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REVIEW ARTICLE

Primary malignant tumors of the spine

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Received 20 February 2014; *Accepted* 5 March 2014

Citation: Kakarla Subbarao (2014) Primary malignant tumors of the spine. J Med Sci Res 2(2):76-83. DOI: http://dx.doi.org/10.17727/JMSR.2014/2-015

Introduction

Most common malignant tumors involving the vertebral column are metastatic. Primary malignant tumors involving the axial skeleton are rare. The primary function of spine is anatomical, and is involved in the erect posture of human beings. The physiological function is primarily hemopoietic. The segments of the spine include cervical with seven vertebrae, dorsal with twelve vertebrae, lumbar with five vertebrae, sacral with fused five vertebrae and coccygeal with fused four vertebrae.

Imaging methods of spine include conventional, CT, MRI, Radio nuclide, PET CT and Angiography. Conventional radiology most often reveals the nature of the lesion. However, when the lesions are multiple routine skeletal survey may not be adequate. MRI Radionuclide scintigraphy and PET CT scan are more sensitive and may pickup multiple lesions.

Myeloma, a common malignant tumor in old age manifests as 1) Solitary – plasmacytoma, 2) Multiple myeloma, and 3) Poems syndrome

Plasmacytoma

It is a solitary lesion and can be encountered in any bone. However, it is common in the spine. Most of the benign looking plasmacytomas when followed may develop into multiple myeloma eventually. Radiologically, a plasmacytoma is an expanding lytic lesion without any calcification or new bone formation. When it involves the vertebral body a

compression fracture may be seen. When marked compression is seen vertebral plana is noted (Figure 1abc). To differentiate from metastasis, it is stated that in metastasis pedicles are involved earlier as they are vascular (2ab). However, when a large part of the body is involved in myeloma, pedicles also may be destroyed (Figure 3abc).

CT and MRI demonstrate lytic areas with struts of bone interspersed. In MRI, plasmacytoma shows a classical minibrain appearance (Figure 4abc). At times, it is difficult to differentiate hemangioma from plasmacytoma (Figure 5ab). It may remain localized for many years but more than 30% progress quite rapidly to generalized myelomatosis with a latent interval of 5-10 years. Radiological features are listed in table 1.

Table 1: Radiological features of plasmacytoma

- It is typically lytic and destructive. Sclerosis is rare.
- Bone expansion, which may be considerable with thinning of the overlying cortex, is common.
- The margins are usually well defined and sharply demarcated and an associated soft tissue mass is frequently seen.
- Large lesions in flat bones may assume a soap –bubble appearance.
- "Solitary osteolytic lesion in a vertebral body in a patient in late middle age should always be considered as a Plasmacytoma".
- Following the diagnosis of plasmacytoma, MRI of the spine and pelvis is indicated to identify additional lesions, to rule out multiple myeloma, which may be seen in up to 80% of cases.
- Occult lesions may also be demonstrated by [F-18] FDG-PET.

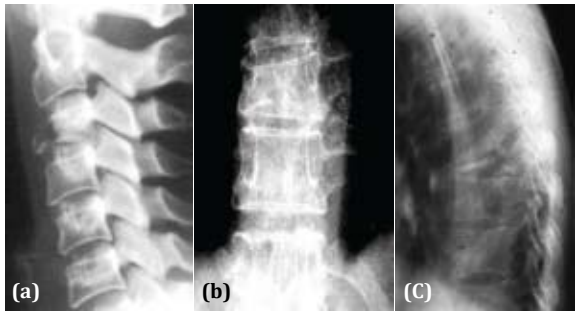

Figure 1ab: Plasmacytoma with collapse of C3, T10
Figure 1c: Plasmacytoma – vertebra plana T8

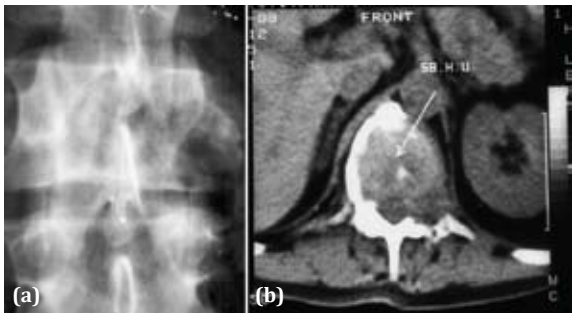

Figure 2ab: a. Plasmacytoma of L2, b. CT guided biopsy

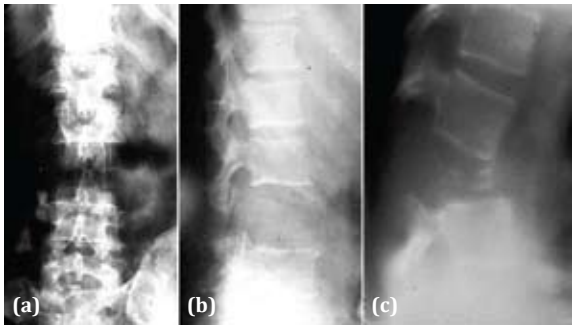

Figure 3abc: Plasmacytoma - Complete destruction of body of L3 including the posterior elements.

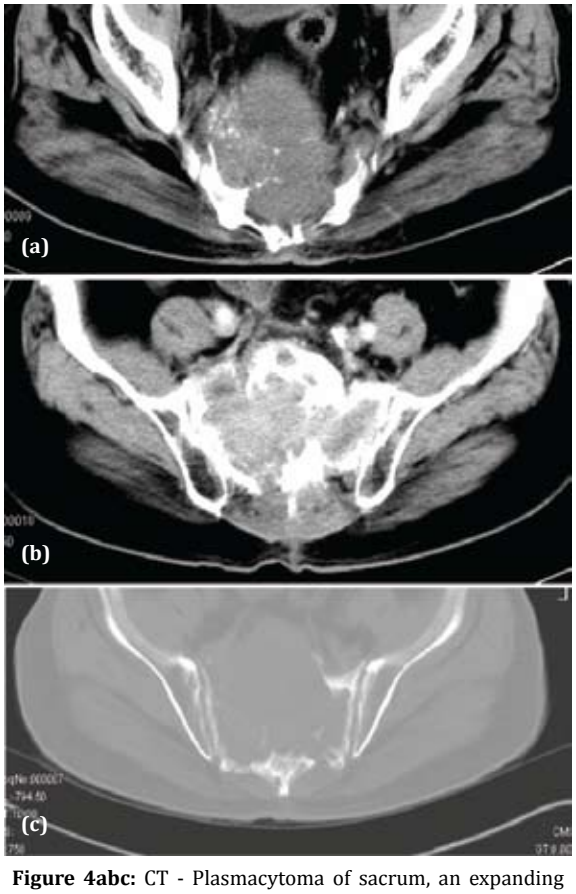

Figure 4abc: CT - Plasmacytoma of sacrum, an expanding lytic lesion

Multiple myeloma (MM)

MM is a clonal B-lymphocyte neoplasm of terminally differentiated plasma cells and it is most common primary malignant tumor of bone. The median age at diagnosis is 55 years. Higher incidence is noted in men. Clinical symptoms include fatigue, generalized bone pains with or without associated fracture. The types of multiple myeloma are listed in table 2.

Table 2: Four distinct forms of involvement have been described:

- Diffuse skeletal involvement (myelomatosis),
- Diffuse skeletal osteopenia
- Sclerosing myeloma
- Plasmacytoma, distribution of malignant plasma cells in MM includes sites that show normal areas of active hematopoiesis in adults, lesions are osteolytic with discrete margins and uniform size, common sites are vertebrae, pelvis, skull, ribs, sternum and proximal appendages.

It radiologically shows a spectrum of manifestations. The bones particularly, in axial skeleton may present with multiple compression fractures. In 10-15% of cases, classical punched out lesions of the skull may be noted. In long tubular bones, pathological fractures are common with associated soft tissue swelling. In early skeletal lesions, moth eaten type of appearance is observed without any periosteal reaction (Figure 6ab). MRI is more sensitive to depict bone marrow lesions, whereas radionuclide scintigraphy with technetium 99 compounds is not that sensitive (Figure 7abcdef). The radiological appearances are listed in Table 3.

Table 3: Multiple Myeloma Radiological Spectrum

- Normal
- Osteopenia
- Compression #s of spine
- Moth eaten
- Punched out lesions
- Geographic areas of lysis
- Sclerotic lesions (Poems)
- Soft tissue masses
- Plasmacytoma

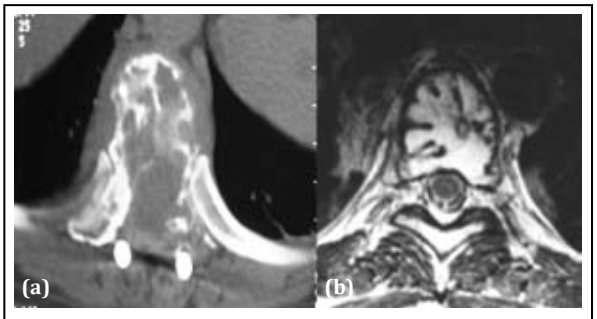

Figure 5ab: a. CT (post op.) – b. MRI – Minibrain appearance

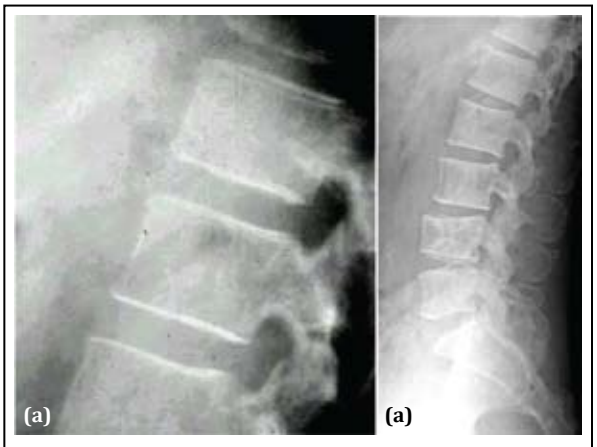

Figure 6ab: Multiple Myeloma – a. Diffuse osteopenia with loss of trabeculae, b. lytic lesions in the bodies.

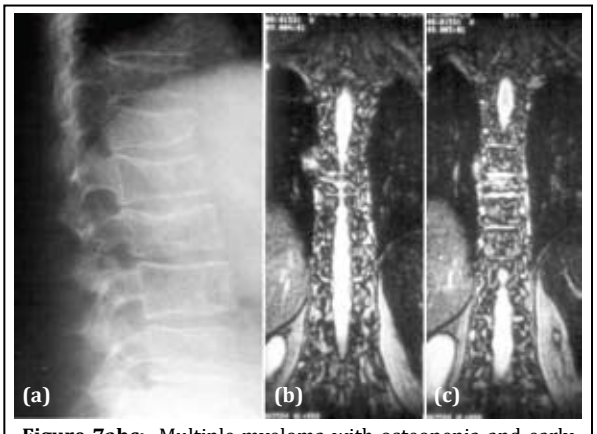

Figure 7abc: Multiple myeloma with osteopenia and early collapse of vertebral bodies BC MRI of the same patient.

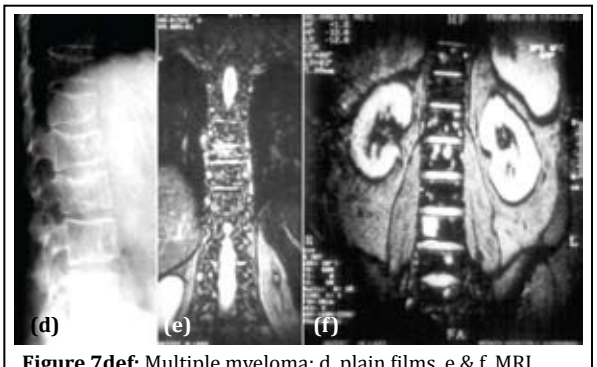

Figure 7def: Multiple myeloma: d. plain films, e & f. MRI

Poems syndrome

'P' stands for polyneuropathy, 'O' stands for organomegaly, 'E' stands for endcrinal changes, 'M' stands for M protein and 'S' stands for sclerotic changes in the bone. All of these may not be present all the time.

Radiologically, sclerotic changes in the bone may be unifocal or multifocal. At times, lytic lesions may be also noted (Figure 8ab).

In the differential diagnosis particularly in older people metastasis must be considered. Table 4 shows the radiological differentiation between myeloma and metastases. Differential diagnostic features are listed in table 4.

Table 4: Myeloma Vs Metastases

	Myeloma	***Metastasis***
Vertebral pedicle involvement	Late	Early
Soft tissue	Masses ++	Mass only with fracture
Mandible involvement	Involved	Late
Mixed lesions	Unusual	Not unusual
IV disc involvement	Involved	Rare
Facial bones involvement	Involved	Rare

Figure 8a: Sclerotic myeloma – diffuse sclerosis of all the lumbar vertebrae

Figure 8b: Sclerotic lesion in the femur in Poems syndrome

Chondrosarcoma

This is also a rare tumor and may involve the vertebral column. Radiological findings include lytic lesion with sclerosis and cartilaginous calcifications (Figure 9abcd). CT helps in identification of early calcifications. CT allows optimal detection and characterization of matrix mineralization, endosteal scalloping. MR imaging depicts the extent of marrow involvement and demonstration of soft tissue extension with mass formation. MRI shows the cartilaginous nature of the lesion by bright signals in T2 (Figure 10abcd).

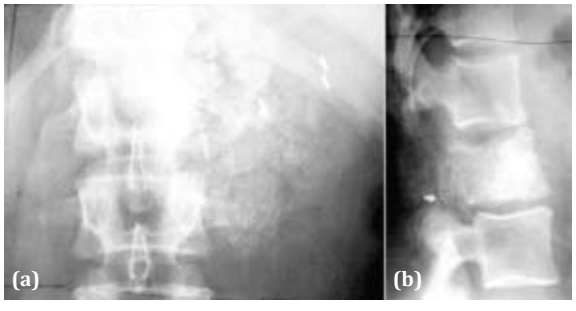

Figure 9ab: Chondrosarcoma of L1. Note the paravertebral ring like calcifications

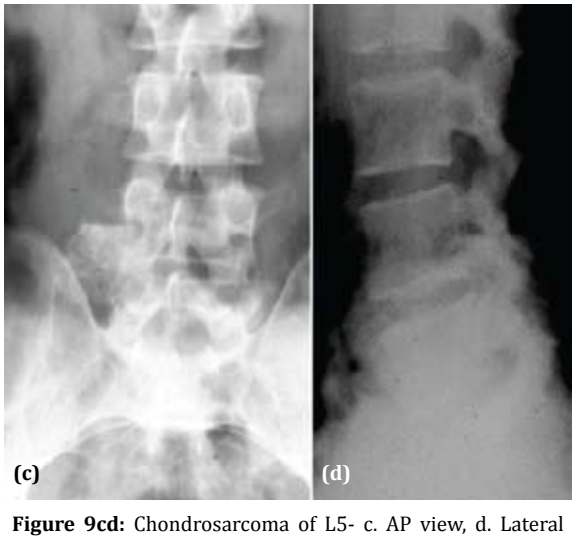

Figure 9cd: Chondrosarcoma of L5- c. AP view, d. Lateral view

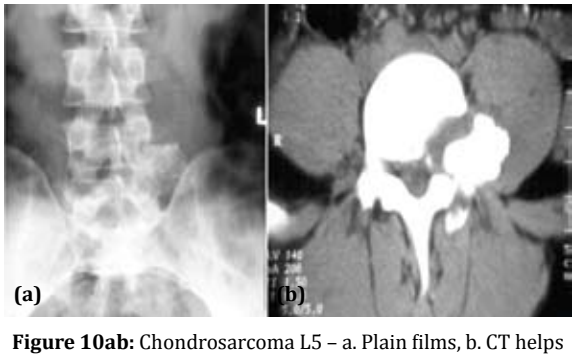

Figure 10ab: Chondrosarcoma L5 – a. Plain films, b. CT helps in identification of small calcifications

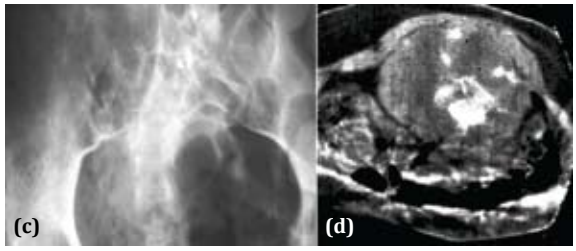

Figure 10cd: Chondrosarcoma of sacrum –c. Plain, d. MRI

Ewing's sarcoma

It constitutes 10% of all primaries. The incidence of M:F :: 2:1. It is not common in the vertebral column. 75% of the lesions occur in pelvic girdle and long bones. Chromosomal abnormality is translocation of (t11;22). Biopsy is the final confirmation for diagnosis.

Radiological findings include permiative lytic lesion with a large soft tissue mass, on occasion diffuse sclerosis is noted simulating an ivory vertebra (Figure 11ab)

"The gross anatomy (as revealed in radiographs) is often a safer guide to a correct clinical conception of the disease than the variable and uncertain structure of a small piece of tissue"- Ewing - 1922.

The differential diagnosis of Ivory vertebra seen on plain films is listed in table 5.

Table 5: Differential diagnosis of ivory vertebra.

- Ewing's sarcoma
- Metastasis - Solitary
- Lymphoma
- Paget's disease
- Osteosarcoma
- Chronic osteomyelitis
- Melorheostosis
- Chordoma (rare)
- Myeloma (rare)
- Osteoblastoma

Primary osteosarcoma

This is another rare tumor that involves the vertebral column. Several instances have been reported involving single vertebral body of spine more commonly sacrum. Radiologically, both lytic and sclerotic changes are noted with new tumor bone formation. When it occurs in the sacrum it may simulate an ivory body (Figure 12). When sclerotic metastasis occur in a solitary vertebra it is difficult to differentiate from primary osteosarcoma.

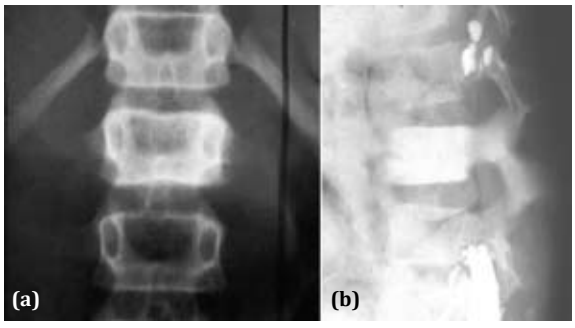

Figure 11ab: Ewing's sarcoma – Ivory vertebra of L1.

Figure12: Primary osteosarcoma involving the sacrum.

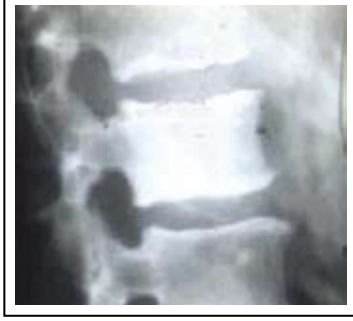

Figure 13a: Lymphoma (Ivory vertebra) involving the body of L1

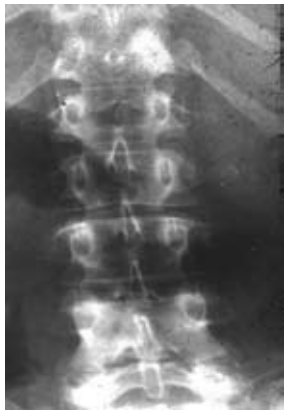

Figure 13b: Hodgkin's lymphoma. Note the ivory pedicles

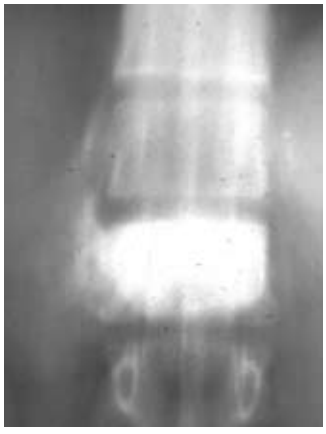

Figure 13c: Hodgkin's lymphoma – conventional tomogram.

Primary lymphoma

Primary lyphoma of bone is uncommon. However, when the bone is involved, radiological findings include both lytic and sclerotic changes with soft tissue swelling. In Hodgkin's type of lymphoma sclerotic lesions predominate. Occasionally, only pedicles of the spine may be involved (Figure13abc).

Primary osseous malignant fibrous histiocytoma (MFH)

Formerly used to be called fibrosarcoma which does not form osteoid or chondroid matrix, it is rare in the vertebral column.

Radiologically, most primary osseous MFHs are purely osteolytic and range in aggressiveness from geographic, with a variable width in the zone of transition, to permeative patterns. Internal calcification, bone formation, periosteal reaction and sequestra within osseous lesions are uncommon findings (Figure 14ab). Histiologically, storiform type of spindle cells are noted.

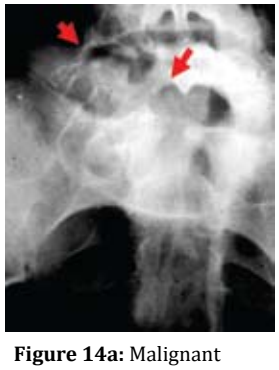

Figure 14a: Malignant fibrous histiocytoma involving L5 and sacrum on right side.

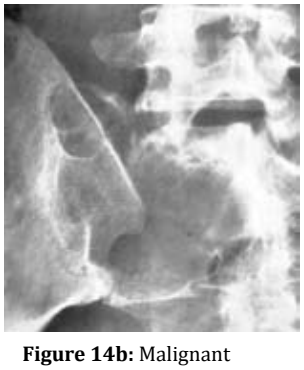

Figure 14b: Malignant fibrous histiocytoma of the sacrum involving right iliac bone

Chordoma

This locally malignant lesion occurs in the vertebral column in 90% of the cases, 55% in sacrum and 25% in clivus and rest of the spine 20%. It is common in the cervical spine as compared to the rest of the spine. The age group is between 45 & 70, but no age is exempt.

Radiologically, majority of the cases show extensive osteolysis of the sacrum with few areas of osteoblastic reaction. A large soft tissue mass is noted with few calcifications. The calcifications of chondroid nature are common in chondroid type of chordoma and are extensive (Figure 15abcde). In the differential diagnosis, plasmacytoma, metastasis and chondrosarcoma may be considered. Aggressive benign lesions such as aneurysmal bone cyst, myxopapillary ependymoma and aggressive giant cell tumor may also be under consideration particularly when it occurs in the sacrum. In the cervical spine TB of the spine comes under differential diagnosis. Histiologically, the physalipherus cell is characteristic (Figure 15f).

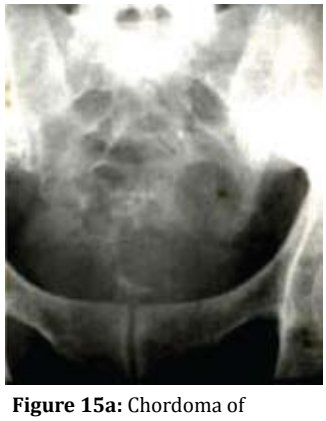

Figure 15a: Chordoma of sacrum

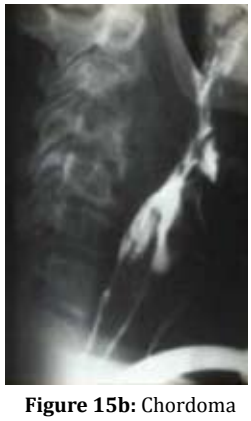

Figure 15b: Chordoma C. Spine with soft tissue swelling compressing the pharynx

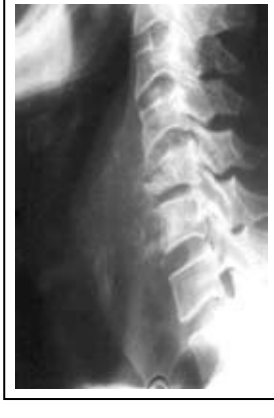

Figure 15c: Chordoma C. Spine

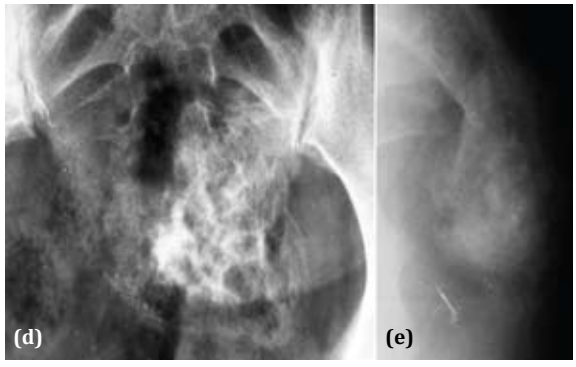

Figure 15de: Chondroid chordoma – d. AP, e. Lateral of sacrum.

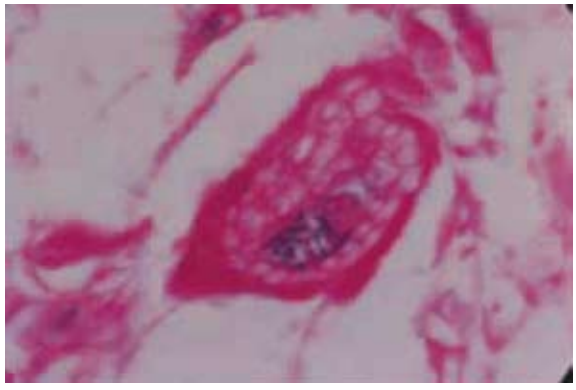

Figure 15f: Chordoma – Physalipherus cell.

Myxopapillary ependymoma

It is a rare tumor involving the sacrum. Radiologically, it is a lytic lesion which is lobulated and is difficult to make a diagnosis (Figure 16). CT and MRI may help in tissue differentiation. Histology is the only way to make a diagnosis (Figure 17abcd). An admixture of fibrillated & epitheloid cells with myxoid connective tissue stroma and fusiform cells in palisading arrangement mixed with mucin deposition are noted. Glial fibrillary acidic protein (GFAP), S-100 protein is positive.

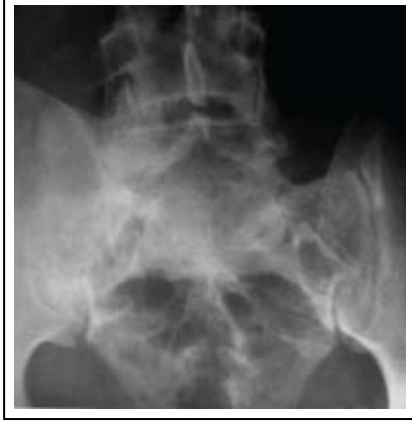

Figure16: Myxopapillary ependymoma of the sacrum – Note lytic and sclerotic changes of non – specific nature.

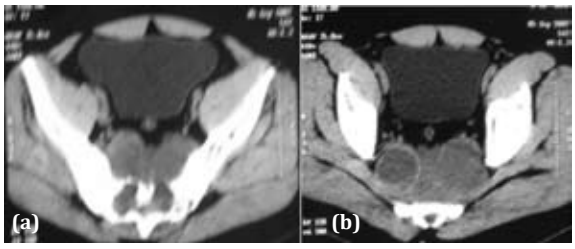

Figure 17ab: CT – Myxopapillary ependymoma of sacrum. Lytic and sclerotic changes with cyst like soft tissue masses.

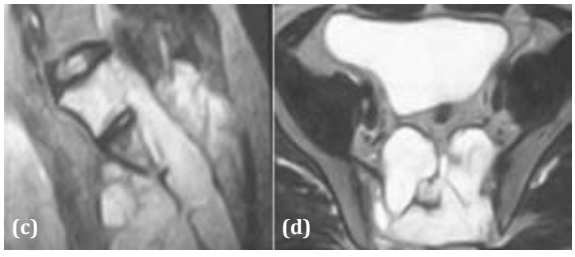

Figure 17cd: MRI – Myxopapillary ependymoma of sacrum. Cyst like soft tissue masses in a lytic sacrum.

Conclusion

Primary malignant tumours of the vertebral column are rare. Whenever, an isolated vertebral lesion is noted radiographically a systemic examination for other lesions should be done, preferably radionuclide scintigraphy. Most of the time, plain films would help. However, small lesions may be missed. Hence, CT scan and MRI should be used to identify the small lesions and early calcifications. Radionuclide scintigraphy would help in identifying multiple lesions, but this may not be helpful in multiple myeloma and other bone marrow lesions. Finally biopsy should be done under the imaging guidance for confirmation of the diagnosis.

Conflict of interest

The author wishes to express that he has no conflict of interest.

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