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Editors

Bone Tumors

Diagnosis and Therapy Today

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Preface

The volume's two editors have dealt with patients with bone tumors for more than 40 years; Dominique G. Poitout in Marseille, France, and Jaime Paulos in Chile.

The aim of this book is to offer clinical and surgical knowledge from the simplest to the most advanced approach in bone tumors. It provides an actual knowledge for beginners in orthopedics to the most advanced surgical techniques of resection and reconstruction. To complete this aim, collaborators of great experience in bone tumors have contributed chapters in helping to reach our purpose.

We hope this book will assist orthopedic surgeons and residents for teaching and learning about bone tumors.

We must agree that the final decision in the diagnosis and treatment of a particular patient must come from the doctor him/herself or the team in charge of the patient.

Santiago, Chile — Jaime Paulos
Marseille, France — Dominique G. Poitout

Acknowledgements

We would like to thank all the people who made this book possible.

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Our gratitude also goes to Oscar Contreras MD, Professor of Radiology at the Clinical Hospital of Universidad Católica, Chile, who facilitated a great number of images of cases submitted to surgery in the above mentioned institution.

A special word of thanks goes to Prof. Juan Fortune, who with his teachings initiated Prof. Jaime Paulos into the study of bone tumors from the very beginning of his residency.

Finally, we would especially like to thank each and every one of the authors of the chapters, who together with their experience and knowledge contributed to the edition of this book.

Prof. Jaime Paulos, M.D.
Prof. Dominique G. Poitout, M.D.

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Introduction

1 Introduction

Jaime Paulos

Abstract

This chapter discusses the low frequency of bone tumors present in a general hospital, the importance of knowledge about them, their classification and methods of diagnosis, and the complementary study tests.

Keywords

Bone tumors • Orthopedic surgery

This book is focused on bone tumors. We will examine:

- the diagnosis
- the clinical and radiological aspects
- the therapy.

Sometimes it is very difficult to precisely diagnose a bone tumor, and so especially for children we have an obligation to always consider it and to give a diagnosis only when we are sure of it. Also the adapted treatment is given only if we are totally sure of our diagnosis. The diagnoses with their differences and numerous images have been included to explain them. We must have the results of the surgical biopsy.

The clinical study, the radiological exams, can only give an orientation but no more than that. Many different tumors have very similar aspects. We have insisted on their differences.

We have chosen the usual histological classification: the Jaffe and Lichtenstein classification, [1, 8] which is an histological one, but it could be more interesting to use a genomic classification, especially for certain types of tumors such as giant cell tumors or Ewing's sarcomas (World Health Organization Classification).

We have proposed different types of treatment following a perfect extracompartimental resection. Chemotherapy and/or radiotherapy can be used before the surgery and after if it is necessary to destroy the tumoral cells which can be disseminated throughout the body.

Some years ago the only way to treat these malign tumors was by a major amputation; now, most of the time we can make, after a cancerological resection, a massive reconstruction with new techniques such as human cryopreserved bone stored in liquid nitrogen at −196°, or a massive metallic prosthesis made of titanium porometal directly cut in the operating room.

Bone tumors are a special chapter in orthopedics due to their high importance and low frequency of cases. Their high importance relates to the diagnosis of a bone tumor being always traumatizing for the patient. In a general hospital with orthopedic services no more than 3–5% of the orthopedic patients will be diagnosed with a bone tumor. Of course, centers dedicated to cancer and bone tumors concentrate this pathology and will develop the diagnoses, treatments and research on them.

Different levels of expertise about bone tumors in the physicians are necessary. General practitioners must know about the existence of the different types of bone tumors and it will often be the first physician to suspect a bone tumor in a patient and so, send the patient to an orthopedic surgeon. Orthopedic surgeons must know about the correct diagnosis and send this patient to a center or to a surgeon who treats bone tumors. And in addition to this, these centers must be carrying out research in this field.

In our personal experience, we see that very often the patient is not correctly oriented and is treated without a diagnosis and with unuseful treatments for his pathology, for example with anti-inflammatory drugs, physiotherapy, alternative therapies, etc.

Bone tumors have been studied at the Hospital of the Catholic University of Chile since 1962 with the formation of the National Register of Bone Tumors [4], where most of the bone tumors of the country were studied from the clinical, radiologic and anatomopathologic aspects, and providing guidelines for treatment when patients were not

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treated in our own hospital. From this group of cases in the register, benign bone tumors made up 38% of cases, malignant bone tumors 20%, pseudotumoral lesions 35%, and metastasis 7% (the data exclude patients with known primary tumors). This data can give the reader an idea of the low frequency of bone tumors in a general hospital.

In our study approximately half of the total of primary bone tumors are malignant bone tumors and a third of the total are pseudotumoral lesions. In a study of 3345 bone tumors and tumor-like lesions, the distribution of them was the following:

Benign bone tumors

- Osteochondroma 544
- Chondroma 242
- Giant cell tumors 189
- Chondroblastoma 88
- Osteochondromatosis 50
- Hemangioma 49
- Chondromatosis 35
- Osteoma 34
- Fibroma 28

Malignant bone tumors

- Osteosarcoma 299
- Chondrosarcoma 130
- Ewing's Sarcoma 117
- Myeloma 107
- Malignant fibrous histyocitoma 37
- Limphoma primitive of bone 46
- Fibrosarcoma 23
- Parosteal osteosarcoma 19
- Adamamtinoma 11

Metastasis 269
Pseudotumoral lesions

- Unicameral cyst 303
- Osteomyelitis 300
- Metaphyseal fibrous defect 189
- Aneurysmal bone cyst 159
- Osteofibrous dysplasia 140
- Histiocytosis X 127
- Myositis ossificans 51

Others

- Osteopoiquilosis 2
- Hydatidosis osea 4
- Tumoral calcinosis 3
- Osteopetrosis 3
- Paget's disease 6
- Hyperparathyroidism 4
- Charcot arthropathy 2
- TBC (in bone) 4

Clinical Study of Bone Tumors

Bones can be affected with neoplastic lesions through three different mechanisms [1]:

1. Bone tumors which arise from cells or tissues of the bone considered like an organ. These are the primitive bone tumors.
2. Bone tumors coming from a non-osseous malignant tumor, called secondary bone tumors or metastasis.
3. Tumors that invade the bone like an extension of a malignant tumor around the bone.

Primitive Bone Tumors

The cells which form the different tissues of the bone like an organ have the capacity of forming a bone tumor as osteocytes, chondrocytes, fibroblastic cells, myeloreticular tissue, vascular tissue, fat cells, etc. These cells have the potential of forming bone tumors. On this basis Jaffe and Lichtenstein classified bone tumors according to a histogenetic classification. In that way bone tumors are classified like a series of osteogenetic, chondroblastic, fibroblastic, myeloreticular, fat, vascular, etc., bone tumors. However, there is not always only one type of cell present, and the histological diagnosis can become more complex.

Examples of this situation are the chondromyxoid fibromas, dedifferentiated osteogenic sarcomas, pleiomorfic sarcomas, etc. Sometimes the cells are so undifferentiated that the histologist cannot discover the origin of the tumor. Today, the best studies using histopathology, histochemistry techniques, and genetics studies can differentiate new lineages of bone tumors. Also sometimes the differentiation of a benign bone tumor and a malignant one is not easy, and the families of tumors whose origin are cartilage or fibrous cells or like the giant cell tumors are examples of this fact.

The basis of a final diagnosis depends on the conjunction of the clinical history of the patient, the imaging, and the histopathological study. It is necessary at this point to insist on this trio of methods for the diagnosis, and although sometimes one of them makes the final diagnosis the sum of these three elements are decisive for it. The clinical study depends on a good anamnesis and a physical examination and most of the time this tool provides a very good orientation to think in a bone tumor or not.

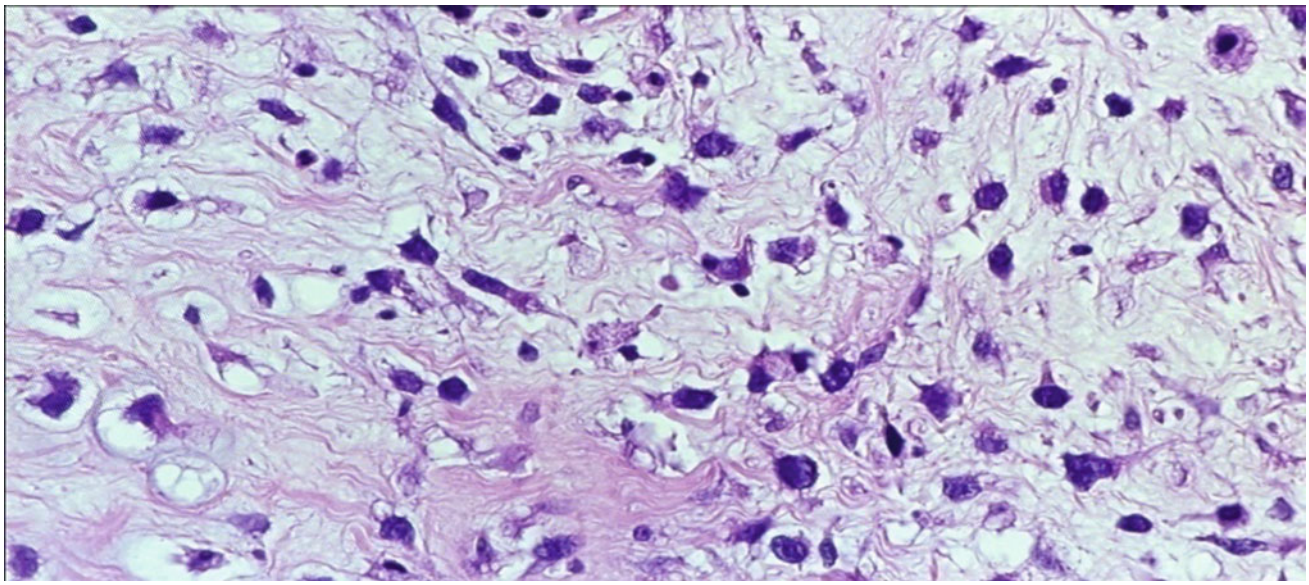

Fig. 1.1 Histology of a chondrosarcoma

Bone tumors in general are closely aligned to age. Specific bone tumors, most of them, appear between determinated ages and only very occasional cases appear out of these age ranges. For example, a primitive osteosarcoma frequently appears in adolescents or young people and it is rarely seen over the fifth decade of life. When it appears in later life it is an osteosarcoma secondary to an irradiated bone, or Paget's disease, or varieties of it like parosteal osteosarcoma or a dedifferentiated sarcoma. Giant cell tumors are very rare before 15 years old, being more frequent between the third decade and fourth decade of life. Chondrosarcoma (Fig. 1.1) is more frequent between the fourth and fifth decade of life. Myeloma, like metastasis, is more frequent after the fifth decade. And so, each bone tumor is presented between determinated ages and therefore is a great tool for the diagnosis.

A bone lesion of malignant aspect on a patient younger than 30 years is suspicious of a primitive bone tumor. On the other hand, in a patient older than 50 years the clinician must think of metastasis or myeloma (Fig. 1.2).

Symptoms and Signs of Bone Tumors

Symptoms that may be helpful for the diagnosis of a bone tumor are: (1) pain (2) swelling (3) functional disability (i.e., limping) (4) spontaneous fracture.

Although these signs are unspecific and can be found in many musculoskeletal diseases, clinical experience shows that patients with these findings appearing without a logical reason or a traumatic event may be affected by a bone tumor. In any of these cases the clinical physician must order an X-ray of the affected segment as the first laboratory examination.

Pain

Mostly the patient feels the pain on the site of the tumoral lesion. Sometimes the pain is related to a joint. In this case, it is representative of epiphysial bone tumors, for example giant cell tumors or juxtarticular bone tumors like osteosarcomas, typically located in the metaphysis of long bones. In the spine, metastasis, myeloma or hemangioma are most frequently presented with axial or radicular pain. Many times the pain is dull, not a heavy pain, and can be well tolerated by the patient for a long time. We very often see this kind of pain in benign tumors like osteochondromas (Fig. 1.1), enchondromas or also in the intraosseous growth of a malignant bone tumor. In those last cases, when the growth of the tumor reaches the periosteum, the pain becomes more intense and precise.

For this reason, bone tumors can be silent and ending in a late diagnosis.

Swelling

Benign bone tumors can grow slowly and without local signs. On the other hand, fast-developing tumors like

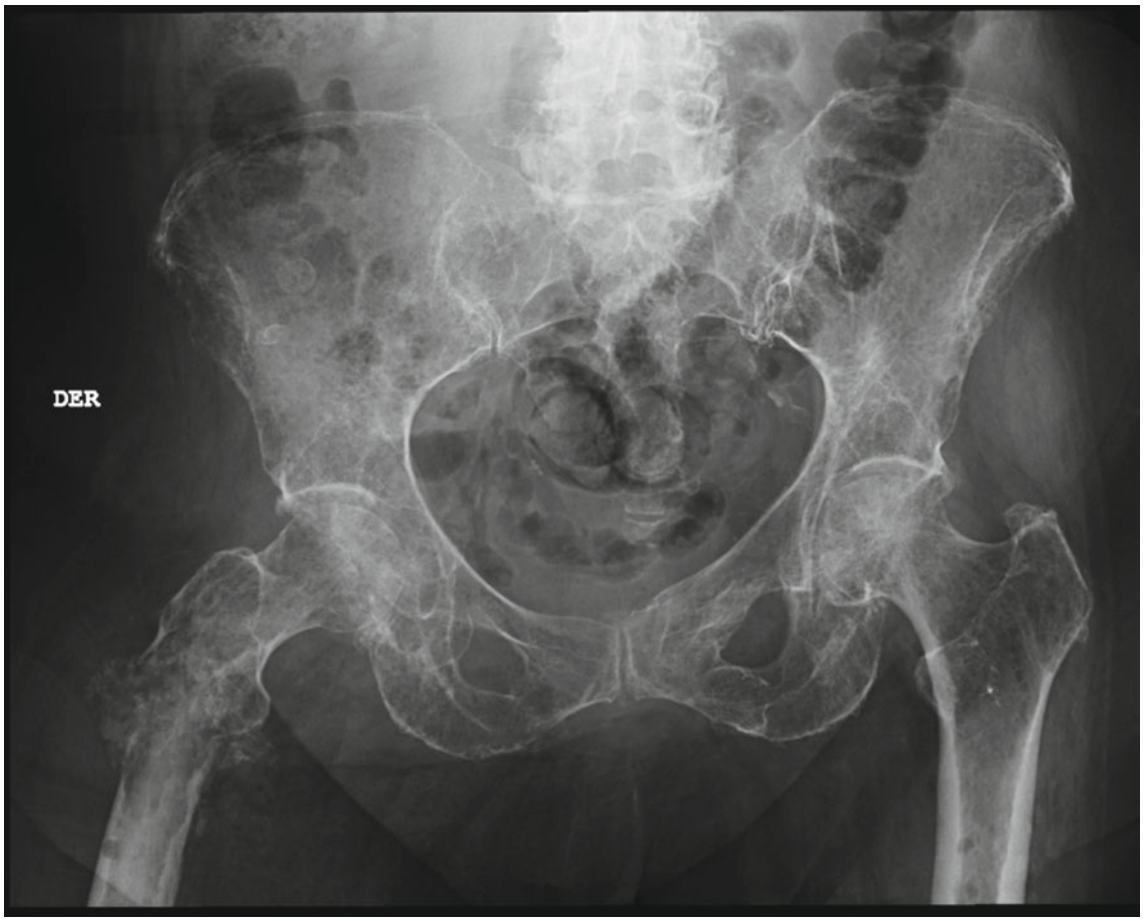

Fig. 1.2 Myeloma pelvis X-ray

Ewing's sarcoma or osteosarcoma quickly develop pain and swelling. This makes the patient seek clinical assistance sooner. In these cases the clinician must be aware of these signs as being suspicion of a tumor.

Disability

The disability produced by pain and sometimes by inflammatory signs can be clear signs of an aggressive bone tumor (i.e., bone tumor of giant cells or Ewing's sarcoma).

The diagnosis of bone tumors is frequently made late because these findings and symptoms may not be present, or only with a very low intensity. For these reasons the patient does not look for medical attention and on the other hand, the physician does not take in to consideration the signs and symptoms of the patient or doesn't think of a bone tumor as a differential diagnosis.

Sometimes bone tumors are found on an X-ray made for another reason (Fig. 1.3).

Spontaneous Fractures

These are produced by a weak structure of bone typically seen in osteoporotic fractures and bone tumors and pseudotumoral bone lesions (Fig. 1.4). There is no relationship between the energy involved and the resulting fracture. Usually the energy involved would be insufficient to achieve this fracture. An example of this is a patient that throws an object and suffers a humeral fracture or a patient that walking or by jogging suffers a lower extremity fracture. We can see this in children with bone cysts or osteolytic bone tumors in adults [7].

Diagnosis of Bone Tumors

The efficiency of the final diagnosis with only the clinical behavior of bone tumors in general is poor. However, the suspicion of the existence of a bone tumor should be very high.

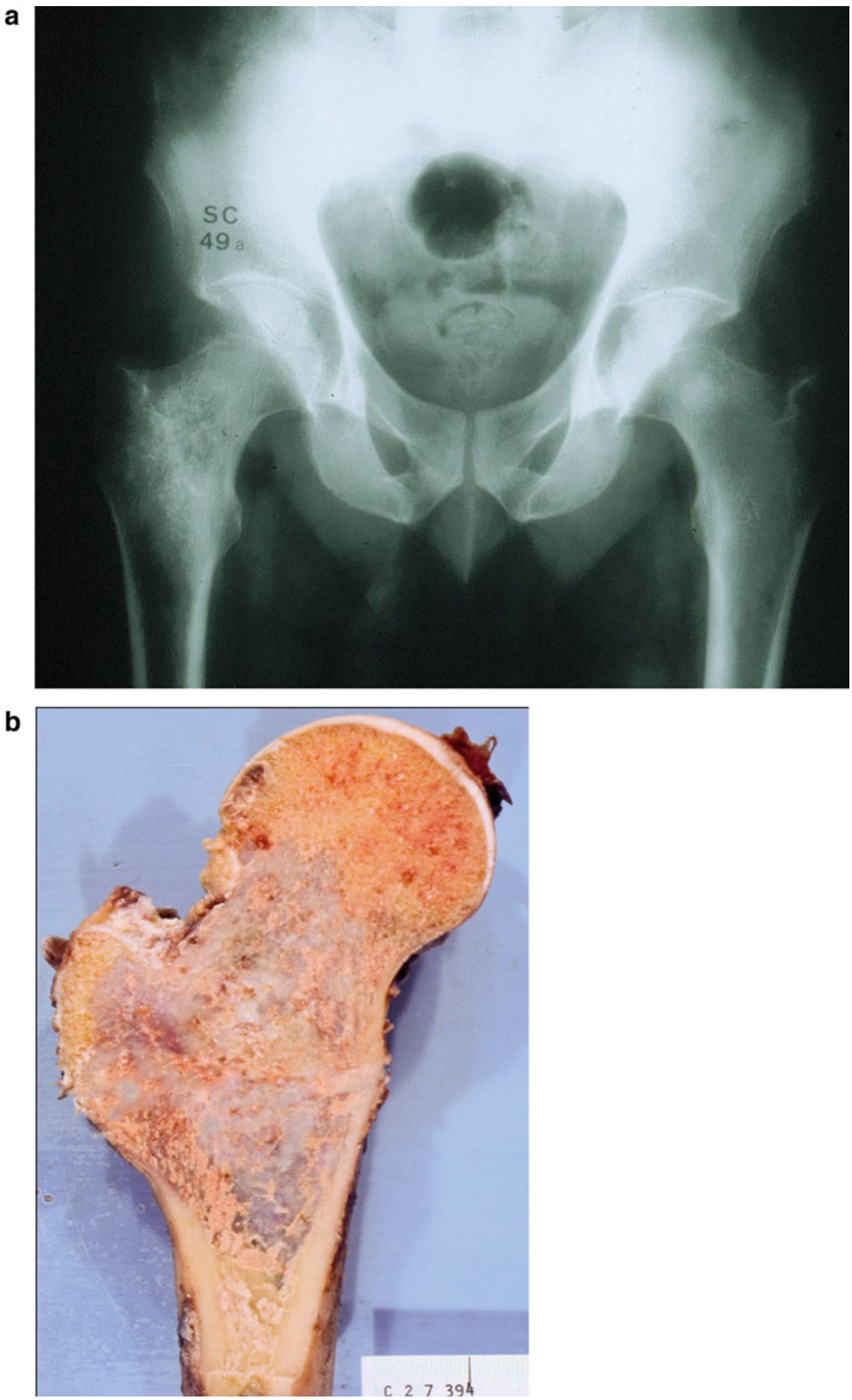

Fig. 1.3 Asymptomatic chondrosarcoma found after a hip contusion **a** radiological aspect **b** macroscopic aspect

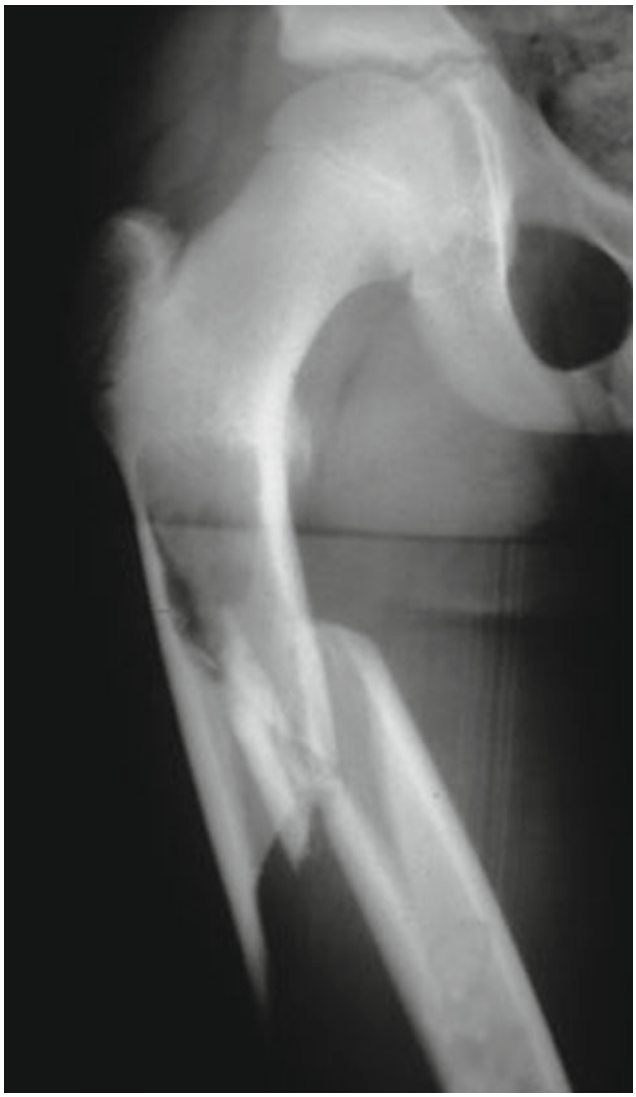

Fig. 1.4 A 12 year old patient with pathological fracture in an unicameral bone cyst

If the general physician is the first person to see the patient, he must correctly drive the patient to the final diagnosis.

The second step for the diagnosis of bone tumors is the imaging studies [9]. The first image must be a conventional X-ray of the segment where the physician thinks the bone tumor is located. There are a lot of typical images of bone tumors, but there are no pathognomonic images of a bone tumor. Also a false image can be found (Fig. 1.5).

With these two elements, clinical findings and X-rays, it is possible to achieve a hypothetical diagnosis, but the final diagnosis must be confirmed with a bone biopsy. Secondary studies must also be made such as scanner (CT, computed tomography), MRI which will give precise data about intraosseous extension and soft tissues around the tumor (Fig. 1.6). Scintigraphy (with Tc 99) or PET-CT (positron emission tomography) will be useful for the study of polyostotic lesions or the spreading of bone metastasis.

However, the histology studies must also be correlated with clinical and radiological findings.

A classic sample of this is the histopathological finding in myositis ossyficans (Fig. 1.7) and osteosarcoma. Both microscopic features can be very similar and for the final diagnosis of the patient's clinical history will be definitive for making the diagnosis. In brief, the final diagnosis must be made with the participation of the clinical orthopedic surgeon, the radiologist and the pathologist.

Brief of the Procedure of Diagnosis

The suspicion of the diagnosis of a bone tumor begins with the physician that has contact with the patient for the first time; then a radiological study must be done and finally the biopsy study by the pathologist. The physician, not the specialist, should be able to think of the diagnosis of a bone tumor and then send the patient to the correct place for the correct final diagnosis. The biopsy and the interpretation of the whole case must be in the hands of a specialist; however, the general physician must think in a tumoral bone lesion to continue the correct process. A lot of time could be wasted if the general physician would not consider a bone tumor and so the patient would not receive the correct diagnosis.

Then, the radiologist discovers the presence of a lesion and gives its description, proposes if it is a benign or malignant lesion and makes a diagnosis (Fig. 1.8).

A specialist surgeon dedicated to bone tumors must do a correct biopsy, taking a good piece of tissue to identify the tumor lesion.

Malignant bone tumors like Ewing's Sarcoma, osteosarcoma, fibrosarcoma, malignant fibrous histiocytoma and myeloma are some of the most malignant bone tumors of our body.

We cannot wait for a spontaneous change in the lesion since almost always they continue to grow, and perhaps the identification of the tumor does not reflect its potential growth rate (Fig. 1.9).

Some conditions could reflect a more severe bone tumor lesion. There are some very aggressive bone tumors which are a condition of their biological behavior, for example, the aggressiveness of giant cell bone tumors, although they are classified as benign bone tumors.

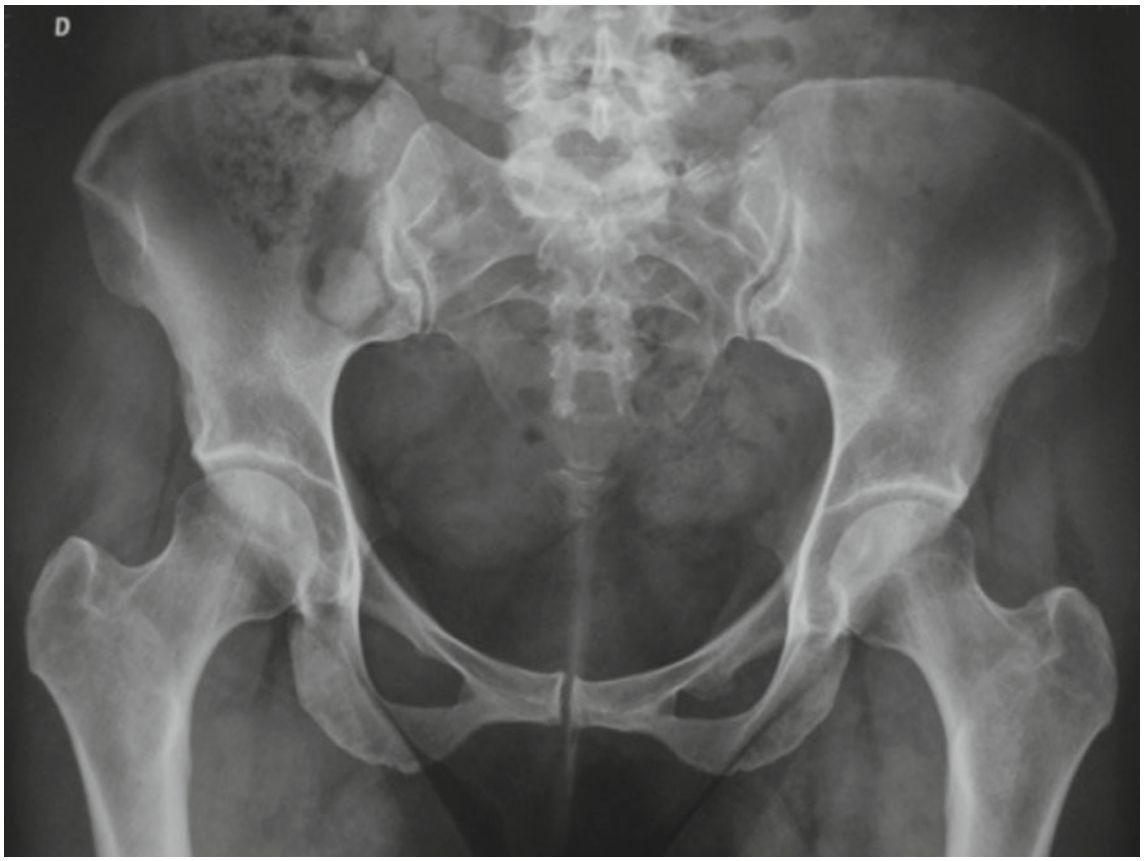

Fig. 1.5 False image of gas inside a bowel simulating an osteolytic bone lesion

The size of the bone tumor can also be a feature for prognosis condition. Big tumors are more difficult to remove.

The location of the tumor is also an important factor. A benign tumor in a critical place can be very dangerous for the patient. The invasion of the tumor is also a bad prognosis and making a resection very difficult or impractical.

Bone Biopsy

Two possible ways to make a bone biopsy are [5]:

(1) Open surgical biopsy
(2) A needle biopsy. The biopsy size must be the correct one for the histological interpretation (Fig. 1.10).

Inside a bone tumor it is possible to find different types of tumor tissues, for example, in a dedifferentiated sarcoma you can find tissues of a chondrosarcoma and also of an osteosarcoma. The incision of the biopsy must be made in such a way that it can be excised, when the definitive resection of the whole tumor is done.

The surgeon performing the biopsy must be familiar with incisions of limb salvage surgery.

Many surgeons prefer an open biopsy to be sure that they will take a good amount of tissue for the histological study. A biopsy can be done with a needle biopsy, but sometimes that can be insufficient. However, in experienced hands core biopsy can provide an accurate diagnosis in 90% of cases.

Classification of Bone Tumors

The World Health Organization (WHO) has made an effort to classify bone tumors. Nowadays, is the most useful classification as described below [6].

1. **Bone Tumors Forming Bone Tissue**
   – BENIGN Osteoma
     Osteoid osteoma
     Osteoblastoma

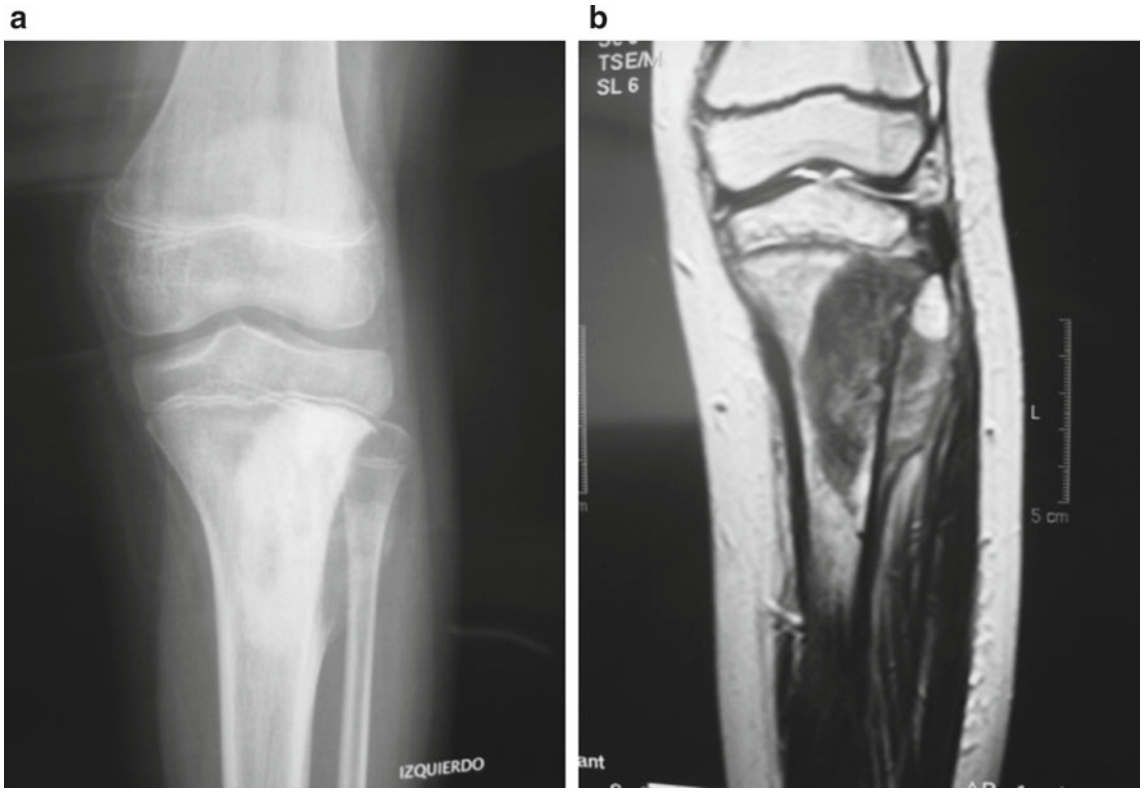

Fig. 1.6 Radiological aspect of an osteosarcoma **a** X-ray **b** MRI

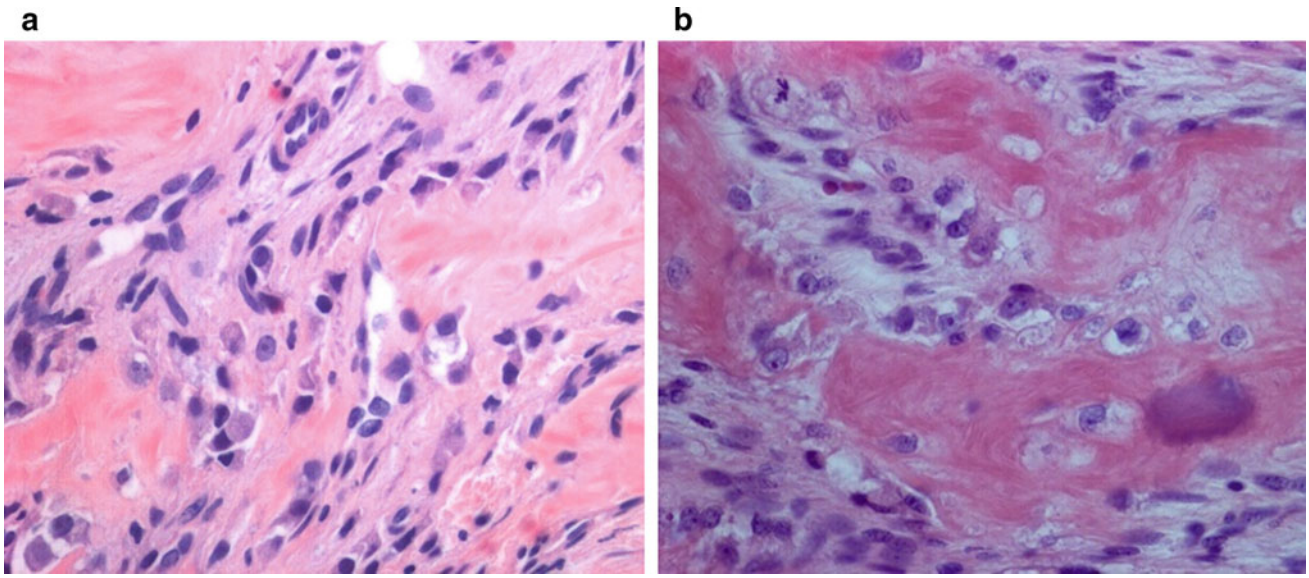

Fig. 1.7 Histologic aspect of an osteosarcoma: **a** osteoid tissue with cytological atypia without mitosis related to a myositis ossificans or an osteosarcoma **b** typical histopathology of an osteosarcoma with typical tumoral cells with mitosis

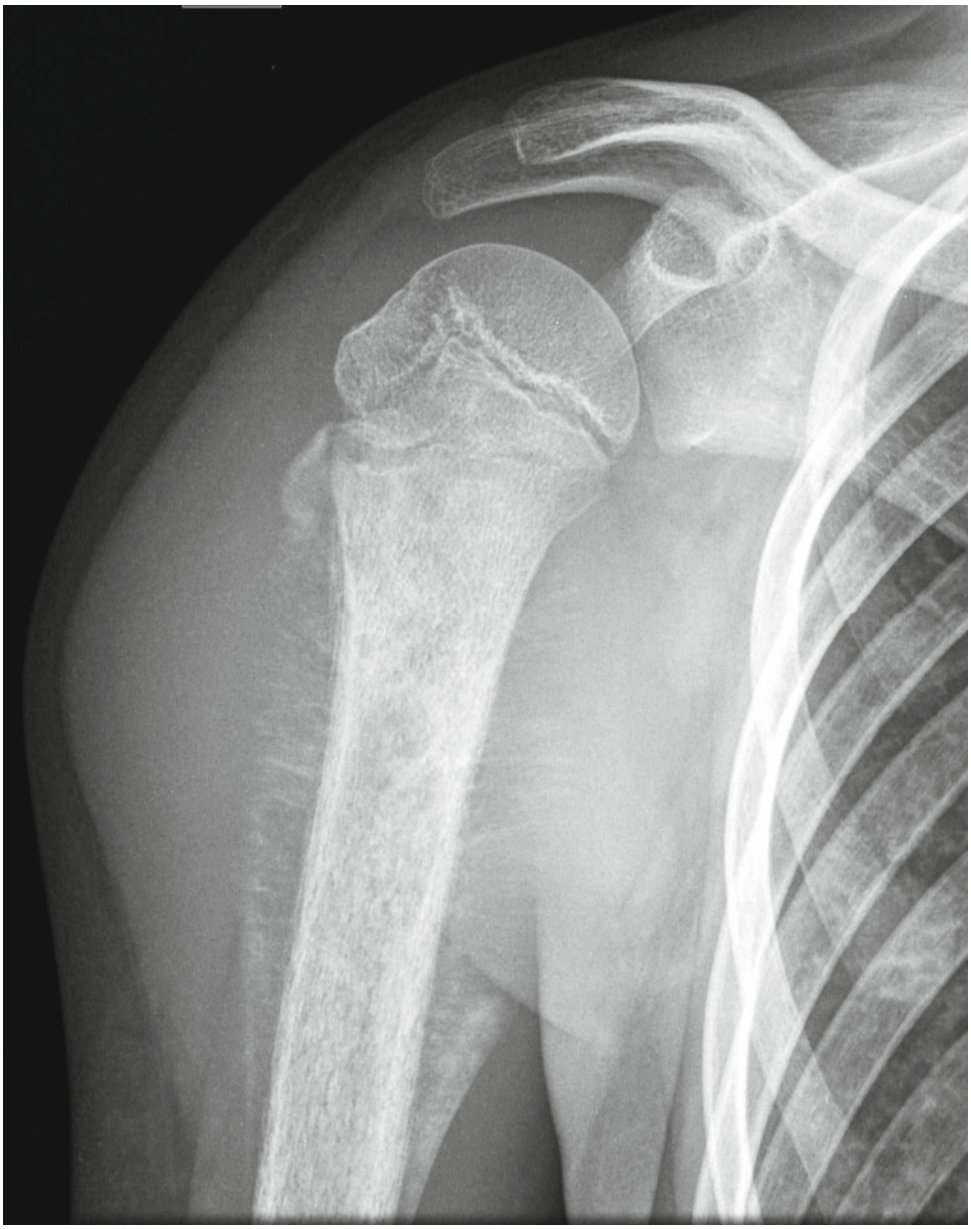

Fig. 1.8 Bone sarcoma showing periosteal reaction, radiated image and Codman's triangle, all signs of a malignant lesion

- MALIGNANT
  Osteosarcoma

2. **Bone Tumors Forming Cartilagenous Tissue**
   - BENIGN
     Chondroma
     Osteochondroma
     Chondroblastoma
     Chondromyxoid fibroma
   - MALIGNANT
     Chondrosarcoma
3. **Giant Cell Tumors**
4. **Bone Marrow Tumors**
   Ewing's sarcoma and reticulosarcoma
   Lymphosarcoma
   Myeloma
5. **Vascular Tumors**
   Hemangioma
   Hemangiosarcoma
6. **Tumors of Connective Tissue**
   Desmoid fibroma
   Lipoma
   Fibrosarcoma
7. **Other Bone Tumors**
   Chordoma
   Adamantinoma
   Neurofibroma

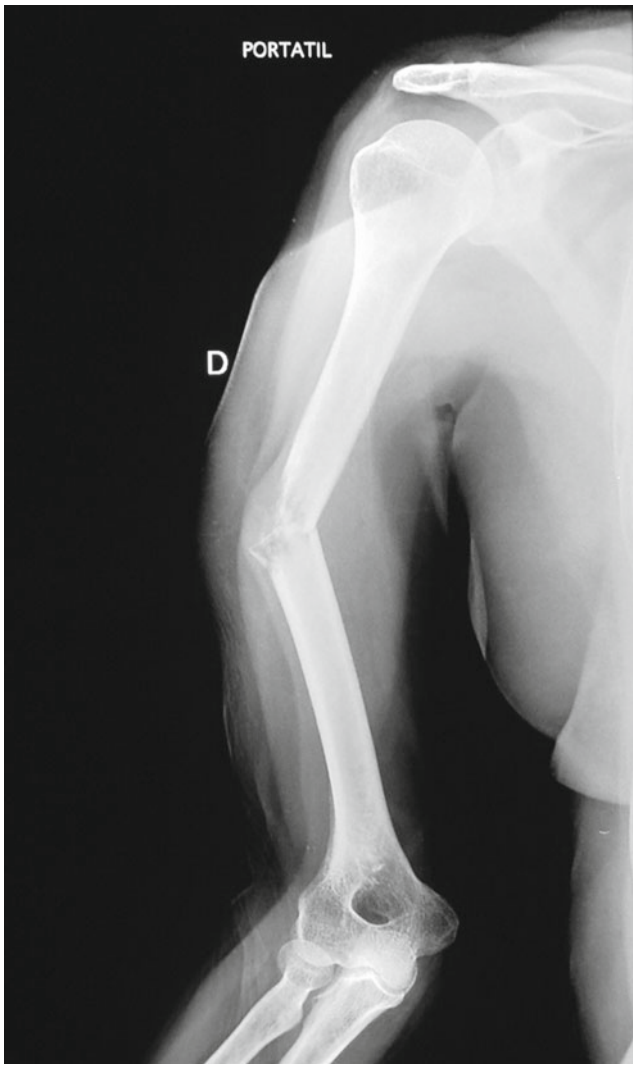

Fig. 1.9 Pathological humerus fracture on breast metastatic carcinoma

8. **Pseudo-Tumoral Lesions**
   Unicameral bone cyst
   Aneurysmal bone cyst
   Juxta-articular cyst
   Metaphyseal lagoons
   Eosinophylic granuloma
   Fibrous dysplasia
   Ossyficant myositis
9. **Secondary Cancer of Bone**
   Metastasis

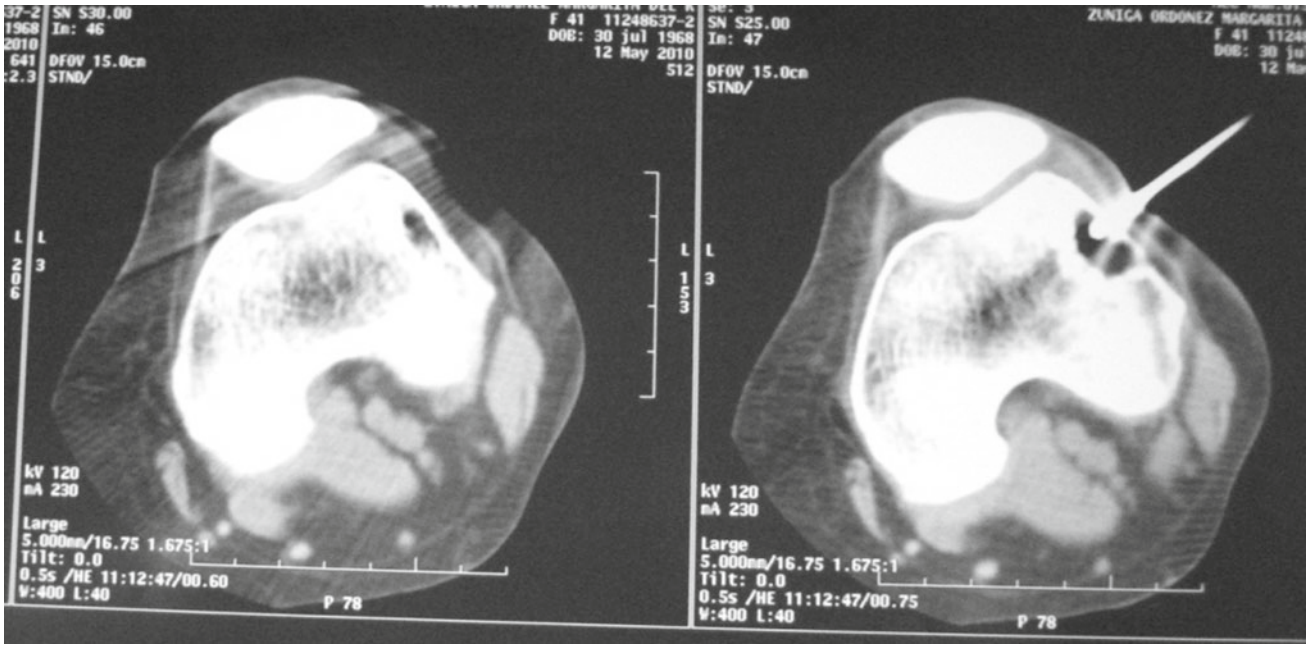

Fig. 1.10 TAC assisted biopsy

Staging

The most well-known staging of malignant bone tumors is the Enneking [2, 3] grading system, classifying it in three main stages:

Stage	Grade	Site	Metastasis
IA	low grade	intracompartment-T1	M0 (none)
IB	low grade	extracompartment-T2	M0
IIA	high grade	extracompartment-T2	M0
IIB	high grade	extracompartment-T2	M0
IIIA	Metastatic	intracompartment-T1	M1 (regional or distant)
IIIB	Metastatic	extracompartment-T2	M1 (regional or distant)

For example, Fig. 1.11 represents a humerus radiography, where a heterogeneous lesion that goes through the cortical bone can be seen; according to the Enneking classification it would be in stage IIB.

Another system for staging bone tumors is the AJCC (American Joint Committee on Cancer) or TNM System. This system is based on the origin and the tumor size, lymph node involvement and the presence or absence of metastasis as follows (Table 1.1):

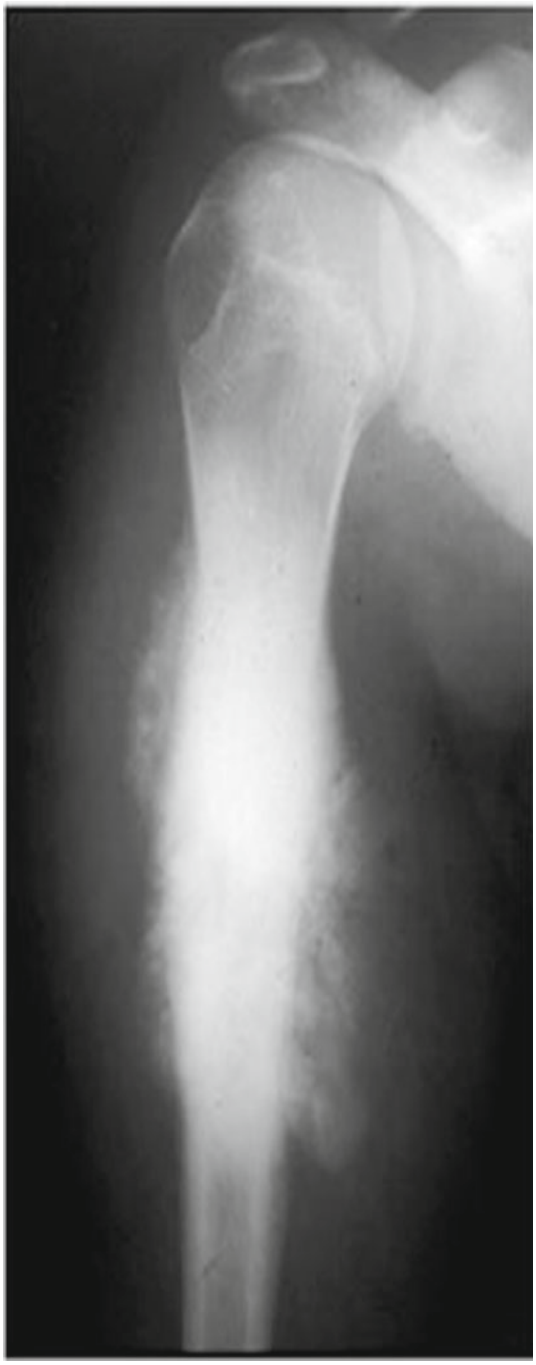

Fig. 1.11 Osteosarcoma stage IIB (high grade, extracompartment)