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Review Article

Types and Management of Bone Tumour

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Abstract

Introduction: A bone tumor is an abnormal growth of bone cells and often occurs in the area where bone grows rapidly. The main causative factor of bone tumor is unknown but familial heredity, genetic defects, radiation, and injuries are known to be the possible causative factor that initiates tumor formation in bone. With the advancement in data of genetic factors on bone, the tumor had led to show its role in diagnosis and classification. The classification of bone tumor vary according to the source, type of tissue involved, and histopathological features and is treated accordingly. Bone tumors are broadly divided as benign or malignant and primary or secondary.

Aim of the Study: The aim of the review is to understand the classification of the bone tumor with a brief insight into the treatment of the same.

Methodology: The review is the comprehensive research of PUBMED since the year 1995 to 2012.

Conclusion: The primary and secondary tumor of bone is widely categorized from osteogenic and fibrogenic to vascular, and lipogenic. The most important factor in dealing with the bone tumor is its correct diagnosis and treatment that follows which in turn depends on determining clinical staging, histopathological examination, radio-imaging, surgical excision, post-surgical treatment (chemotherapy, radiotherapy, hormonal and immunotherapy) and the prognosis depends on the metastases behavior. The longer survival may lead to a higher risk of metastases and pathological fractures. Thus, a multidisciplinary follow-up of the patient is essential for long term survival.

Keywords: *Primary Management of Osteoarthritis*

Introduction

For the prediction of the biological potential of the tumor, histopathological classification is widely accepted which represents a guide for treatment. The WHO classification does not include the concept of histogenesis and cell of origin of the tumor but rather focuses on a combination of parameters such as morphology, phenotype and genotype [1]. According to the recent WHO classification, the tumors are grouped in 15 different categories such as cartilage, osteogenic, fibrogenic, fibrohistiocytic, hemopoietic, notochordal, giant cell, vascular, smooth muscle and neural tumors, Ewing sarcoma, primitive neuroectodermal tumor etc [2].

WHO classification of bone tumour [3]

Behavior	Type of Tissue	Tumour
Benign	Bone Cartilage Misc	• Osteoma • Osteoid Osteoma • Osteoblastoma • Osteochondroma • Enchondroma • Chondroblastoma • Giant cell tumor • Solitary Aneurysmal bone cyst
Malignant	Bone Cartilage Fibrogenic tumor Fibrohistiolytic tumor Medullar tumor Misc	• Osteosarcoma with subtypes (conventional, chondroblastic, fibroblastic, osteoblastic, telangiectatic, small cell, low-grade central, secondary, para and periosteal, high-grade surface. Chondrosarcoma with subtypes (central, primary, secondary, peripheral, dedifferentiated, mesenchymal, clear cell)Fibrosarcoma • Malignant fibrous histiocytoma • Ewing sarcoma/ primitive neuroectodermal tumor • Giant cell tumour (malignant giant cell tumour, osteoclastoma) • Haematopoietic tumour (plasma cell myeloma, malignant lymphoma) • Vascular tumour (angiosarcoma) • Smooth muscle tumour (leiomyosarcoma) • Lipogenic tumour (liposarcoma) • Notochordal tumour (chordoma) • Adamantinoma

Osteoid osteoma

Osteoid osteoma is a benign bone-forming tumor with tumor cells directly forming mature bone. It accounts for 5% of all bone tumors. It predominantly occurs in the appendicular skeleton. The lower extremity is commonly affected, long bone such as femur and tibia are involved, juxta or intraarticular region of the femoral neck. Histologically the lesion is central vascular osteoid tissue and peripheral sclerotic bone. Clinically patient gives a history of pain. A radiolucent cortically-based nidus features less than 2 cm in size is seen in radiograph with sclerosis. Surgical treatment (en-bloc resection, CT guided percutaneous technique) is an option for a patient having severe pain and not responding to NSAIDs [4,5].

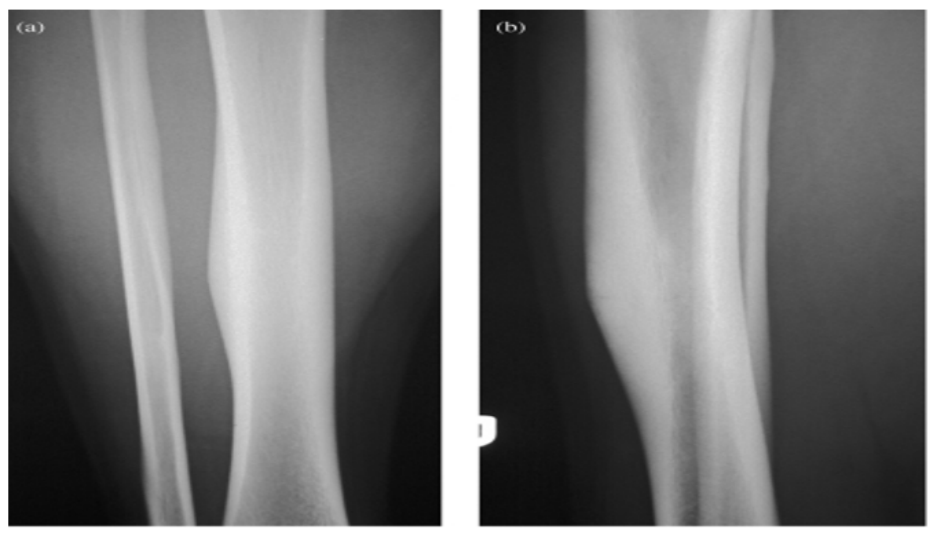

Figure 1: *Showing A. anteroposterior B. lateral aspect of tibia showing florid sclerosis and periosteal new bone formation [4].*

Osteoblastoma

Osteoblastoma is one rare primary bone neoplasm which closely resembles osteoid osteoblastoma. Bone forming lesion is found within medullary canal, periosteal tissue, cortex. It differs from osteoid osteoma in its ability to grow larger than 2 cm in size and aggressive behavior which often makes it difficult to differentiate it from osteosarcoma. It occurs in the first 3 decades of life and present with pain characterize as dull and achy which do not subsides with NSAIDS unlike osteoid osteoblastoma. The most common location is a vertebral column and long tubular bones. A well-circumscribed radiolucent lesion in the bony cortex with a thin shell of peripheral new bone separating soft tissue is seen in the radiograph. Surgical treatment is complete excision of the lesion [6].

Osteosarcoma

Osteosarcoma is a malignant mesenchymal bone tumor, primary in origin where tumor cells form from bone or osteoid or both. Osteosarcoma is the second most common bone tumor after multiple myeloma [4]. They are classified as follow.

Conventional: Most common among all other osteosarcomas, occur in the second or third decade of life, situated in the metaphyseal region of a long bone. Radiographically, the lesion appears sclerotic, lytic, or have mixed pattern, 'sun-burst appearance or Codman's triangle' is a characteristic feature appear as a periosteal reaction. Soft tissue infiltration may be seen [4].

Telangiectatic osteosarcoma: Accounts for 5% of all osteosarcoma and is more aggressive than the conventional one. They occur in metaphysis and diaphysis of bone with lesion contain blood clot filled large cystic cavity which differentiates it from conventional osteosarcoma. Cortical destruction, periosteal reaction and Codman's triangle may be seen in the radiograph. MRI may show fluid-filled cysts [4].

Parosteal osteosarcoma: Occurs in the second to fifth decade of life and accounts for 5% of all osteosarcoma. A common site is metaphysis of long bones; femur is most common among them. The clinical presentation is mild pain with a limited range of motion due to joint interference. On a radiograph, the lesion is dense, ovoid and rounded mass connected to the cortex of underlying bone by a stalk initially. A thin line separating tumor from underlying bone is the classic radiographic feature of the tumor [4].

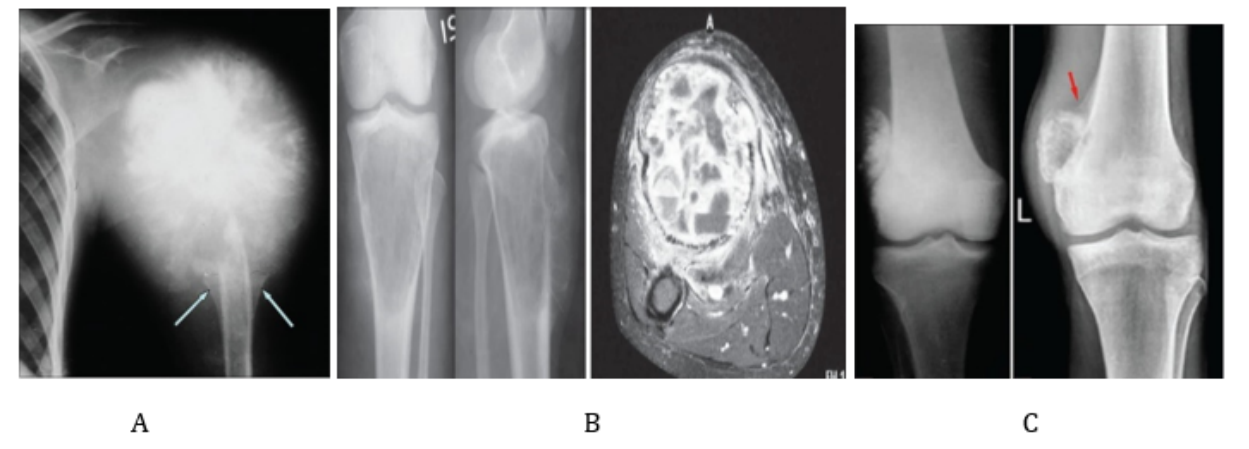

Figure 2: *Showing A. typical sunburst appearance of conventional osteosarcoma B. telangiectatic of proximal tibia showing lysis and expansion and MRI is showing a cystic fluid-filled cavity C. Paraosteal osteosarcoma underlying the cortex and periosteal osteosarcoma with the lifting of the periosteum (red arrow) [9].*

Periosteal osteosarcoma: It is another surface variant of osteosarcoma causing erosion of cortex, evident on the radiograph [7].

Metastatic long bone often represents impending or complete bone fracture, thus open internal fixation of long bones is usually preferred treatment which is further stabilize using locked intramedullary devices followed by radiation therapy. Replacement arthroplasty is done for pathologic fractures of the femoral and humeral head of neck, distal femoral or proximal tibial condylar defects. The radiotherapy after surgery can be critical and may aid in the healing of the pathological fracture [8].

Giant cell tumor

It is locally aggressive tissue mainly composed of connective tissue, stromal cells and giant cells. The most common clinical presentation is pain and swelling. On a radiograph, a giant cell tumor is seen involving metaphysis and epiphysis that extends to the subarticular border, bony expansion, cortical thinning and erosion. The surgical intervention is by intralesional curettage and autograft reconstruction by placing the iliac graft in cavity post curettage [10].

Osteochondroma

Osteochondroma/exostosis is bony protuberance covered with cartilage, arising from the surface of the bone(metaphyseal regions). The lesion is non-tender and slow-growing and is confined to the childhood and adolescent period but may be discovered after 20 years. Osteochondroma can occur in bone arise from endochondral ossification. On a radiograph, osteochondromas appear as bony protuberance arising from the external surface of long bones. The asymptomatic lesion does not require any treatment and should be monitored periodically for further changes. The symptomatic lesion needs to be excised leaving no cartilaginous margin since it may lead to recurrence [11].

Enchondroma

Enchondroma is benign in nature, cartilage forming tumor, arising within the medullary cavity of the bone. Lesions are usually asymptomatic. On a radiograph, they appear as a medullary lesion with geographic margin and endosteal scalloping and calcification [4].

Chondrosarcoma

Chondrosarcoma is another cartilage producing but malignant tumor of bone. Primary chondrosarcoma arises de novo and most of them are secondary in nature, arising from benign lesions such as osteochondromas or enchondromas. It usually occurs in the fifth or sixth decade of life with a male predominance. The lesion can be central or peripheral. The lesion arises in the metaphysis and may extend into the epiphysis. The high-grade tumor is often fast-growing with painful swelling, while the low-grade tumors are mild in pain with swelling. Pathologically it is seen as large grayish-white lobulated masses with ill-defined border, firm in consistency and area of focal calcification, mucoid degeneration or necrosis is present. The calcification is the important characteristic feature of chondrosarcoma on the radiograph. Periosteal reaction, scalloping of the inner cortex, cortical thickening and soft tissue invasion is seen. Wide surgical excision is the preferred treatment for chondrosarcoma [12].

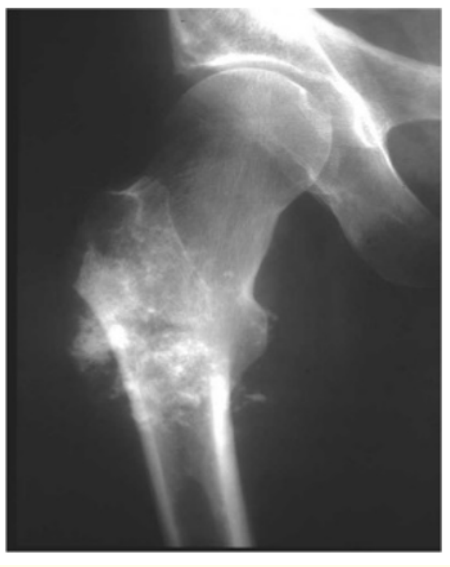

Figure 3: *Showing secondary chondrosarcoma developing in diaphyseal aclasis [4].*

Multiple myeloma

Multiple myelomas are the most common primary malignant tumor of bone. Plasma cells with the monoclonal proliferation of B-cells results in marrow infiltration of the skeleton and osteolytic damage of bone. The tumor may affect any bone with a hematopoietic red marrow. The common presentation of the disease is in the fifth decade of life with men are affected twice as women. Pain is a common symptom along with the symptoms of anemia. Radiographic features may vary, it may appear normal or show diffuse demineralization or s single osteolytic lesion or widespread lytic lesions. These osteolytic lesions are well-defined punched out lesions of varying sizes [4].

Fibrosarcoma

Fibrosarcoma is a malignant tumor showing the presence of fibrous tissue not associated with osteoid production, bone or cartilage. The most common site is metaphysis or metadiaphysis of long bones and most prevalent in the fourth to sixth decade of life. The lesion is grayish-white in color with rubbery inconsistency. On a radiograph, the lesion appears as a moth-eaten osteolytic lesion with ill-defined margins. Periosteal reaction is not seen but cortical destruction and soft tissue extension are evident. Surgical intervention includes en bloc resection or wide resection depending on the reach of the tumour to muscle origin. Chemotherapy and radiotherapy are other adjuncts [13].

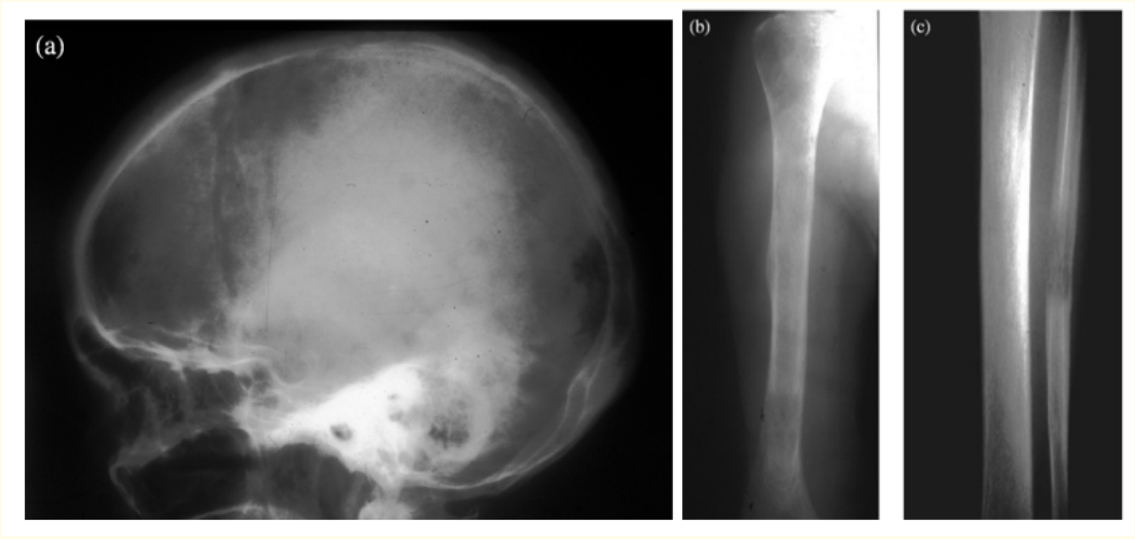

Figure 4: *Showing multiple myeloma of A. skull B. humerus C. fibula with multiple small punched-out osteolytic lesions [4].*

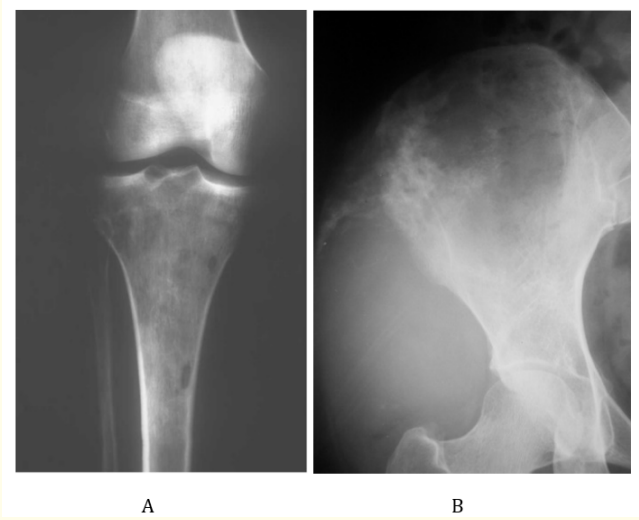

Figure 5: *Showing A. Fibrosarcoma B. Malignant fibrous histiocytoma [4].*

Malignant fibrous histiocytoma

The primary osseous malignant lesion is seen in the fifth to seventh decade of life with a slight male predilection. The most common location is metaphysis and diaphysis of long bones. The patient may present with acute or chronic pain, tenderness and swelling over the lesion. Tumour is usually located within the medullary cavity and radiographically appears as an aggressive lesion associated with soft

tissue with little presence of periosteal reaction and calcification at the periphery of the mass. En-bloc resection is the principal treatment. The role of chemotherapy and radiotherapy is limited [14].

Lymphoma

The intraosseous lymphoma accounts for 5% of malignant bone tumors, seen in fourth to the fifth decade of life with men being more commonly affected. Clinically patient presents with pain or swelling over the site of involvement. On a radiograph, the lymphoma appears as a vague, mottled lesion with cortical destruction but no periosteal reaction. Treatment of lymphoma may include a combination of multi-agent chemotherapy (CHOP- cyclophosphamide, doxorubicin, vincristine and prednisolone). Surgical removal of the tumor is not a common practice; however, pathological fractures need to be corrected surgically [15].

Conclusion

The bone tumor can be broadly divided into primary or secondary on the basis of origin or benign or malignant on the basis of behaviors of a tumour. The appropriate diagnosis using conventional radiography, MRI and CT as well as the clinical staging of the tumor, is essential for treatment planning. Most of the benign and malignant tumor requires surgical excision, chemotherapy, and radiotherapy depending on the condition. Infiltrative tumor requires wide resection. A primary malignant tumor is rare; therefore it is difficult to appropriately recognize and classify them. On the other hand, bone sarcomas are commonly affecting children and adolescents hence have a major impact on the life of patients and their families. With the advancement in surgical and other treatment modalities, it is possible to improve the outcome and survival rate of these bone tumors.

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