Malignant bone tumors often present with nonspecific symptoms and are frequently discovered on standard radiographs incidentally. The recognition of the standard radiographic signs of an aggressive bone lesion is important, because early diagnosis and prompt treatment will improve the outcome dramatically.

Once a malignant bone tumor is detected, further diagnostic work-up is required in order to characterize the lesion and to determine the extent of the disease.

There is a whole range of malignant bone tumors, including osteosarcoma, Ewing’s sarcoma, lymphoma, chondrosarcoma, fibrosarcoma, malignant fibrous histiocytoma, plasmocytoma/multiple myeloma or metastasis.

In the first part of this review, the merits of each imaging modality in this diagnostic work-up will be highlighted.

The second part will focus more specifically on the imaging features of malignant bone tumors, with special emphasis on standard radiographic findings.

Discussion

Imaging strategy

Lesion detection and characterization

Clinical symptoms are nonspecific and include local swelling and tenderness, not infrequently after a minor trauma or pathological fracture.

The next step in the diagnostic setting starts with conventional radiographs, which is regarded as the gold standard for lesion detection. However, in more complex anatomical areas, such as the spine or skull base, CT or MRI may be complementary.

Besides lesion detection, standard radiography plays a pivotal role in lesion characterization as well. Other important factors that may contribute to the characterization are the patient’s age, lesion location and multiplicity.

Indeed, some tumors, such as osteosarcoma and Ewing’s sarcoma tend to occur exclusively in children and adolescents, whereas metastases, chondrosarcoma and multiple myeloma are mostly found in the elderly population.

CT may be used for depicting small matrix calcifications and appreciating cortical destruction.

MRI plays only a minor role in characterization.

MRI features mostly are nonspecific, showing a high signal intensity (SI) on T2-weighted images (WI) and a low SI on T1-WI. Nevertheless, some tumors have specific findings on MRI or CT, based on their histological composition and/or location (1).

Staging and follow-up after treatment

MRI is the imaging technique of choice for local tumor staging. Local staging parameters include the presence of skip metastases, joint and soft-tissue invasion and involvement of the neurovascular bundle.

Dynamic MRI, as part of a routine MRI protocol, is used for short term follow-up after neo-adjuvant chemotherapy, since tumor vascularization is a better parameter in evaluating tumor response than tumor volume (1-3).

A precise local staging and short term follow-up after neo-adjuvant chemotherapy have led to an increase in limb-salving surgery (1, 2).

CT remains the gold standard for distant staging (lung, liver and lymph nodes) and in guiding biopsy. Angiography has been largely surpassed by other imaging techniques as adjuvant imaging technique for diagnostic purposes. The only residual use is local assessment of chemotherapy in isolated limb perfusion (ILP) (2).

Whole body bone scintigraphy will document bone metastases or lesion multiplicity.

Table I. – Most frequent malignant bone tumors.

| Osteosarcoma          |
| Ewing’s sarcoma       |
| Chondrosarcoma        |
| Lymphoma              |
| Plasmocytoma/Multiple myeloma |
| Chordoma              |
| Metastasis            |
| Fibrosarcoma          |
| Malignant fibrous histiocytoma |
| Hemangio-endothelioma and hemangio-endotheliosarcoma |
| Hemangiopericytoma    |
| Adamantinoma          |
| Ameloblastoma and Adenoameloblastoma |

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Imaging features of malignant bone tumors

Only the most frequent histological subtypes will be discussed. Other more rare malignant bone tumors are summarized in Table I and are extensively described in various textbooks.

Osteosarcoma

Osteosarcomas can be divided into different subtypes. Depending on their specific location or histological composition, the standard radiographic appearance can be very variable.

Grossly osteosarcomas are divided into three major groups: intramedullary, juxtacortical or extraskeletal. There are further subdivided according to their specific location or histology (Table II) (3).

Discussion of the different subtypes is beyond the scope of this brief review. Only the conventional osteosarcoma, which accounts for 75% of all osteosarcomas, will be discussed here. For a more in depth review of other subtypes, we refer to reference 3.

Conventional osteosarcoma

This tumor is preferentially located in the metaphysis of long bones around the shoulder and knee. They are almost exclusively found in children and adolescents.

Although they can present as pure lytic or pure osteoblastic lesions, they are most frequently mixed osteosclerotic and osteolytic on standard radiographs.

A Codman triangle, sunburst or hair-on-end periosteal reaction is seen, indicating the aggressive nature of the lesion (Fig. 1). A soft tissue component is almost always present (3).

Ewing’s sarcoma

Ewing’s sarcoma is the second most frequent primary bone tumor after osteosarcoma.

It occurs almost exclusively in young children and adolescents.

Unlike osteosarcoma this tumors may exhibit systemic symptoms, mimicking osteomyelitis (4).

In contrast with osteosarcoma, Ewing’s sarcoma has a variable location. Mostly the long bones, but also the flat bones, such as the ribs or more frequently the pelvic bones, may be involved.

Location in the midshaft rather than at the end of the long bones is more frequent (4).

The overall prognosis is poor.

The radiographic hallmark is an osteolytic lesion with an onion-skin periosteal reaction, although the radiographic appearance may be extremely variable and in 25 percent of cases the lesion can be sclerotic. A soft-tissue component is common (4).

Chondrosarcoma

A chondrosarcoma is a malignant mesenchymal tumor, embedded in a chondroid matrix, produced by the tumor cells. They tend to occur more frequently in the older population.

Chondrosarcomas can be subdivided based on their origin (primary of secondary), location within the bone and cellular pattern, reflecting their biological behavior (Table III).

Chondrosarcomas developing from a preexisting lesion, mostly an...
enchondroma or osteochondroma, are called secondary chondrosarcomas.

Differentiating an enchondroma or osteochondroma from their malignant counterparts, can be difficult (5).

Pain and change in the calcification pattern of an enchondroma or osteochondroma are highly suggestive for malignant degeneration. Due to their chondroid matrix, chondrosarcomas have some typical imaging features.

"Popcorn-like" calcifications best seen on standard radiographs (Fig. 2A) or CT and 'ring and arc' enhancement on MRI with gadolinium (Fig. 2B), suggest a chondroid tumor (5).

Furthermore, cortical scalloping, cortical disruption, an aggressive periosteal reaction and an inhomogeneous distribution of calcifications with large lucent areas, are suggestive for malignancy (5).

Central chondrosarcoma

Central chondrosarcomas are located in the medullary cavity, mostly in the metaphysis or diaphysis.

The femur is the most frequently affected bone, although chondrosarcomas can be found anywhere in the skeleton.

The appearance of a chondrosarcoma on standard radiographs largely depends on the degree of malignancy, but in most cases, a geographic, moth-eaten or permeative osteolytic lesion is seen.

Grade I and II sarcomas cause scalloping of the cortex with a fusiform expansion, whereas grade III lesions penetrate and destroy the cortex.

An aggressive periosteal reaction, like a Codman's triangle, onion-skin or hair-on-end periosteal reaction, is suggestive for grade III lesions (5).

Peripheral chondrosarcoma

Peripheral chondrosarcomas are the malignant counterparts of osteochondromas.

They are mostly located in the scapula, iliac bone, tibia, femur, pubic bone or rib.

On standard radiography, continued growth of an exostosis after skeletal maturity, changes in surface delineation with previous radiographs, internal lytic areas, erosion or destruction of the adjacent bone, the presence of a soft tissue mass, or an irregular distribution of calcifications, should raise the suspicion of a malignant degeneration. These chondrosarcomas originate in the cartilaginous cap, which is not visible on standard radiographs.

For superficially located lesions, the thickness of this cartilage cap can be easily measured by ultrasound.

For deeper seated lesions—however—MRI is the modality of choice to evaluate the cartilage thickness. On T2-WI, the hyaline cartilage cap is of high signal intensity, in contrast to the underlying cortical bone. The normal thickness should not exceed 3 cm before skeletal maturity, whereas in adults a cartilage more than 1.5 cm thick should raise the possibility of malignant transformation (6).

Therefore, MRI is imaging modality of choice to detect malignant degeneration of an osteochondroma (5).

Primary bone lymphoma

Primary bone lymphoma (PBL) is an uncommon malignancy that accounts for less than 5% of all primary bone tumors. It should be distinguished from a secondary lymphomatous invasion of the bone. In PBL, there is a lack of lymphadenopathies.

Symptoms include insidious and intermittent bone pain, systemic
Symptoms and pathological fracture. Spinal involvement can cause radicular symptoms and even spinal cord compression.

The metadiaphysis of the femur and the spine are the most affected sites.

Primary bone lymphomas (PBL) may have variable imaging features but, most often a solitary, permeative osteolytic lesion with a multilamellar periosteal reaction is seen. In a minority of cases, PBL presents as a mixed sclerotic-lytic lesion.

Sometimes standard radiographs can have a ‘near-normal’ appearance, requiring a bone scintigraphy or MRI to demonstrate tumoral involvement.

Most PBL have a soft tissue extension with surprisingly little cortical destruction.

Plasmocytoma/multiple myeloma

Multiple myeloma is the most frequent primary malignant bone tumor.

Fig. 3. – Multiple myeloma. Standard radiograph (lateral (A) and frontal (B) view) of the skull: multiple punched out lesions are seen in the skull vault. Standard radiograph (frontal view) of the femur (C) shows punched out lesion in the distal third of the femur. Standard radiograph (frontal view) of the pelvis (D): osteolytic destruction of the left iliopectineal ramus.
It arises from malignant transformed plasmacells with an overabundance of paraprotein.

A solitary lesion is designated as plasmacytoma, whereas multiple lesions are characteristic for multiple myeloma.

Clinical symptoms include infections, bone pain, renal insufficiency, pathological vertebral fractures with possible spinal cord compression.

In contrast to osteosarcoma and Ewing’s sarcoma, these tumors occur in the elderly population.

Multiple myeloma and plasmacytoma present on standard radiographs as well circumscribed osteolytic lesions, better known as punched-out lesions. They are most frequently located in the skull vault (fig. 3A,B), spine, ribs, femora (fig. 3C) and pelvis (fig. 3D).

In the spine, lesions may be shallow and may even not be visible on standard radiographs. In these cases, CT and MRI are the methods of choice to detect vertebral lesions (8).

The only visible radiographic sign may be diffuse demineralization of the skeleton.

It is important to remember that scintigraphy is negative in case of diffuse multiple myeloma and can not be advocated as a useful imaging technique.

Therefore CT and MRI are the methods of choice to detect spinal lesions (8).

Chordoma

Chordomas are rare tumors. Accounting for 2-4% of all primary bone tumors, they originate from embryonic notochordal remnant along the length of the neuraxis.

In rare cases they can also occur in extra-axial locations.

The most frequently involved area is the sacral bone, followed by the clivus. The rest of the spine is less frequently involved.

Chordomas can be found at every age. Although they almost never metastasize, they are locally aggressive.

Symptoms depend on the location. Rectal dysfunction and urinary incontinence are associated with sacral chordomas, whereas headache and cranial nerve palsy are seen in clival chordomas.

On standard radiographs, bone destruction can be seen, but is better appreciated on CT.

On MRI chordomas, typically show a very high SI on T2-WI because of their high fluid contents of vacuolated cellular components (9).

Chordomas of the spine extend over more levels with typical sparing of the intervertebral discs (10).

Metastasis

Metastases are mostly found in patients older than 40 years.

Standard radiographic features range from lytic to sclerotic, depending on the primary tumor (fig. 4).

Metastases may arise anywhere in skeleton, but mostly those areas containing red bone marrow are affected.

The best screening method for metastases is bone scintigraphy. Hot areas are initially further examined by standard radiographs. However, if less than 50 percent of bone is destroyed or the metastases are located in anatomical complex regions, standard radiographs may be normal. In these cases, adjuvant CT or MRI is mandatory.

A more recent technique to screen for bone metastases, is whole body MRI. The drawback of this method is its high costs and relatively low availability compared to scintigraphy.

Conclusion

Malignant bone tumors are frequently discovered by coincidence on standard radiographs after minor trauma. Despite the availability of multiplanar imaging, standard radiographs are still the mainstay in detection and characterization of malignant bone tumors.

Histology is still the gold standard for definitive characterization of a malignant bone tumor. However, the combination of age, location, multiplicity and features on standard radiographs can narrow the differential diagnosis.

Except for anatomical difficult regions, cross-sectional imaging has only a minor role in the detection and characterization of malignant bone lesions.

CT is used to screen for distant metastases, whereas bone scintigraphy is the method of choice to demonstrate bone metastases.

MRI has a major role for local tumor staging and long term follow-up for recurrence. Dynamic MRI should be a part of every routine MR protocol, to evaluate tumor response to neo-adjuvant chemotherapy.

The higher accuracy of MRI in local staging and evaluation of tumor response to neo-adjuvant chemotherapy have led to a dramatic increase in limb-salving surgery.

Every patient, with a possible malignant bone lesion, should undergo both plain radiographs and MRI, before invasive procedures are performed.

References


