BONE TUMOURS , CLASSIFICATION & ASSESSEMENT

Dr. Mayilvhanan Natarajan*

Tumours arising from the skeletal system are uncommon neoplasms accounting for a very small fraction of the malignant and benign lesions of the body. Despite this rarity they hold an unusual fascination for clinicians and scientists, based in part on the broad panoply of possible diagnoses, the prevalence of the tumour in younger people, and the dramatic virulence of the malignant lesions.

Since each component of the osseous system may give rise to several forms of benign and malignant tumours, the variety of radiographic and histologic presentations presents an enormous challenge to the orthopaedist, radiologist, and pathologist who deal with the tumours. Over the past several decades a series of strategies has evolved for the surgical, oncological, and radiotherapeutic management of the lesions, which enormously add to the complexity and excitement of dealing with tumours.

Benign bone tumours are more frequent in incidence, perhaps outnumbering their malignant counterparts three to fourfold. It is obvious with such small numbers that the smaller medical centers have difficulty gaining experience in dealing with these difficult problems.

* M.B.B.S. D.ORTHO.M.Ch. Trauma(Eng.) Ph.D. As. Prof. of Ortho.Surgery Govt.Gen.Hospital & Madras Medical College, M.G.R. Medical University, Madras- 600 003.

The distribution of tumours varies considerably with age. Most benign bone lesions, osteosarcomas, and Ewing's sarcomas occur in the second and third decade, while giant cell tumours, chondrosarcomas, fibrosarcomas, myelomas, lymphomas, and metastatic disease all predilect the older age groups. With the exception of GCT, most benign and malignant tumours of bone are slightly more common in the male.

The etiology of bone neoplasms remains largely unknown. A genetic basis for some tumours is suggested by the increased incidence of bone sarcomas in patients with hereditary multiple osteocartilaginous exostoses, osteogenesis imperfecta, and in survivor's of bilateral retinoblastoma. Osteosarcomas have also been reported in siblings and in greater frequency in first cousins of patients with osteosarcoma. Other diseases such as solitary osteochondroma, Ollier's disease [multiple enchondromatosis], Mafucci's syndrome, bone infects, chronic osteomyelitis, Paget's disease, and fibrous dysplasia are associated with a higher incidence of sarcoma than in the general population. The true incidence of sarcomas in Paget's disease is considerably less than 1% of patients affected with the disorder, but it contributes to a second peak occurrence of osteosarcomas noted in patients of the fourth, fifth and sixth decades. An increased incidence of bone sarcomas is seen in bone irradiated for bone tumours or in bone that fell within the

radiation fields employed in treatment of other diseases.

Strict criteria for identifying such sarcomas have been established and include a relatively long symptom-free latent period between the radiation exposure and the occurrence of the sarcoma. In addition to sarcomas, benign osteochondromas also occur following radiation of immature bone. Trauma has been implicated in the etiology of bony neoplasms but, in facts most lesions are "discovered" after a trauma. There is no evidence that a single, uncomplicated injury causes cancer.

A viral etiology for bone sarcomas has been postulated and several animal models of viral induced disease have been investigated. Support for such a finding in human osteosarcomas has been provided by Morton and Malgren, who demonstrated cytotoxic anti-osteosarcoma antibodies in associates and family members of patients with osteosarcoma, and studies by Finkel et al, have shown that osteosarcomas can be induced in hamsters by injection of extracts of human osteosarcoma tissue. In spite of these investigative efforts osteosarcoma tissue. In spite of these investigative efforts no viruses have been recovered from any human osteosarcomas.

The types of tumours occurring within bone reflect its various components including the osseous tissues, the bone marrow and supporting connective tissues, along with its component nerves, blood vessels, and fat.

A generally accepted classification system is based on the predominant matrix component and type of cell differentiation within the lesion. Thus each type of tissue contained within the bone may give rise to one, or in most cases several, clinically, radiologically and histologically distinct benign and malignant neoplastic lesions, each with its own pattern of biologic behaviors. In many instances the lesion arises directly from the same type of tissue (e.g. osteosarcoma arises from osseous tissue), but this is not always the case.

Chondrosarcoma, for example, frequently occurs in locations which normally contain no cartilage. The classification system neatly divides all lesions into "benign" and "malignant" categories for each tissue, but there is little evidence to suggest that the malignant lesions occur as a dedifferentiation of their benign counterparts. Furthermore the distinction between benign and malignant tumours is not always clear with certain tumours such as the osteoblastoma, giant cell tumour, or chondroblastoma occasionally behaving in a very aggressive and rarely frankly malignant manner.

An even more difficult problem is that some of the malignant tumours have variable degrees of histological aggressiveness so that a chondrosarcoma may be classified as low, intermediate, or high grade and show a conforming pattern of biological behavior. The system as defined however, provides a useful framework in which to organize these heterogeneous lesions. For the most part it is possible to categorize all but about 1% of tumours using the system.

PRINCIPLES OF DIAGNOSIS AND STAGING

Considering the problems presented by these highly variable groups of neoplasms, it should be obvious that the evaluation of the patient with a bone tumour is a complex procedure, often requiring consultation and discussion amongst orthopaedists, radiologists, pathologists, radiotherapists, and medical oncologists to arrive at the correct diagnosis and institute the appropriate treatment. Much of this depends on the pre-biopsy workup or "first-order screen" which generally includes a detailed history, physical examination, laboratory tests, plain roentgenograms or xeroradiograms of the lesion, radionucleide bone scans, and a chest X-ray.

The goal of this first set of procedures is to arrive at a tentative decision whether the lesion is a benign or malignant primary bone tumour, a metastatic deposit, or a marrow cell lesion.

Except rarely, the history presented with most benign and malignant neoplasms is non-specific. Most often the patient complains of pain, which is first noted after minor trauma, but many benign, and occasionally malignant lesions are discovered incidentally on X-ray obtained for other purpose. On occasion a lump or pathological fracture may be the presenting problem. The pain is usually dull and aching unless a pathological fracture has occurred. A notable exception is the osteoid osteoma, in which the pain is described as sharp and boring often worse at night, and characteristically completely relieved by aspirin.

On examination, benign lesions may have minimal findings. An osteoid osteoma may show local tenderness and joint limitation or, in children, may present with a scoliosis, limp, or growth disturbance. Malignant lesions are often associated with a palpable soft tissue mass which is tender to palpation. The mass should be assessed accurately for its size, the distinctness of its margins, its consistency, mobility, and location. Large lesions are more likely to be malignant, as are tumours that are tender to palpation.

In some malignant, and occasional benign tumours, heat, redness, edema, venous distention, and even lymphangitis may be present, at times making the differentiation from infection difficult. Systemic findings are usually lacking except patients with Ewing's sarcoma or lymphoma, who may present with fever, chills, anorexia, and weight loss consistent with a chronic or subacute infectious process.

Laboratory tests include a complete blood count and sedimentation rate, which are helpful in excluding diseases such as myeloma, leukaemia, and infection. Calcium and phosphorus determinations are useful in determining the presence of metabolic bone disease as well as the hypercalcemia which occurs in some patients with metastatic bone disease, but it may be significantly elevated in patients with osteosarcoma, lymphoma, or Ewing's sarcoma. A serum immuno-electrophoresis helps to determine cases of multiple myeloma. Plain roentgenorgrams of the lesion in two or more planes are very helpful in the screening process. The lesion can be assessed for location, size, cortical integrity, margination, and the presence or absence of a soft tissue mass. It is often

possible with a simple roentgenogram or xeroradiograph to make a preliminary determination whether the lesion contains bone, chondroid, fibrous or "other" tissues, and some non-neoplasmic disorders such as bone infarct, infection or stress fracture can be excluded. The xeroradiogram is particularly helpful in delineating a soft tissue extension of the lesion. The radionucleide bone scan [usually Tc 99M di- or poly- phosphonate] is important not only in assessing the activity of the lesion relative to its bone production and blood flow, but also in determining the presence or absence of bony lesions at other sites. One should note that on occasion lesions such as eosinophilic granuloma, simple bone cyst, and multiple myeloma may appear normal or actually decreased in activity on bone scan. A chest X-ray is obtained on any patient with a suspected malignant bone tumour to search for evidence of metastatic disease, or a primary focus from which a metastasis may have arisen.

After these preliminary studies a decision is made. If the lesion is believed to be a primary benign bone tumour one may elect to observe it or biopsy it for confirmation. If the lesion is thought to be a primary malignant bone tumour further second order studies are required. These include a computed tomogram [CT] of the lesion, an arteriogram [in cases presumed to be resectable or to assess the extent of the soft tissue extension], and tomograms of the lesion. These tests are principally directed towards determining the extent of the lesion within the bone and adjacent soft tissues.

The relationship of the soft tissue mass to the surrounding anatomical structures, especially the neurovascular structures, is critical. It is important to obtain this iinformation before performing the biopsy, because these tests may be affectedly the hematoma associated with the biopsy.

Tomograms or computed tomograms of the chest are also obtained preoperatively on patients suspected of having malignant bone tumours.In lesions thought to be metastatic, preliminary workup seeking the site of the primary tumour is helpful. In addition to the history and physical examination the studies may include mammograms, urinalysis, intravenouspyelography, acid phosphatase determination, a thyroid scan, and chest tomograms or CT. On occasions it is not possible to find the primary tumour, and the diagnosis will depend upon the histologic diagnosis of the bony lesions determined by biopsy.

For tumours thought to be round cell lesions [Ewing's tumor, myeloma, and lymphoma], further staging studies should include a gallium scan, abdominal CT, and lymphangiography. Myeloma patients require a serum immunoelectrophoresis, a skeletal bone survey, and a bone marrow biopsy before biopsy of the bony lesion.