

**Primary malignant bone tumors of the Spine ñ a pictorial review**

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## 1. Introduction

Primary malignant bone tumors of the spine are relatively rare when compared with metastatic disease and multiple myeloma. The last two usually manifest with multifocal lesions and thus pose little diagnostic dilemma.

The aim of this presentation is to review the imaging findings of primary malignant bone tumors of the spine using a multimodality approach (X-Ray, CT, MR).

### 2.jpg

**Distribution**

General bone			Spine		
freq.	tumor	%	freq.	tumor	%
1 <sup>st</sup>	Osteosarcoma	40	1 <sup>st</sup>	Chordoma	40
2 <sup>nd</sup>	Chondrosarcoma	20	2 <sup>nd</sup>	Chondrossarcoma	20
3 <sup>rd</sup>	Ewing Sarcoma	15	3 <sup>rd</sup>	Ewing Sarcoma	8
4 <sup>th</sup>	Lymphoma	8	4 <sup>th</sup>	Lymphoma	8
5 <sup>th</sup>	MFH / Fibrosarcoma	8	5 <sup>th</sup>	Plasmacytoma	5
6 <sup>th</sup>	Chordoma	5	6 <sup>th</sup>	Osteosarcoma	1
7 <sup>th</sup>	Plasmacytoma	1	7 <sup>th</sup>	MFH / Fibrosarcoma	1
8 <sup>th</sup>	angiosarcoma / hemangiopericytoma	1	8 <sup>th</sup>	angiosarcoma / hemangiopericytoma	1

MFH – Malignant Fibrous Histiocytoma. Repair in the different distribution of malignant neoplasm in general bone and spine. For example osteosarcoma is the most frequent neoplasm in general bone, but it is rare in spine. On the other hand, chordoma is the most frequent in spine, and is relatively rare in general bone.

3.jpg

## PRIMARY MALIGNANT LESIONS OF THE SPINE

- Chordoma
- Chondrosarcoma
- Ewing Sarcoma
- Osteosarcoma
- Primary osseous lymphoma
- Plasmacytoma

## 2. Discussion

Discussion.

3.jpg

## PRIMARY MALIGNANT LESIONS OF THE SPINE

- Chordoma
- Chondrosarcoma
- Ewing Sarcoma
- Osteosarcoma
- Primary osseous lymphoma
- Plasmacytoma

4.jpg

## Chordoma

- Uncommon tumor, accounting for 2-4 % of all primary malignant bone neoplasms
- The most common primary malignant tumor of the spine in adults (excluding lymphoproliferative neoplasms)
- Distribution:
  - 50 % - sacrococcygeal region
  - 30% - spheno-occipital region
  - 15 % - other spinal segments

5.jpg

## Chordoma

- +++ Middle-aged patients (30-60 years)
- Spinal chordomas – 2:1 male-to-female ratio
- Chordomas are slowly growing lesions, with a gradual onset of neurologic symptoms, and are often bulky masses when initially discovered

6.jpg

## Chordoma

### □ Radiology:

- The most frequent radiographic appearance is a destructive lesion of a vertebral body centered in the midline, with a large, associated soft-tissue mass
- Osseous expansion is frequent
- Evidence of **intratumoral calcification** is seen in 50-70% of sacrococcygeal lesions on radiographs and in as many as 90% on CT

7.jpg

## Chordoma

### □ Radiology:

- Lesions of spinal segments above the sacrum are generally less expansile, have calcification in only approximately 30% of cases, and may contain sclerosis
- There may be intervertebral disk involvement, with narrowing, which is unusual for most spinal tumors and simulates infection and may lead to involvement of two adjacent vertebral levels

8.jpg

## Chordoma

- CT:
  - Coronal oblique CT of sacrococcygeal lesions is the optimal method for detecting intervertebral foramen and sacroiliac joint involvement
  - > 50% of cases with soft-tissue mass of low attenuation, which reflects the myxoid-type tissue present pathologically
- MR:
  - low to intermediate signal intensity on T1 and very high signal intensity on T2. MR is superior to CT in depicting the extent.
- Enhancement of chordoma is commonly seen after intravenous administration of contrast

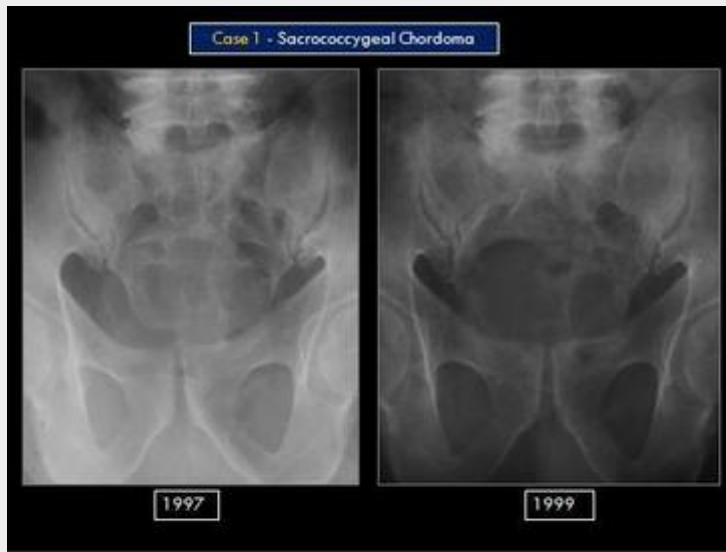
9.jpg

## Chordoma

- Prognosis depends on whether the tumor can be completely resected
- Adjunct radiation therapy may also be employed
- Death is often related to local recurrence and invasion rather than to metastatic disease



10.jpg



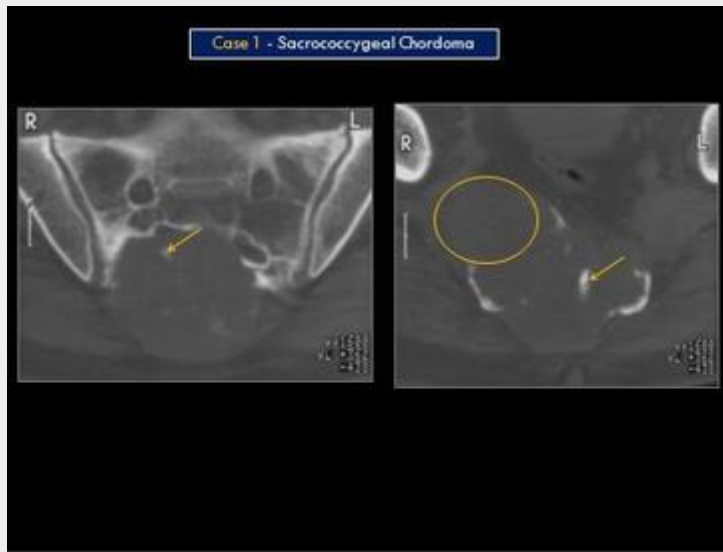
X-Ray and CT – Pelvis X-Ray obtained with two years interval (patient refused treatment in 1997) reveals a slow growing tumor. This case appears in the midline of sacrococcygeal region (the most frequent location for Chordoma) as a expansil lytic lesion, that spares the upper portion of sacrum. The soft-tissue mass has low density (circle), due to the myxoid-type tissue present pathologically. There are intra-tumoral calcifications (arrows) that usually represent destructed bone sequestrum (and not matrix calcification).

11.jpg



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13.jpg



X-Ray and CT – In lumbar spine the lesion usually arises from the transition between the anterior 2/3 with the posterior 1/3 of the vertebral body (place of remnants of notochord). Usually, lumbar chordoma, as seen in this case, has a mixed pattern of lysis and sclerosis. There is a soft tissue mass with calcifications, intervertebral disk involvement with narrowing (can simulate infection!) and partial vertebral body collapse.

14.jpg



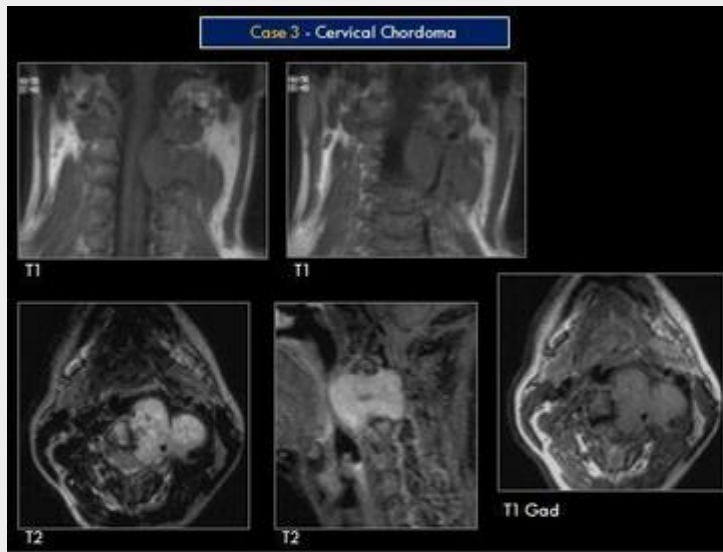
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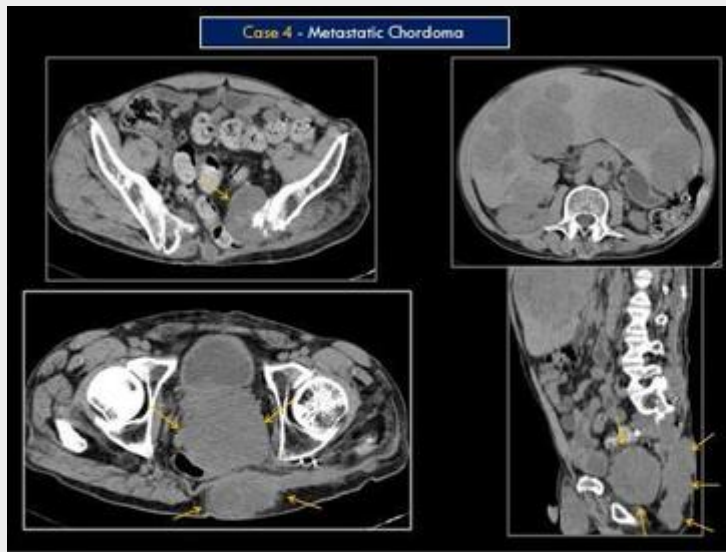
X-Ray, CT and MR – Osteolytic lesion, with a big and lobulated soft-tissue mass, that goes through an enlarged neural foramina extending to epidural and paravertebral space. There is low signal on T1, very high signal on T2 (myxoid content) with a weak but heterogeneous enhancement after gadolinium.

16.jpg



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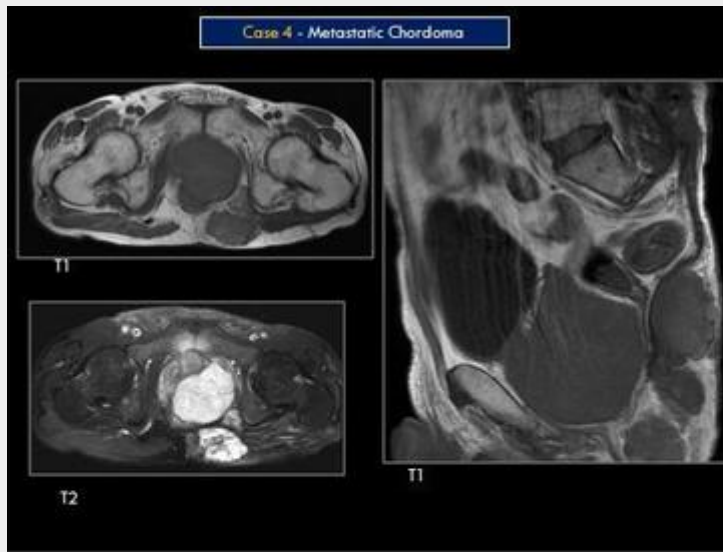
17.jpg



CT and MR – We can see several volumous soft-tissue masses in the pelvis (arrows). The lesions have intermediate signal on T1 and high signal on T2. There are also multiple liver metastasis.



18.jpg



CT and MR – We can see several volumous soft-tissue masses in the pelvis (arrows). The lesions have intermediate signal on T1 and high signal on T2. There are also multiple liver metastasis.

19.jpg

## Chondrosarcoma

- The 2nd most common nonlymphoproliferative primary malignant tumor of the spine
- Spine (+++ Thoracic spine) represents the primary site in 3-12 % of all chondrosarcomas
- Patients typically present with pain and a palpable mass
- 2-4:1 male-to-female ratio
- Mean age is 45 years
- Most lesions represent primary chondrosarcoma
- May also occur when osteochondroma undergoes malignant transformation

20.jpg

## Chondrosarcoma

- Radiography typically reveals bone destruction
- May arise in vertebral body (15%), the posterior elements (40%) or both (45%)
- Characteristic chondroid matrix mineralization (rings and arcs) is apparent on radiographs in 70% of patients, but is better delineated by CT
- Cortical destruction invariably present
- Extension into the surrounding soft tissues is common (best seen by CT/RM), with mineralization usually apparent in the soft-tissue component
- Involvement of adjacent vertebral level by extension through the disk is seen in 35% of lesions

21.jpg

## Chondrosarcoma

- When arising from osteochondroma is seen as thickening at the peripheral cartilaginous cap
- CT – the attenuation of the nonmineralized portion of the lesion is often lower than muscle (high water content of hyaline cartilage)
- MR – low to intermediate signal intensity on T1 and very high signal on T2

## Chondrosarcoma

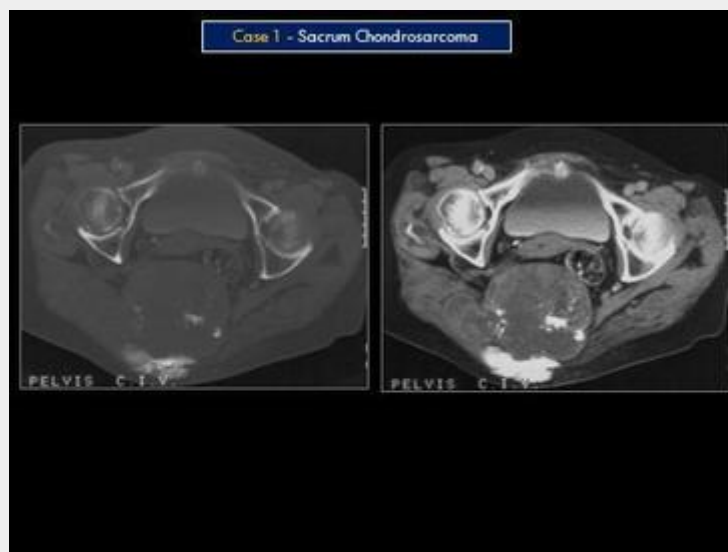
- Treatment is surgical resection, with cure possible in as many as 26 %
- When wide marginal excision cannot be performed, tumor recurrence ultimately causes death in 74% of cases
- Radiation therapy is also used as adjunct treatment
- Because it is frequently a low-grade lesion, survival is often prolonged (mean survival of 5,9 years) and distant metastases are not common

23.jpg



X-Ray and CT – There is a osteolytic lesion associated with a volumous soft-tissue mass, which has several mineralized areas (resulting not only from chondroid matrix mineralization but also due to bone sequestra that result from destruction of the previous osteochondroma). The low attenuation seen at CT in the nonmineralized portion of the mass is typical, and reflects the high water content of hyaline cartilage.

24.jpg



X-Ray and CT – There is a osteolytic lesion associated with a volumous soft-tissue mass, which has several mineralized areas (resulting not only from chondroid matrix mineralization but also due to bone sequestra that result from destruction of the previous osteochondroma). The low attenuation seen at CT in the nonmineralized portion of the mass is typical, and reflects the high water content of hyaline cartilage.

25.jpg

## Ewing Sarcoma

- The most common nonlymphoproliferative primary malignant tumors of the spine in children
- Spine account for 3-10 % of all primary sites of Ewing sarcoma. However metastatic foci of Ewing sarcoma involving the spine are much more common than primary lesions
- +++ 10 – 30 years
- Clinical symptoms are pain and neurologic changes

26.jpg

## Ewing Sarcoma

- Most common location is the sacrococcygeal region, followed by the lumbar and thoracic segments
- Typically centered in the vertebral body
- Radiographs may reveal permeative bone lysis, osseous expansion or sclerosis
- Paraspinal soft-tissue masses are often a prominent feature
- MR – intermediate signal intensity on T1 and intermediate to high signal intensity on T2

## Ewing Sarcoma

- RT and QT are the current mainstays of treatment for spinal lesions, with results approaching 100% for local control and 86% for long-term survival (non-sacral tumors)
- Sacrococcygeal tumors have a worse prognosis, with 62,5 % local control and 25 % long-term survival

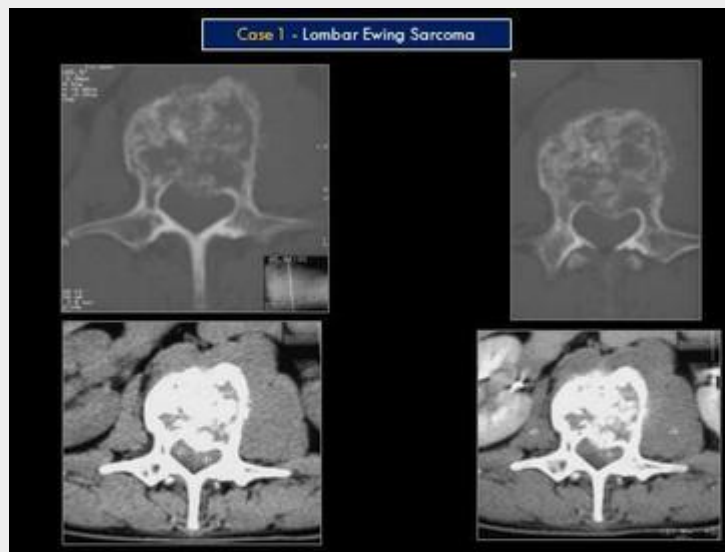


28.jpg



X-Ray, CT and MR – This case has a mixed pattern (lytic and sclerotic), more often seen after treatment. The lesion is centered in the vertebral body – a typical location for Ewing Sarcoma. There is a prevertebral soft-tissue mass, also a prominent feature of these lesions. The MR appearance is nonspecific – intermediate signal intensity on T1 and intermediate to high signal intensity on T2.

29.jpg



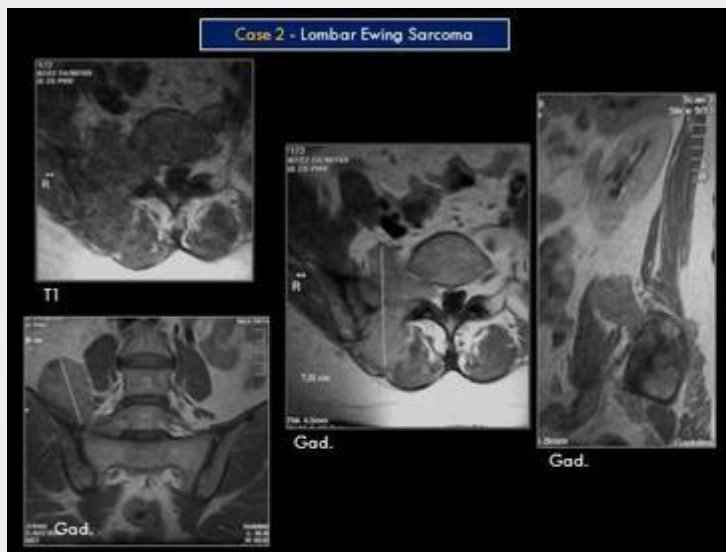
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30.jpg



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31.jpg



MR – The associated soft-tissue mass is the prominent feature of this tumor. The MR findings are nonspecific, with low/intermediate signal on T1, and a weak homogeneous enhancement after gadolinium.

32.jpg

## Osteosarcoma

- Rare, accounting for 0,6% - 3,2% of all osteosarcomas and 5 % of all primary malignant tumors of spine
- Older age (4th decade) than those with appendicular lesions
- +++ Male predominance
- Patients often present with pain and a palpable mass and 80% have neurologic symptoms

33.jpg

## Osteosarcoma

- Predilection for the lumbosacral segments
- In most cases, the vertebral body is primarily involved, albeit eccentrically, but secondary extension into the posterior elements is also common
- May be a secondary lesion: Radiation (5-20 year latent period); associated with Paget disease.

34.jpg

## Osteosarcoma

### □ Radiology:

- Radiographs usually reveal densely mineralized matrix, and an ivory vertebral body may be recognized
- Loss of vertebral height and sparing of the adjacent disk are common
- Purely lytic lesions is rare
- CT and MR – useful for evaluating lesion extent and the soft-tissue involvement
- Lesions with large amounts of matrix mineralization may remain low signal intensity on all MR images

35.jpg

## Osteosarcoma

- The prognosis is dismal, because the lesions are large at presentation and cannot be completely excised in this location
- Adjuvant QT and RT are also often used

36.jpg

## Primary osseous lymphoma

- Uncommon presentation of lymphoma, and must be distinguished from secondary osseous involvement by extraosseous primary disease
- +++ 30 – 60 years
- The lesion is lytic, most frequently moth-eaten or permeative, but it can appear to be of mixed density because of reactive bone formation and prominent endosteal thickening

37.jpg

## Primary osseous lymphoma

- The lesion can enlarge rapidly, giving rise to two features seen on radiography, CT or MRI that can be suggestive of dx:
  - Very large soft tissue mass without extensive cortical destruction.
  - Bone sequestra can be seen
- MR is needed for staging because radiographs do not show the true size and extent of lesion
- Can metastasize to lymph nodes, bone and lung
- Treatment is whole-bone radiation with chemotherapy reserved for disseminated disease



38.jpg



X-Ray and CT – This lesion is difficult to characterize on X-Ray. However CT demonstrates well the typical permeative pattern seen in primary bone lymphoma.

39.jpg



X-Ray and CT – This lesion is difficult to characterize on X-Ray. However CT demonstrates well the typical permeative pattern seen in primary bone lymphoma.

40.jpg



X-Ray, CT and MR – The pattern of this lesion is mainly osteolytic, with partial vertebral body collapse. At MR there is low signal on T1 and high signal on T2. After gadolinium there is little but heterogeneous enhancement. In this case there isn't an associated soft-tissue mass (frequently seen with this kind of tumors).

41.jpg



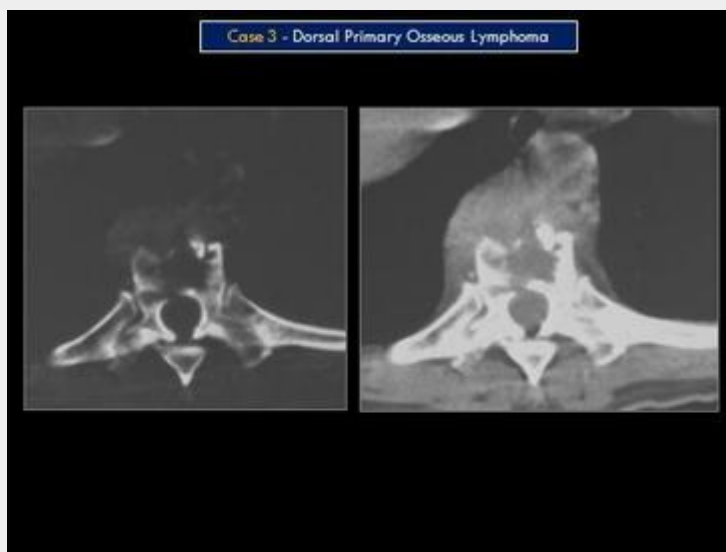
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42.jpg



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43.jpg



CT – Osteolytic lesion located in the vertebral body associated with a soft tissue mass.

44.jpg

## Plasmacytoma

- Neoplastic proliferation of plasma cells. The solitary form of this type of proliferation is called plasmacytoma (as opposed to multiple myeloma)
- Usually patients with > 40 years
- Most cases progress to multifocal or generalized disease within a few years

45.jpg

## Plasmacytoma

- Radiology:
  - Vertebral bodies are a frequent location because they are rich in hematopoietic marrow
  - Lytic expansile geographic lesion, with a relatively narrow zone of transition without sclerotic margins
  - No matrix calcification is present
  - MR – “mini-brain” appearance

46.jpg



X-Ray, CT and MR – Osteolytic lesion affecting a lumbar vertebral body, with relative preservation of cortical bone – “empty vertebra appearance”. We can see several lytic areas, separated by vertical bone bridges, with preservation of vertebral body height. The lesion has low signal on T1 and high signal on T2, with enhancement after gadolinium.

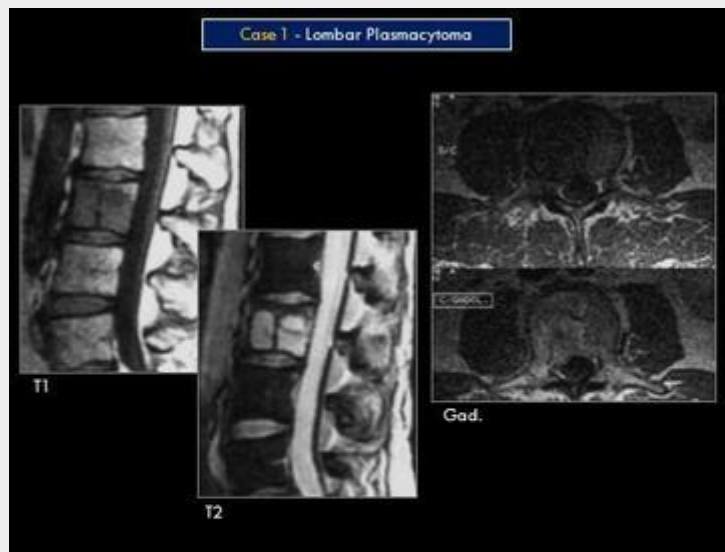


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48.jpg



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49.jpg



X-Ray and CT – Purely lytic lesion with associated soft-tissue mass.

50.jpg



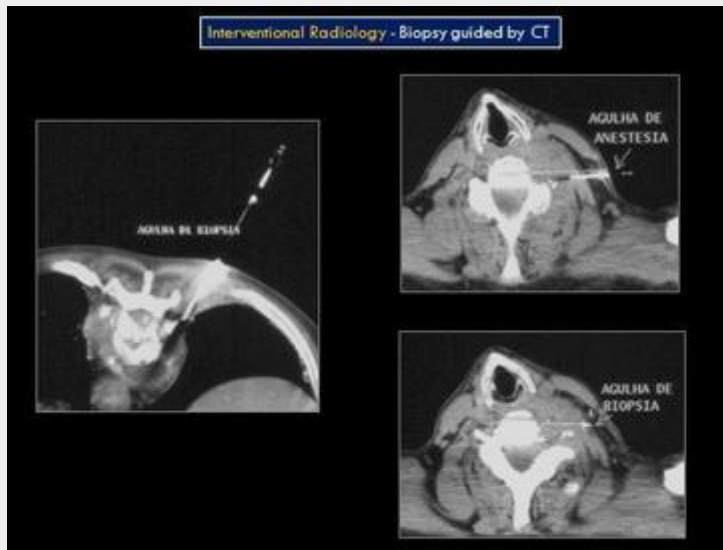
X-Ray and MR – There is fracture with vertebral body collapse, being apparent a medullary osteolytic pattern with relative preservation of cortical vertebral plates. The lesion is low signal on T1 and high signal on T2, with good enhancement after gadolinium.

51.jpg



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52.jpg



Interventional Radiology – CT guided biopsy is instrumental for obtaining tissue for histologic analysis. Biopsy can be made in the bone lesion or in the often associated soft-tissue mass, at any level of the spine.

Conclusions:

- 1) The identification of a solitary spinal bony lesion challenges the differential diagnosis, and imaging is instrumental for clinical decision making.
- 2) Radiological and clinical findings quite often are sufficiently specific to address the correct diagnosis.
- 3) When necessary CT guided biopsy is the best way for obtaining tissue for histologic analysis.

### 3. References

References:

## References

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## 4. Mediafiles

44.jpg

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36.jpg

## Primary osseous lymphoma

- Uncommon presentation of lymphoma, and must be distinguished from secondary osseous involvement by extraosseous primary disease
- +++ 30 – 60 years
- The lesion is lytic, most frequently moth-eaten or permeative, but it can appear to be of mixed density because of reactive bone formation and prominent endosteal thickening

37.jpg

## Primary osseous lymphoma

- The lesion can enlarge rapidly, giving rise to two features seen on radiography, CT or MRI that can be suggestive of dx:
  - Very large soft tissue mass without extensive cortical destruction.
  - Bone sequestra can be seen
- MR is needed for staging because radiographs do not show the true size and extent of lesion
- Can metastasize to lymph nodes, bone and lung
- Treatment is whole-bone radiation with chemotherapy reserved for disseminated disease

45.jpg

## Plasmacytoma

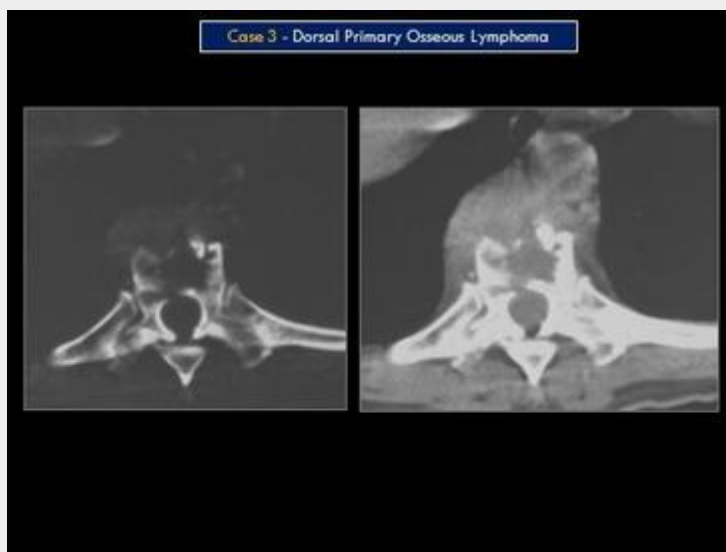
- Radiology:
  - Vertebral bodies are a frequent location because they are rich in hematopoietic marrow
  - Lytic expansile geographic lesion, with a relatively narrow zone of transition without sclerotic margins
  - No matrix calcification is present
  - MR – “mini-brain” appearance

53.jpg

## References

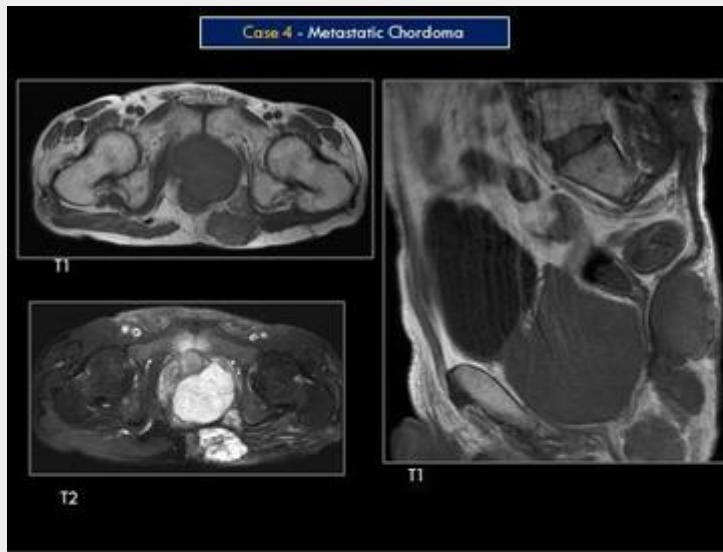
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43.jpg



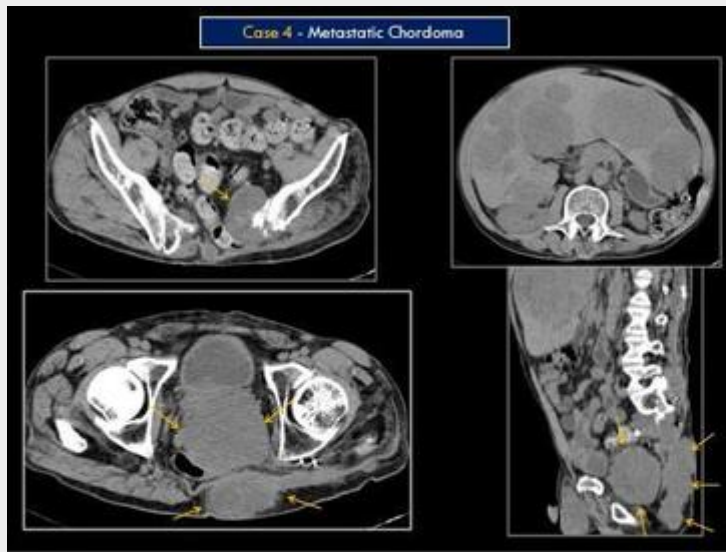
CT – Osteolytic lesion located in the vertebral body associated with a soft tissue mass.

18.jpg



CT and MR – We can see several volumous soft-tissue masses in the pelvis (arrows). The lesions have intermediate signal on T1 and high signal on T2. There are also multiple liver metastasis.

17.jpg



CT and MR – We can see several volumous soft-tissue masses in the pelvis (arrows). The lesions have intermediate signal on T1 and high signal on T2. There are also multiple liver metastasis.

52.jpg



Interventional Radiology – CT guided biopsy is instrumental for obtaining tissue for histologic analysis. Biopsy can be made in the bone lesion or in the often associated soft-tissue mass, at any level of the spine.

2.jpg

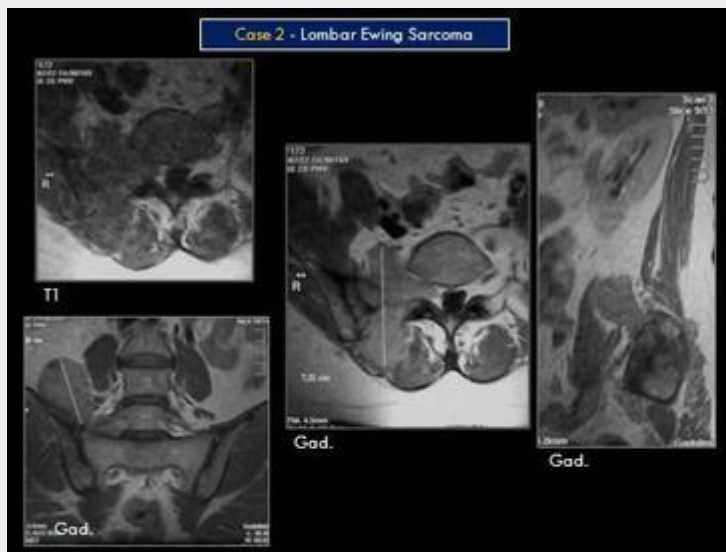
### Distribution

General bone			Spine		
freq.	tumor	%	freq.	tumor	%
1 <sup>st</sup>	Osteosarcoma	40	1 <sup>st</sup>	Chordoma	40
2 <sup>nd</sup>	Chondrosarcoma	20	2 <sup>nd</sup>	Chondrossarcoma	20
3 <sup>rd</sup>	Ewing Sarcoma	15	3 <sup>rd</sup>	Ewing Sarcoma	8
4 <sup>th</sup>	Lymphoma	8	4 <sup>th</sup>	Lymphoma	8
5 <sup>th</sup>	MFH / Fibrosarcoma	8	5 <sup>th</sup>	Plasmacytoma	5
6 <sup>th</sup>	Chordoma	5	6 <sup>th</sup>	Osteosarcoma	1
7 <sup>th</sup>	Plasmacytoma	1	7 <sup>th</sup>	MFH / Fibrosarcoma	1
8 <sup>th</sup>	angiosarcoma / hemangiopericytoma	1	8 <sup>th</sup>	angiosarcoma / hemangiopericytoma	1

MFH – Malignant Fibrous Histiocytoma. Repair in the different distribution of malignant neoplasm in general bone and spine. For example osteosarcoma is the most frequent neoplasm in general bone, but it is rare in spine. On the other hand, chordoma is the most frequent in spine, and is relatively rare in general bone.

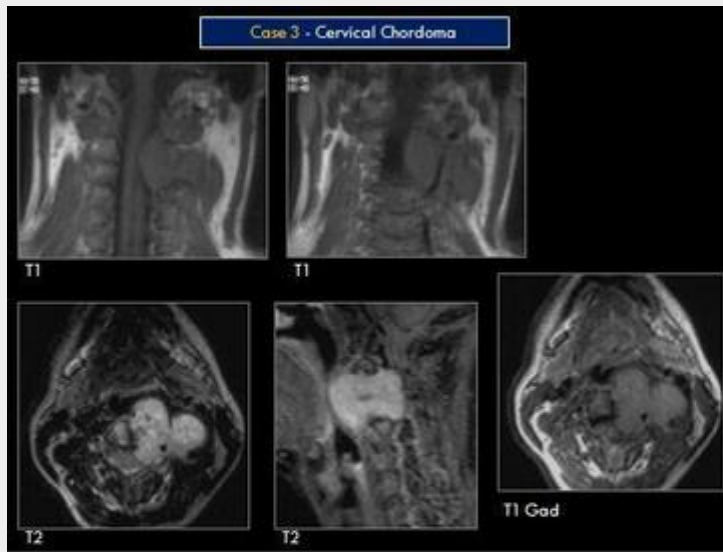


31.jpg



MR – The associated soft-tissue mass is the prominent feature of this tumor. The MR findings are nonspecific, with low/intermediate signal on T1, and a weak homogeneous enhancement after gadolinium.

16.jpg



X-Ray, CT and MR – Osteolytic lesion, with a big and lobulated soft-tissue mass, that goes through an enlarged neural foramina extending to epidural and paravertebral space. There is low signal on T1, very high signal on T2 (myxoid content) with a weak but heterogeneous enhancement after gadolinium.

15.jpg



X-Ray, CT and MR – Osteolytic lesion, with a big and lobulated soft-tissue mass, that goes through an enlarged neural foramina extending to epidural and paravertebral space. There is low signal on T1, very high signal on T2 (myxoid content) with a weak but heterogeneous enhancement after gadolinium.

47.jpg



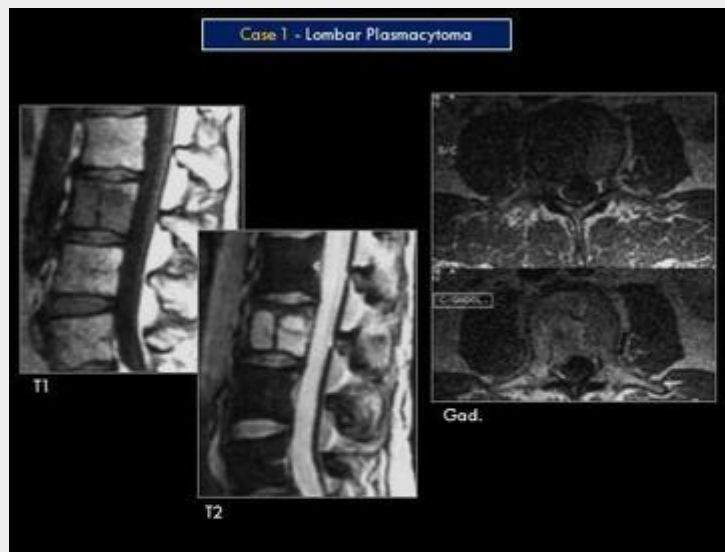
X-Ray, CT and MR – Osteolytic lesion affecting a lumbar vertebral body, with relative preservation of cortical bone – “empty vertebra appearance”. We can see several lytic areas, separated by vertical bone bridges, with preservation of vertebral body height. The lesion has low signal on T1 and high signal on T2, with enhancement after gadolinium.

46.jpg



X-Ray, CT and MR – Osteolytic lesion affecting a lumbar vertebral body, with relative preservation of cortical bone – “empty vertebra appearance”. We can see several lytic areas, separated by vertical bone bridges, with preservation of vertebral body height. The lesion has low signal on T1 and high signal on T2, with enhancement after gadolinium.

48.jpg



X-Ray, CT and MR – Osteolytic lesion affecting a lumbar vertebral body, with relative preservation of cortical bone – “empty vertebra appearance”. We can see several lytic areas, separated by vertical bone bridges, with preservation of vertebral body height. The lesion has low signal on T1 and high signal on T2, with enhancement after gadolinium.

42.jpg



X-Ray, CT and MR – The pattern of this lesion is mainly osteolytic, with partial vertebral body collapse. At MR there is low signal on T1 and high signal on T2. After gadolinium there is little but heterogeneous enhancement. In this case there isn't an associated soft-tissue mass (frequently seen with this kind of tumors).

41.jpg



X-Ray, CT and MR – The pattern of this lesion is mainly osteolytic, with partial vertebral body collapse. At MR there is low signal on T1 and high signal on T2. After gadolinium there is little but heterogeneous enhancement. In this case there isn't an associated soft-tissue mass (frequently seen with this kind of tumors).



40.jpg



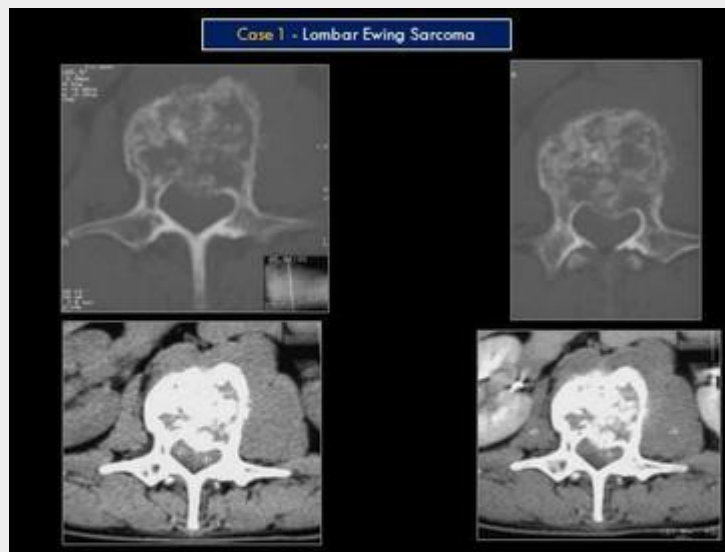
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30.jpg



X-Ray, CT and MR – This case has a mixed pattern (lytic and sclerotic), more often seen after treatment. The lesion is centered in the vertebral body – a typical location for Ewing Sarcoma. There is a prevertebral soft-tissue mass, also a prominent feature of these lesions. The MR appearance is nonspecific – intermediate signal intensity on T1 and intermediate to high signal intensity on T2.

29.jpg



X-Ray, CT and MR – This case has a mixed pattern (lytic and sclerotic), more often seen after treatment. The lesion is centered in the vertebral body – a typical location for Ewing Sarcoma. There is a prevertebral soft-tissue mass, also a prominent feature of these lesions. The MR appearance is nonspecific – intermediate signal intensity on T1 and intermediate to high signal intensity on T2.

28.jpg



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13.jpg



X-Ray and CT – In lumbar spine the lesion usually arises from the transition between the anterior 2/3 with the posterior 1/3 of the vertebral body (place of remnants of notochord). Usually, lumbar chordoma, as seen in this case, has a mixed pattern of lysis and sclerosis. There is a soft tissue mass with calcifications, intervertebral disk involvement with narrowing (can simulate infection!) and partial vertebral body collapse.

14.jpg



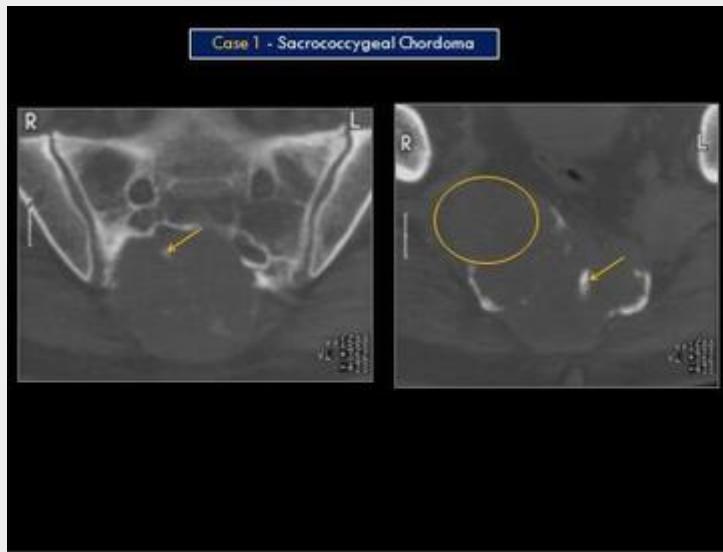
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11.jpg



X-Ray and CT – Pelvis X-Ray obtained with two years interval (patient refused treatment in 1997) reveals a slow growing tumor. This case appears in the midline of sacrococcygeal region (the most frequent location for Chordoma) as a expansil lytic lesion, that spares the upper portion of sacrum. The soft-tissue mass has low density (circle), due to the myxoid-type tissue present pathologically. There are intra-tumoral calcifications (arrows) that usually represent destructed bone sequestrum (and not matrix calcification).

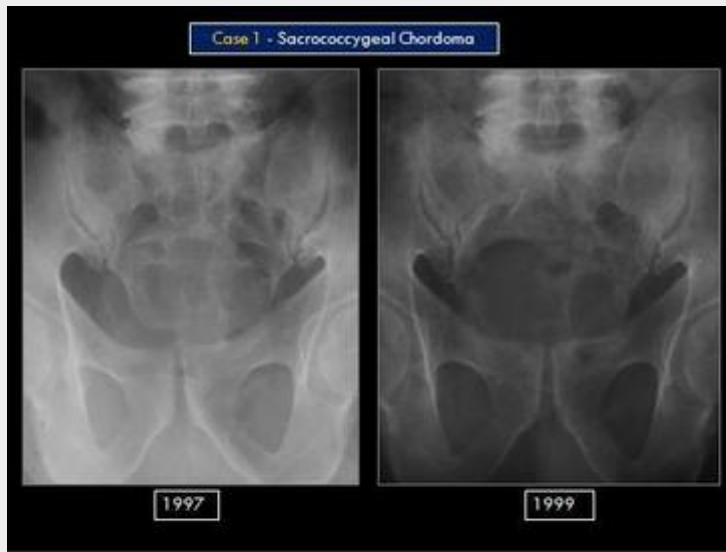
12.jpg



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10.jpg



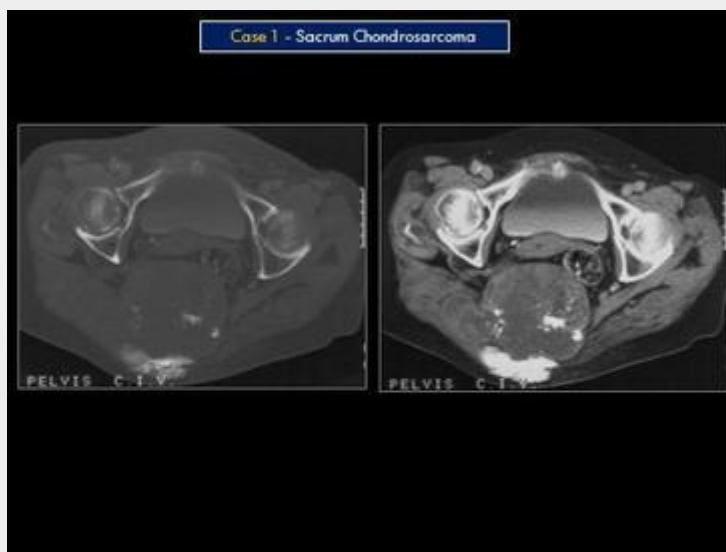
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49.jpg



X-Ray and CT – Purely lytic lesion with associated soft-tissue mass.

24.jpg



X-Ray and CT – There is a osteolytic lesion associated with a volumous soft-tissue mass, which has several mineralized areas (resulting not only from chondroid matrix mineralization but also due to bone sequestra that result from destruction of the previous osteochondroma). The low attenuation seen at CT in the nonmineralized portion of the mass is typical, and reflects the high water content of hyaline cartilage.

23.jpg



X-Ray and CT – There is a osteolytic lesion associated with a volumous soft-tissue mass, which has several mineralized areas (resulting not only from chondroid matrix mineralization but also due to bone sequestra that result from destruction of the previous osteochondroma). The low attenuation seen at CT in the nonmineralized portion of the mass is typical, and reflects the high water content of hyaline cartilage.

39.jpg



X-Ray and CT – This lesion is difficult to characterize on X-Ray. However CT demonstrates well the typical permeative pattern seen in primary bone lymphoma.

38.jpg



X-Ray and CT – This lesion is difficult to characterize on X-Ray. However CT demonstrates well the typical permeative pattern seen in primary bone lymphoma.

51.jpg



X-Ray and MR – There is fracture with vertebral body collapse, being apparent a medullary osteolytic pattern with relative preservation of cortical vertebral plates. The lesion is low signal on T1 and high signal on T2, with good enhancement after gadolinium.

50.jpg



X-Ray and MR – There is fracture with vertebral body collapse, being apparent a medullary osteolytic pattern with relative preservation of cortical vertebral plates. The lesion is low signal on T1 and high signal on T2, with good enhancement after gadolinium.