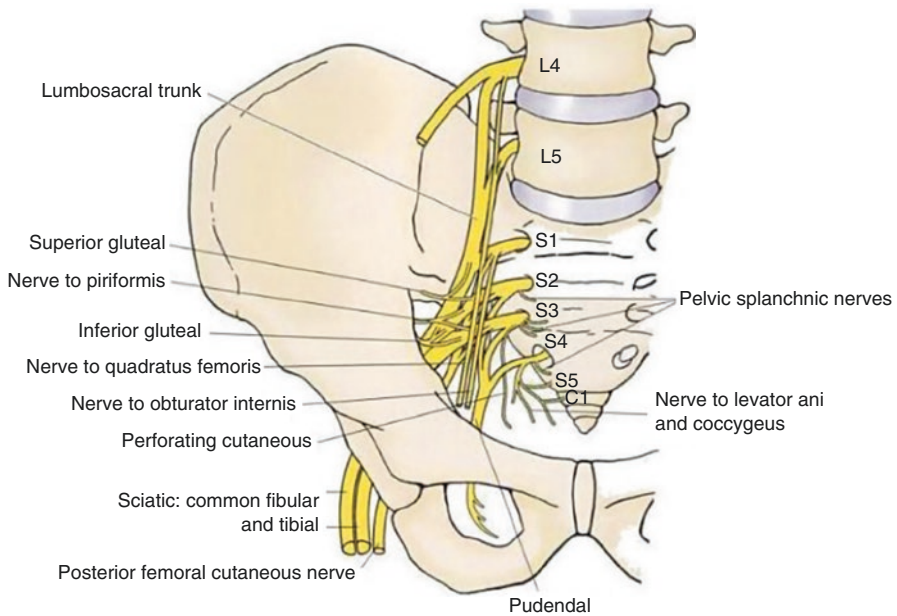
Sacral tumors are generally diagnosed late and can present as large, advanced neoplastic masses because of mild initial symptoms. The clinical pattern depends on the anatomic location of the lesion within the sacrum, its extension, and whether it compresses or invades neighboring structures [1]. The pain may initially be nonspecific and as clinical examination is usually poor, these tumors may remain clinically silent for long periods of time. The most common initial symptom of a sacral tumor is local pain due to its mass effect and compression. Occasionally smaller lesions could become symptomatic secondary to involvement of critical structures, such as nerves or ureters, or because of pathologic fractures. Generally, however, these tumors remain asymptomatic until they are quite large, and lower sacral tumors can grow large enough for their anterior portion to be palpated during a rectal examination [1–3]. While lateral extension of sacral tumors across the sacroiliac joints causes local pain at the joint, invasion of the origin of the gluteus maximus and piriformis muscles leads to local pain and subsequently decreases hip extension and external rotation strength [3–7].

Subsequently, as nerve roots become increasingly compressed or infiltrated by tumor, multiradicular sensory deficits develop and can include radicular pain radiating uni- or bilaterally into the buttocks, posterior thigh or leg, external genitalia, and/or perineum (Fig. 3.1).

As this continues to progress, motor deficits, and eventually, bladder, bowel, and/or sexual dysfunction from anterior extension of the tumor into the presacral space can be noted [1].

---

A. Gangi, M.D. • R. Gonzalez, M.D. (✉)  
Sarcoma Department, H. Lee Moffitt Cancer Center,  
12902 Magnolia Drive, Tampa, FL 33602, USA  
e-mail: [Alexandra.Gangi@moffitt.org](mailto:Alexandra.Gangi@moffitt.org); [Ricardo.Gonzalez@moffitt.org](mailto:Ricardo.Gonzalez@moffitt.org)



**Fig. 3.1** Sacral nerve roots (2016, June). Retrieved July, 2016, from <http://wiki.ahuman.org/index.php/HumanNervesSpinalRoots>

Involvement of lumbosacral nerve roots in sacral lesions leads to certain specific deficits. A lesion involving the L-5 nerve root, commonly in its L5-S1 foraminal or extraforaminal course, may cause radicular pain and hypesthesias in the lateral thigh and calf as well as dorsum of the foot to the great toe [1, 2, 4]. Motor weakness of the L-5 nerve root may result in weakened ankle dorsiflexion, great toe extension, knee flexion, and hip abduction. The straight-leg raise test, or Lasegue's sign, which involves raising the patient's leg with a straight knee while the patient is supine, would result in sciatic pain and render a positive result. A lesion involving the S-1 nerve root, in its canalicular, S1-2 foraminal or extraforaminal course, typically causes radicular pain and hypesthesias in the posterior thigh and calf as well as at the lateral and plantar face of the foot and the small toe. A motor deficit due to an S-1 lesion may result in weakened ankle plantar flexion, knee flexion, and hip extension. A unilateral lesion to the S2 or S3 nerve root usually leads to mild or moderate bladder, bowel, and/or sexual dysfunction [8, 9]. A bilateral lesion of the S2 or S3 roots almost always results in complete bladder, bowel, and sexual dysfunction, and although a unilateral lesion at the same nerve root may cause symptoms, they are generally more nonspecific. However, unilateral or even bilateral lesions of the S4 and/or S5 roots do not result in autonomic dysfunction, although anatomical work has shown some S4 and S5 root contribution to bladder and bowel function [10]. Performance of a thorough physical exam in such patients is critical and can significantly aid in diagnosis and ancillary testing.