Special Clinical Aspects of Certain Bone Tumors and Tumor-Like Lesions

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2.1 Introduction

A bone tumor is an abnormal growth of cells within the bone, which can be classified as benign bone tumor, malignant bone tumor, or pseudotumoral lesions. The primary care physician should make a correct diagnosis in a timely manner, relying on a thorough medical history and physical examination. Then multidisciplinary interactive work between imaging specialists, orthopaedic surgeons, oncologists, pathologists, and internists will help to reach a final diagnosis.

2.2 Reason for Consultation

The initial symptom of bone tumors is usually pain that fails to improve with rest, tends to awaken the patient at night, and usually becomes constant. Patients say that the pain yields for a few hours after taking analgesics. Such is the case with osteoid osteomas, which present with prevalence of night pain that responds to salicylates. In high-grade sarcomas, which tend to spread rapidly, the pain develops before the physical manifestation of the tumor. On occasion, bone tumor manifestations are very poor, as is often the case with benign tumors, but they can grow and compress adjacent tissues, causing pain. Generally, pain diminishes with salicylates, but sometimes even opiates are required.

2.3 The Tumor

At consultation, it is possible that the presence of a palpable mass may be manifest. The tumor can cause pain and lead to joint limitations and/or muscle atrophy. Sometimes the lump evolves, producing transient pain relief as the progression of the tumor breaks the cortical barrier and reduces the intraosseous pressure. Soft tissue sarcomas may develop without pain or functional impotence, however, even though a palpable mass can be evident.

Before ordering a diagnostic test, three important issues should be addressed:

- The patient's genetic background
- The patient's age, as each kind of bone tumor has a preference for certain ages of presentation
- The location of the lesion and its characteristics, including adherence to upper or lower levels, infiltration of subcutaneous tissue, and blood circulation

All these characteristics are important in evaluating the behavior of the tumor.

2.3.1 Age of Presentation

Tumors arising in the pediatric population are mostly benign, and the incidence of primary malignant bone tumors is low in patients younger than 40 years. In those over 40 years of age, bone tumors are usually multiple myeloma or metastasis (Table 2.1).

2.3.2 Tumor Location

The anatomic location of the lesion is relevant clinical information in approaching a diagnosis. The affected bone and the region around the bone where the lesion is located (segment) should be considered, as well as whether the

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Table 2.1 Classification of tumors according to age

Patient's age	Benign bone tumor	Malignant bone tumor
Under 5 years	Eosinophilic granuloma Essential bone cyst Fibrous dysplasia	Neuroblastoma, metastatic Ewing's sarcoma
5–10 years	Osteoid osteoma Aneurysmal bone cyst Essential bone cyst Osteochondroma	Osteosarcoma Ewing's sarcoma
10–20 years	Chondroblastoma Osteoid osteoma Osteochondroma	Osteosarcoma Ewing's sarcoma
20–30 years	Giant cell tumors Osteochondroma	
30–40 years	Giant cell tumors	Chondrosarcoma Fibrosarcoma
Over 40 years	Angioma	Myeloma Bone metastasis Chondrosarcoma Chordoma

Table 2.2 Tumors and anatomic location

Anatomic region	Tumors
Tibia	Adamantinoma Chondromyxoid fibroma
Sacral bone	Chordoma, chondrosarcoma, lymphoma, aneurysmal bone cyst, giant cell tumors
Pelvis	Metastasis, myeloma, chondrosarcoma, aneurysmal bone cyst, sarcoma (Paget's disease), Ewing's sarcoma, giant cell tumors
Chest wall	Metastasis, myeloma, chondrosarcoma, fibrous dysplasia
Long bones	Chondrosarcoma, osteosarcoma (especially in distal femur and proximal tibia), periosteal chondrosarcoma, osteochondroma
Calcaneus	Intraosseous lipoma, chondroblastoma, osteosarcoma, solitary bone cyst
Small bones of hands and feet	Enchondroma, subungual exostosis
Vertebral bodies	Metastasis, multiple myeloma, sarcoma (Paget's disease), hemangioma, giant cell tumor
Vertebral, posterior elements	Osteoid osteoma, osteoblastoma, aneurysmal bone cyst

tumor is central, eccentric, or cortical in relation to the bone, and whether it affects blood circulation. Bone tumors and pseudotumoral lesions have a predilection for long bones. For example, lesions such as the enchondroma, essential bone cyst, osteosarcoma, and chondrosarcoma, among others, prefer the metaphyseal bone area. Ewing's sarcoma has a high incidence as a pelvic tumor; in long bones, it is preferably located in the diaphysis, as are myeloma, adamantinoma, and solitary lymphoma. Chondroblastomas and giant cell tumors tend to be located in an epiphysis (Table 2.2).

2.3.3 Tumor Behavior

Benign bone tumors have cells with a tendency towards maturation and, in general, are delimited with respect to neighboring tissues. Complete resection achieves a cure without recurrence. Malignant tumors, on the other hand, tend to grow quickly and in a disorderly fashion, infiltrating adjacent tissues and often having a propensity to spread as lung metastases. The bone represents a favorable environment for the location and growth of metastatic tumors. Most frequent metastases are from breast, prostate, lung, thyroid, or renal carcinomas. Physical examination should include the evaluation of lymph nodes, a potential site of metastasis.

2.4 Complementary Tests

The gold standard for the diagnosis of bone tumors is the conventional X-ray of the lesion. It is a simple, non-invasive, economical study, and establishes the basic criteria to diagnose a benign or malignant lesion, because some radiographic features help to determine the lesion behavior. In the analysis of the image, factors to consider include anatomic location, opacity, margins, mineralization, size and number of lesions, periosteal reaction, transition zone (which defines the boundary between the tumor and adjacent normal bone), and the best site for histopathological study. The biopsy will provide the final diagnostic confirmation. Complementary studies allow precise selection of the biopsy area.

In conclusion, it is essential to correlate clinical data, images, and histopathological information to reach an accurate diagnosis. Modern imaging studies include computed tomography (CT), magnetic resonance imaging (MRI), positron emission tomography (PET), and angiography, as well as modern laboratory tests.

2.4.1 CT Scans

CT scans are of great benefit to identify cortical rupture and penetration towards soft structures. It also provides useful topographical information for biopsies.

2.4.2 MRI

MRI is essential to assess the stage of a lesion and its location. In a suspected primary malignant bone tumor, this study is indicated to fully assess the affected bone. Thanks to its resolution, it is possible to observe neoplastic tissue extension, especially as it affects bone marrow, and to detect whether it compromises neurovascular structures.

2.4.3 Bone Scan

^{99m}Tc-MIBI is usually used to assess malignant tumor lesions of the skeleton. Bone lesions in multiple myeloma may be normal in a scan view, however, so this may not be a recommended diagnostic method in this case.

2.4.4 PET/CT

These two imaging tests in a single study, made with the administration of an ^{18}F-FDG (fluorodeoxyglucose) radioisotope, are of great importance to identify the stage of a lesion and to evaluate the response to chemotherapy. These studies can also disclose metastatic activity.

2.4.5 Angiography

This diagnostic method shows the relationship of the tumor blood vessels and vascular abnormalities. This study may help in the surgical plan and in assessing the response of the tumor to chemotherapy.

2.4.6 Laboratory Tests

The lab provides important elements in the recognition of some pathologies. It should be noted that tumors may present with anemia due to bone marrow infiltration. The elevation of lactic dehydrogenase in patients with Ewing's sarcoma is usually associated with a poor prognosis, as are an increase of serum calcium and bone alkaline phosphatase. The increase of bone alkaline phosphatase is a manifestation of osteoblastic action.

Protein electrophoresis alteration and the presence of Bence-Jones protein suggest multiple myeloma, the most common primary bone malignancy. Alkaline phosphatase can be normal, but high levels of β2-microglobulins are important prognostic factors. The presence of hyperkalemia occurs in 20–40% of cases of multiple myeloma.

Tumor markers such as prostatic antigen (PSA), thyroglobulin, Ca125, Ca15-3, and carcinoembryonic antigen (CEA) are useful for the follow-up of the corresponding primary tumors.

2.5 Preneoplastic Lesions

Preneoplastic lesions occur in patients with bone manifestations that have greater or lesser risk of developing a bone neoplasm. High-risk preneoplastic syndromes include Maffucci syndrome, enchondromatosis (Ollier disease), familial retinoblastoma syndrome, and Rothmund–Thomson syndrome. Multiple osteochondromatosis and Paget's disease should be considered moderate-risk lesions, as is osteitis following radiation, among others. Low-risk lesions include fibrous dysplasia, bone infarction, implants, giant cell tumor, chronic osteomyelitis, osteogenesis imperfecta, osteoblastoma, and chondroblastoma.

Suggested Reading

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