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Primary Sacral Lymphoma: A Case Report and Review of the Literature

Primer Sakral Lenfoma: Olgu Sunumu ve Literatürün Gözden Geçirilmesi

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ABSTRACT

AIM: Primary sacral lymphoma is rare. It usually presents in elderly males as low back ache and radiculopathy. Case report: We hereby report a case of primary sacral lymphoma in a 53-year-old male who presented with low back pain. Imaging showed a sacral lesion which on histopathology proved to be a non-Hodgkin's lymphoma of B cell lineage.

MATERIAL and METHODS: The case is presented for its rarity and for the reason that it mimicked a primary bone tumor. Its recognition is important because it has an excellent prognosis.

CONCLUSION: Primary sacral epidural lymphoma should be considered in the differential diagnosis of a sacral mass.

KEYWORDS: Sacral tumors, Sacral lymphoma, Epidural lymphoma, Primary B cell lymphoma

ÖZ

AMAÇ: Primer sakral lenfoma nadir görülür. Primer sakral lenfoma genellikle yaşlı erkek hastalarda bel ağrısı ve radikülopati şeklinde ortaya çıkar.

YÖNTEM ve GEREÇLER: Primer sakral lenfoması olan hasta belağrısı şikayeti ile kliniğimize başvurdu. Görüntüleme çalışmalaında sakral lezyon saptanan hastadan alınan biyopsinin histopatolojik incelemesi sonucunda B hücre orjinli non hodgkin lenfoma tanısı konuldu. Bu vaka hem nadir görülen bir vaka hem de primer kemik tümörü görünümünü taklit etmiş olması nedeni ile sunulmaktadır. Bu hastalığın tanısının konulması hastalığın mükemmel prognozu nedeniyle çok önemlidir.

SONUÇ: Sakral kitlesi olan hastalarda primer sakral epidural lenfoma ayırıcı tanı olarakda düşünülmelidir.

ANAHTAR SÖZCÜKLER: Sakral tümörler, Sakral lenfoma, Epidural lenfoma, Primer B hücreli lenfoma

INTRODUCTION

The commonest primary tumor of sacrum is chordoma. Primary lymphoma of the sacrum is very rare (4). It usually presents in elderly males (13). The usual presentation is low back pain with or without radiculopathy. On imaging, sacral lymphomas can mimic other tumorous lesions and hence differentiating it from these lesions is important as the overall prognosis of primary sacral lymphomas is good (7).

CASE REPORT

Presentation

A 53-year-old male reported to a general physician with history of low back ache of 2 months duration. His general physical, systemic and neurological examinations were normal. He was prescribed analgesics and was advised X-ray lumbosacral spine. X-ray was found to be normal. Patient continued to have pain despite taking analgesics which progressively involved his right lower limb over a period of time. He reported back to his treating physician who on motor examination noticed plantar flexion of right ankle to be 4/5. There was 30% sensory loss along the lateral border of right foot. Straight leg raising test was restricted on both sides. Deep tendon reflexes were normal.

Imaging

He was investigated with MRI lumbosacral spine which revealed altered signal intensity on the right side of S1 and S2 vertebral bodies and a soft tissue extension from L5 to S2 in the sacral epidural space on the right side. The soft tissue was isointense on T1-weighted sequence and hyperintense on T2-weighted.The lesion was causing compression of right S1 nerve root (Figures 1A,B; 2A,B). The patient was referred to our neurosurgical outpatient department. He was further investigated with CT scan of lumbosacral spine and isotope whole body scan. The CT scan revealed osteolytic activity

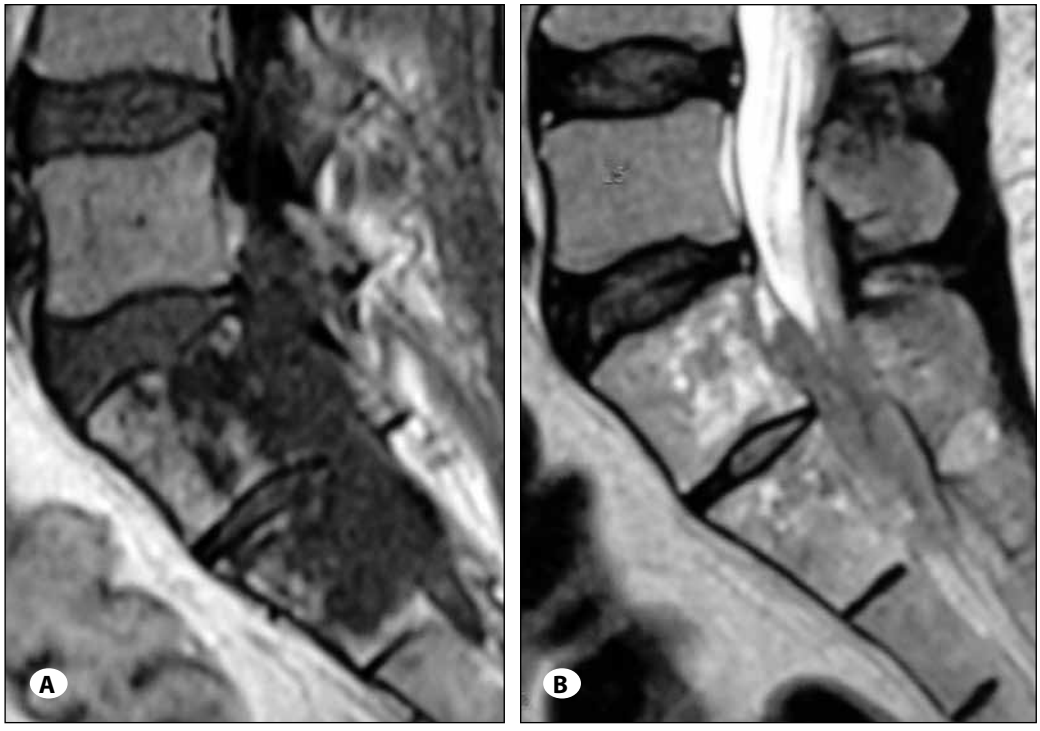

Figure 1: MRI scan of lumbosacral spine **A)** T1-weighted sagittal sections reveal isointense signal changes in S1 and S2 bodies **B)** T2- weighted sagittal sections reveal hyperintense signal changes in S1 and S2 bodies, also noted above is the isointense soft tissue lesion extending into the sacral epidural space.

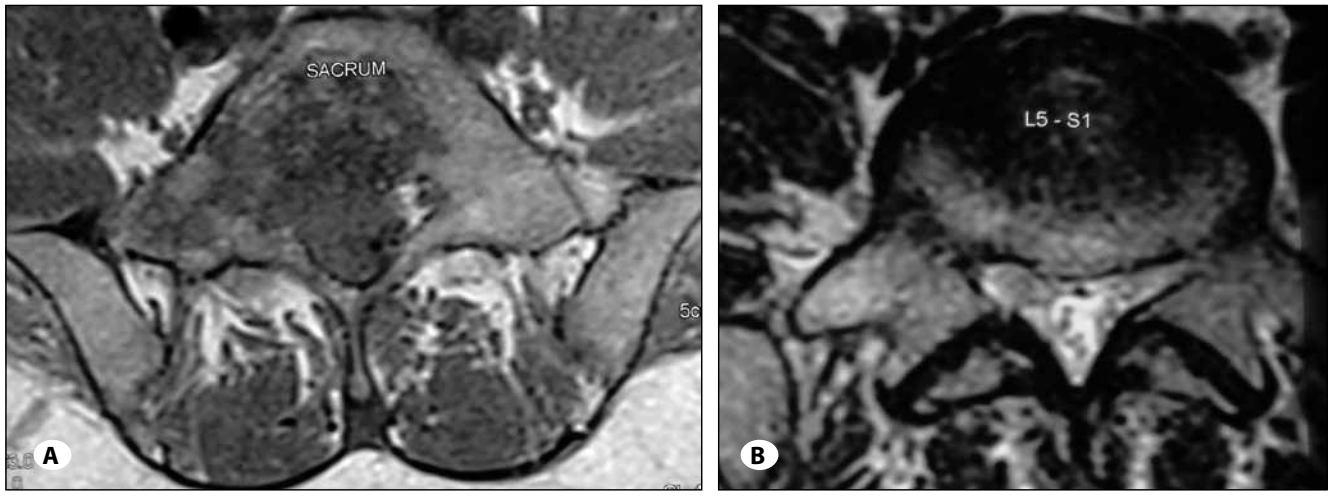

Figure 2: MRI scan of lumbosacral spine **A)** T1-weighted axial sections reveal isointense signal changes in sacrum with breach in the posterior cortical margin of the bone with soft tissue extension causing root compromise on right side **B)** T2- weighted axial sections reveal hyperintense signal changes in sacrum.

involving the right upper half of sacrum (Figure 3). Isotope scan of the whole body with technetium-99^{m} methylene diphosphate (99^{m}TC MDP) showed increased uptake in the right half of the upper sacrum and ala. There was no abnormal uptake of the isotope anywhere else in the body.

Operation

He was operated and S1S2 laminectomy was done. At surgery, the tumor was found in sacral canal and was infiltrating both S1 roots, more on the right side. The tumor was soft, suckable and moderately vascular. The tumor was extending from S1 to S2 level. Decompression of the tumor was done and intraoperative crush biopsy was reported as lymphoma.

Post-operative course

Patient had an uneventful postoperative course. His radicular pain subsided. His sensory deficit improved. However, the planter flexion on right side persisted to be 4/5.

Histopathology

On hematoxylin-eosin staining, the tumor was comprised of sheets of large cells of lymphoid lineage. There were areas

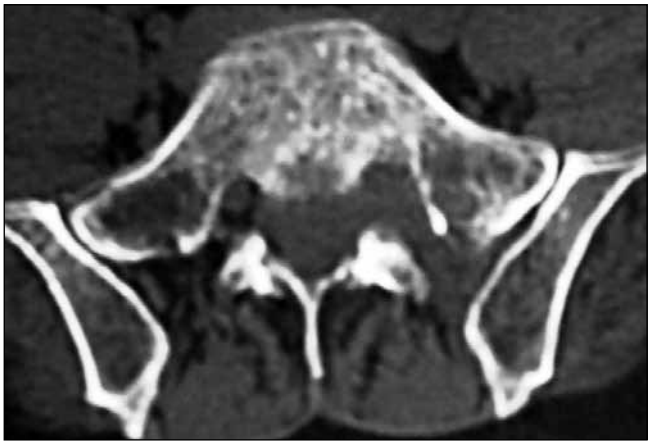

Figure 3: CT scan of lumbosacral spine axial cuts show moth eaten appearance of sacrum.

of hemorrhage and necrosis. Tumor cells had brisk mitotic activity with many apoptotic bodies. Immunohistochemistry showed the tumor cells to be strongly positive for CD-45, CD-20 and CD-19 and negative for CD-3,CD-5, CD-99,Alk and cytokeratin (Figure 4A-C).

Screen for secondary lymphoma

His blood counts and bone marrow were normal. CT chest and abdomen was normal. There were no enlarged nodes or viscera.

Follow-up

Patient is now on 6 months follow up. He has received radiotherapy to the lumbo-sacral spine and is doing well.

DISCUSSION

The commonest malignancy of sacrum is metastasis and commonest primary sacral tumor is chordoma (7). Primary lymphomas of the bone form less than 5% of malignant bone tumors (1). In the spine, usually the lumbar or lower dorsal spine is involved by lymphoma (5). Skeletal involvement by lymphoma is more common in males than females (9).

The usual age of presentation is 5th to 6th decade of life (13), though some series report a higher median age of 70 years (9). The clinical features of spinal lymphomas have been divided into two phases viz. a prodromal phase, in which local pain is common and a second phase characterized by features of compression of cord or cauda equina (5,9). Our patient presented with low back pain and right S1 radiculopathy that was not relieved by analgesics.

On MRI, both high and low signal marrow abnormalities on T-2 weighted images are seen which are consistent with osteolytic and osteoblastic changes respectively. The margins are poorly defined presenting a wide zone of transition (6). Some authors refer to this peculiar type of bone involvement as moth eaten appearance (3). Mascalchi et al after reviewing MRI images of 8 patients of spinal lymphomas concluded that demonstration of a homogenous isointense lesion

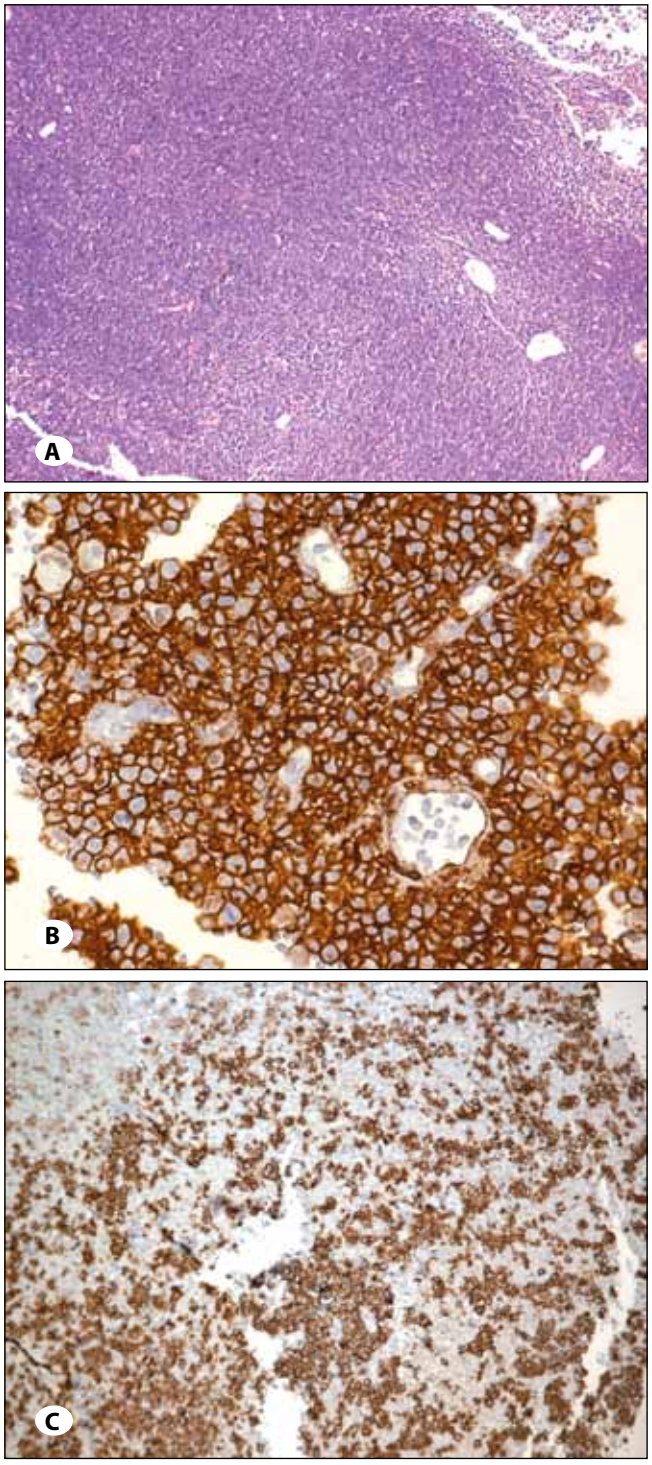

Figure 4: Photomicrograph **A)** showing monomorphic lymphoid cells with intervening blood vessels, **B)** tumor cells showing strong LCA (CD-45) positivity, **C)** tumor cells showing CD-20 positivity.

which extends over more than one segment of the spine, which may have a paraspinal extension and is accompanied by diffuse vertebral marrow signal changes, should raise the suspicion of a primary or a secondary spinal lymphoma.

They also noticed the mean longitudinal extension was 2.6 vertebral segments with a range of 1-4 segments. Our patient had a 2 segment sacral longitudinal extension of the tumor. They also found in 5 of their patients extension of the lesion through intervertebral foramina into the paravertebral soft tissue (10). Such a finding was not observed in our patient. Isotope bone scan reveals a hot spot, suggesting increased uptake by the lesion. In our patient X-ray of the lumbosacral spine was normal and MRI showed signal changes in S1 and S2 vertebral bodies. CT scan showed classical moth eaten appearance of S1 and S2. Isotope scan showed increased activity on right half of upper sacrum and adjoining part of the sacral ala. On imaging the differential diagnosis includes primary bone tumors, metastasis, multiple myeloma. Histologically primary bone tumors have characteristic diagnostic features except for small cell osteosarcoma and Ewing's sarcoma. These were however excluded by doing immunohistochemistry for CD-99 and positivity for leukocyte common antigen (LCA). The osteoid of small cell sarcoma was not seen in our case. Metastatic tumors likely to be mistaken for lymphoma are small cell carcinoma from the lung but immunohistochemistry shall not be positive for LCA, but shall be positive for cytokeratin (8). All the metastatic lesions on MRI will be hypointense on T1-weighted and hyper on T2-weighted sequence. Sacral chordomas and chondrosarcomas have specks of calcifications. Multiple myeloma involving the bone shares the same characteristics on MRI imaging, however on isotope scan there will be no tracer uptake and a cold spot is produced (12).

Epidural involvement of cord by the soft tissue rather than vertebral body collapse is more common in spinal lymphomas. Single epidural lesions are commoner than multiple. Diffuse large-B-cell lymphoma is the most common type of non-Hodgkin's lymphoma. Diffuse large B-cell lymphomas are highly invasive. Bone marrow involvement is present in upto 20% of patients initially, its detection is important because of its strong correlation with later spread to the central nervous system (2,7). In our patient there was no bone marrow involvement.

For localized spinal disease with spinal cord/cauda equina compression, surgical decompression followed by radiotherapy is the treatment of choice. In a large series of 52 patients of primary spinal epidural lymphoma it was found that the ideal dose of local radiotherapy is 36 Gy (11). Lymphomas are very sensitive to radiation and chemotherapy. The results of treatment for localized disease are good (5,7,13).

Primary NHL localized to bone has a better progress than patients in whom bone involvement is secondary to a systemic process. The 5-year overall survival, disease-free survival, and local control reported by Monnard et al in primary spinal epidural lymphoma were 69%, 57% and 88% respectively. About 42% had local relapse. Younger age and complete neurological response after the treatment are favorable prognostic factors (11).

CONCLUSION

Primary sacral lymphoma should be considered as one of the differential diagnosis of a sacral mass in elderly patients.

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