Lec 1

Bone and cartilage Tumors

The rarity of primary bone tumors and the disfiguring surgery often required to
treat bone malignancies make this group of disorders especially challenging.
Therapy aims to optimize survival while maintaining the function of affected
body parts. The predilection of specific types of tumors for certain age groups
and particular anatomic sites provides diagnostic clues. For example,
osteosarcoma peaks during adolescence and most frequently involves the knee,
whereas chondrosarcoma affects older adults and arises most often in the pelvis
and proximal extremities.

Bone tumors may present in a variety of ways:

benign lesions are often asymptomatic incidental findings while other tumors cause pain or are identified as a slow-growing mass or pathologic fracture. Radiographic imaging defines the location and extent of the tumor and can detect features that narrow the differential diagnosis, but biopsy is necessary for definitive diagnosis in almost all cases. Bone tumors are classified according to the normal cell types they recapitulate or the matrix they produce (Table 19.1). Lesions that do not have normal tissue counterparts are grouped according to their clinicopathologic features .

Bone-Forming Tumors

Tumors in this category produce unmineralized osteoid or mineralized woven bone.

Osteoma

An osteoma is a rare benign, slow-growing lesion, regarded by some as a hamartoma rather than a true neoplasm. Osteoma is almost exclusively restricted to flat bones of the skull and face. It may grow into paranasal sinuses or protrude into the orbit.

Morphology : The lesion is composed of well-differentiated mature lamellar bony trabeculae separated by fibrovascular tissue.

Table 19.1 Classification of Selected Primary Bone Tumors

Category	Behavior	Tumor Type	Common Locations	Age (yr)	Morphology
Cartilage forming	Benign	Osteochondroma	Metaphysis of long bones	10–30	Bony excrescence with cartilage cap
		Chondroma	Small bones of hands and feet	30–50	Circumscribed intramedullary hyaline cartilage nodule
	Malignant	Chondrosarcoma (conventional)	Pelvis, shoulder	40–60	Extends from medullary canal through cortex into soft tissue, chondrocytes with increased cellularity and atypia
Bone forming	Benign	Osteoid osteoma	Metaphysis of long bones	10–20	Cortical, interlacing microtrabeculae of woven bone
		Osteoblastoma	Vertebral column	10–20	Posterior elements of vertebra, histology similar to osteoid osteoma
	Malignant	Osteosarcoma	Metaphysis of distal femur, proximal tibia	10–20	Extends from medullary canal to lift periosteum, malignant cells producing woven bone
Unknown origin	Benign	Giant cell tumor	Epiphysis of long bones	20–40	Destroys medullary canal and cortex, sheets of osteoclasts
		Aneurysmal bone cyst	Proximal tibia, distal femur, vertebra	10–20	Hemorrhagic spaces separated by cellular, fibrous septa
	Malignant	Ewing sarcoma	Diaphysis of long bones	10–20	Sheets of primitive small round cells

Adapted from Unni KK, Inwards CY: *Dahlin's Bone Tumors*, ed 6, Philadelphia, 2010, Lippincott-Williams & Wilkins; by permission of Mayo Foundation.

Osteoid Osteoma and Osteoblastoma

Osteoid osteoma and osteoblastoma are benign bone-producing tumors that have similar histologic features but differ clinically and radiographically.

By definition, osteoid osteomas are less than 2 cm in diameter. They are most common in young men. About 50% of cases involve the cortex of the femur or tibia. A thick rim of reactive cortical bone may be the only radiographic clue.

Despite their small size, they present with severe nocturnal pain that is probably caused by prostaglandin E2 produced by osteoblasts and is relieved by aspirin and other nonsteroidal antiinflammatory drugs.

Osteoblastomas are larger than 2 cm and more frequently involve the posterior components of the vertebrae (laminae and pedicles). Any associated pain is unresponsive to aspirin and the tumor usually does not induce a marked bony reaction..

Osteoid osteoma can be treated by radiofrequency ablation, whereas osteoblastoma is usually curetted or removed by en bloc excision

MORPHOLOGY

Osteoid osteoma and osteoblastoma are round-to-oval masses of hemorrhagic gritty tan tissue. They are well circumscribed and composed of delicate interconnecting trabeculae of woven bone that are rimmed by a single layer of

osteoblasts . The stroma surrounding the neoplastic bone consists of loose connective tissue with abundant dilated and congested capillaries. The relatively small size, well-defined margins, and benign cytologic features of the

neoplastic osteoblasts help distinguish these tumors from osteosarcoma

Cartilage-Forming Tumors

These tumors are characterized by the formation of hyaline cartilage. Benign cartilaginous tumors are much more common than malignant ones

Osteochondroma

Osteochondroma, or exostosis, is a benign, cartilage-capped tumor that arises from the bone surface. It may be sessile or pedunculated with a bony stalk.

About 85% are solitary. The remainder are seen as part of the multiple hereditary exostosis syndrome .

Solitary osteochondromas are usually first diagnosed in late adolescence and early adulthood, whereas multiple osteochondromas present during childhood. Men are affected three times more often than women.

Osteochondromas develop in bones of endochondral origin and arise from the metaphysis near the growth plate of long tubular bones, especially near the knee . They are slow-growing masses that can be painful if they impinge on a nerve or if the stalk is fractured. In many cases they are detected incidentally. In multiple hereditary exostoses, the underlying bones may be bowed and shortened, reflecting an associated disturbance in epiphyseal growth.

MORPHOLOGY

Osteochondromas range in size from 1 to 20 cm. The cap is composed of hyaline cartilage covered by perichondrium. The cartilage recapitulates the growth plate and undergoes endochondral ossification, with the newly made bone forming the inner portion of the head and stalk. The cortex of the stalk merges with the cortex of the host bone resulting in continuity between the medullary cavity of the osteochondroma and the host bone.

Clinical Features

Osteochondromas usually stop growing at the time of growth plate closure and,when symptomatic, are cured by simple excision. Secondary chondrosarcoma

develops only rarely, usually in tumors associated with multiple hereditary exostosis.

Chondroma

Chondromas are benign tumors of hyaline cartilage that occur in bones of endochondral origin. Tumors can arise within the medullary cavity (enchondromas) or, rarely, on the bone surface (juxtacortical chondromas).

Enchondromas are usually diagnosed in individuals 20 to 50 years of age. Typically, they appear as solitary metaphyseal lesions of the tubular bones of the hands and feet.

Radiographs show a circumscribed lucency with central irregular calcifications, a sclerotic rim, and an intact cortex .

Ollier disease and Maffucci syndrome are disorders characterized by the development of multiple enchondromas (enchondromatosis).

Most sporadic enchondromas of large bones are asymptomatic and are detected incidentally, but they occasionally cause painful pathologic fractures.

MORPHOLOGY

Chondromas are usually smaller than 3 cm, gray-blue, and translucent. They are composed of benign-appearing chondrocytes embedded in well circumscribed

nodules of hyaline cartilage .

Clinical Features

The growth potential of enchondromas is limited. Treatment depends on the clinical situation and usually includes observation or curettage. Solitary enchondromas rarely undergo sarcomatous transformation; by contrast, those associated with enchondromatosis do so more frequently.

Chondroblastoma

Chondroblastoma is a relatively rare benign tumour arising from the epiphysis of long bones adjacent to the epiphyseal cartilage plate. Most commonly affected bones are upper tibia and lower femur (i.e. about knee) and upper humerus. The tumour usually occurs in patients under 20 years of age with male preponderance (male-female ratio 2:1).

The radiographic appearance is of a sharply-circumscribed, lytic lesion with multiple small foci of calcification.

G/A is a well-defined mass, up to 5 cm in diameter, lying in the epiphysis. Cut surface reveals a soft chondroid tumour with foci of haemorrhages, necrosis and calcification.

M/E The tumour is highly cellular and is composed of small, round to polygonal mononuclear cells resembling chondroblasts and has multinucleate osteoclastlike giant cells.

The End