

Primary Non-Hodgkin's lymphoma of epidural space- A case report of great mimicker and review of literature

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ABSTRACT

Primary Non-Hodgkin's lymphoma of epidural space is a rare entity. Clinical presentation and imaging studies were bizarre and posed the difficulty in diagnosis. Since the management protocol of spinal lymphoma is yet to be standardized, so the prognosis is still looming. Here we are reporting a case of epidural primary Non-Hodgkin's lymphoma in young adult female, who presented with backache and weakness.

Keywords: *Non-Hodgkin's lymphoma, Primary epidural Non-Hodgkin's lymphoma.*

The usual age of presentation of primary epidural Non-Hodgkin's lymphoma (PENHL) is 5th to 6th decade of life with a male preponderance [1]. Literature favors that the triad of motor weakness, back pain and bladder involvement are the most common presenting symptoms [2]. First time Welch and Ginsburg reported the spinal cord compression caused by extra-dural lymphoma [3]. Extranodal (Non-Hodgkin's) lymphoma accounts for 10-20% cases of all form of lymphomas. Vertebral affection of extra-nodal lymphoma is relatively less, and constitutes up to the 2% of all Non-Hodgkin's lymphoma and 10% of all tumors originating from epidural space [4, 5]. So, it seems noteworthy that chronic non-specific backache should be entertained cautiously, because it can be alarming for serious ailment.

Here, we want to bring it to horizon of knowledge, that how a case of PENHL mimicked clinico-radiologically to the pott's spine. In contrast to the literature of male preponderance, our patient was a female of third decade and without any other systemic involvement. She experienced the vague pain in the mid back for 3 weeks which was associated with paraparesis.

CASE REPORT

A 34 years-old female was admitted to the emergency department with chief complaints of pain in the mid of her back with weakness of both lower limbs since six weeks. Over the period of two weeks, she experienced the continuous, localized, vague pain. After that she developed the progressive weakness of both lower limbs and became unable to walk even with support. She did not give a history of fever, night cries, weight loss, and loss of appetite or bladder and bowel involvement.

She complained for the backache corresponding to D12 vertebra. The examination of spinal column was normal, except

the patient discomfort due to pain. The patient had weakness, so gait could not get assessed. Overlying skin was normal, with normal curvature. On palpation, there were no deformity, paraspinal spasm, local rise of temperature and direct or rotational tenderness. The patient had the average nutrition with hypertonia of both lower limbs and absent patellar and ankle clonus. She experienced reduced power in both lower limbs (L2 hip, L3 knee, L4 ankle, L5 great toe and S1 planter flexion - 3/5). Reflexes of lower limbs were exaggerated, with indifferent Babinsky and intact sensory system.

X-ray of the part (dorsolumabr spine) and computed tomography were not informative anymore [Fig. 1]. An MRI was done which revealed the lesion extending from D11 to L1 with hypointense signal on T1 weighted image and isointense signal on T2 weighted image with maintained disc space [Fig. 2].

After clinico-radiological work-up, the differential diagnosis of pott's spine and metastasis of the spine was installed. Other hematological (CBC, Mantoux test, ADA, sputum examination) and radiological (Chest X-ray, ultrasonographic study of breast, abdomen and pelvis) examinations were done for search of tuberculosis and primary, but nothing was suggestive. Finally the decompression and histo-pathology examination was planned.

The patient was surgically managed by laminectomy (D11 to L2) and complete excision of the lesion. Per-operatively it was compressing the dura and adherent to it. It was reddish, vascular and friable in consistency. Histopathology of the lesion revealed the neoplastic infiltrates by small sized lymphocytes (monomorphus). Lymphocytes had the regular nuclei with numerous mitotic figure and clumped chromatin, which suggested the small lymphocytic lymphoma [Fig. 3].

Immunohistochemistry was positive for the marker of LCA, Vimentin and Ki 67(40-50%). The patient showed improved

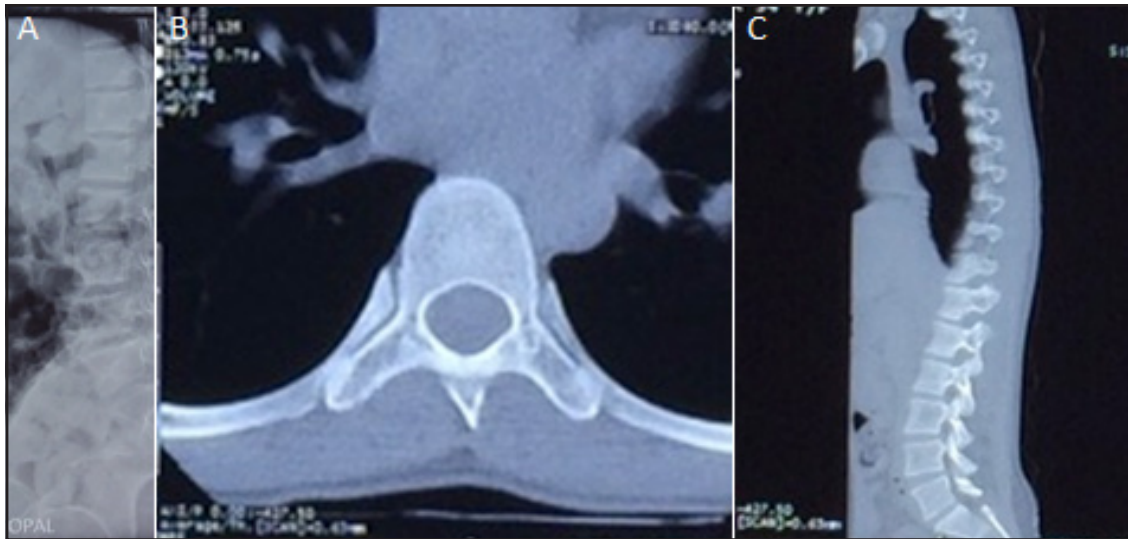


Figure 1: X-ray of the (a) dorso-lumbar spine and computed tomography in coronal (b) as well as coronal (c) section, showing almost normal findings.

comfort on low backache post-operatively. Furthermore, the patient received the chemotherapy of R-CHOP (rituximab, cyclophosphamide, doxorubicin, vincristine and prednisolone, in 6 cycles at three weeks interval) a regime and radiotherapy (4,000 cGy delivered in 25 fractions) over a duration of 4 weeks. While there was no neurological progress till the writing (follow-up of one year), but now the patient is asymptomatic and ambulatory with the help of KAFO (knee ankle foot orthosis).

DISCUSSION

Term primary lymphoma is used when the other possible sites of lymphoma in body have been ruled out. Primary epidural Non-Hodgkin's lymphoma (PENHL) most commonly originated from the epidural or intra-medullary space [6].

Tumorigenesis of PENHL is obscured. One theory conceded that the presence of lymphoid cells in epidural space may provoke antigenic stimulation, followed by transformation algorithm and lymphoma formation [7]. On the contrary, other has suggested that lymphoid tumor cells, from the vertebral and retroperitoneal area, migrate to the epidural space via intervertebral foramina [8]. Dorsal spine is the most common site of lymphoma, followed by lumbar, cervical and lumbo-sacral region [9]. Tumor corresponding to T5-T8 has the bad prognosis because of ischemic tendency due to lesser number of radicular arteries [10]. Our case has the peculiarity that it was located to the least common site of spine.

Plain X-ray of the spine is of meager importance for diagnostic purpose. In 30-42% cases only bony erosion is a diagnostic clue [11]. So, the normal X-ray and normal CT scan in

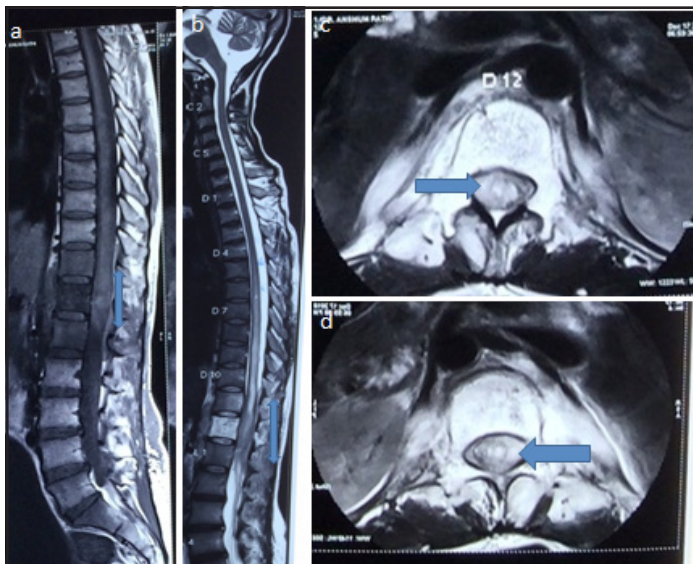


Figure 2: T1 weighted (a) MRI was hypointense and showing isointense signal (b) on T2 weighted image corresponding to D11-L1 with normal disc space. Image (c) and (d) showing the tumor infiltrations at corresponding vertebrae.

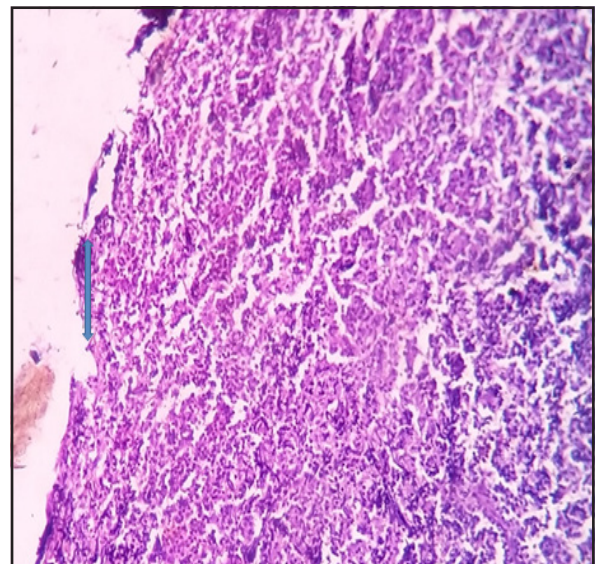


Figure 3: Haematoxylin and eosin stain showing (double headed arrow) small sized lymphocytes with regular nuclei with numerous mitotic figure and clumped chromatin

such case, often gives a significant diagnostic clinch. Henceforth Botterell, *et al.* suggested that extradural compression of the cord without conventional radiological abnormalities, strongly favor for the lymphoma. This typical finding was also corresponded to our case which had a typical clueless normal X-ray and CT scan [12].

Lymphoma has the distinguishing features in MRI. Usually it gives hypointense signal than fat on T1 weighted images and isointense with fat on T2 weighted images. Multiple myeloma, carcinoma and sarcoma are the close differential diagnosis of lymphoma, but they are hyperintense on T2 WI and appear unenhanced lesion on contrast enhanced T1 WI. On the contrary epidural hematoma shows the higher signals in both T1 and T2 WI. Epidural abscess is also a close differential diagnosis of lymphoma, and should be suspected in diabetics. But in such cases the lesion is fusiform in shape, centered over or in contiguity with the diseased disc and adjacent to the vertebral body [11].

Fixed treatment protocol of PNHL does not have the robust evidence and it includes the surgical decompression, chemotherapy and radiotherapy. Eeles *et al.* [13] advocated that surgical intervention (helps by decompressing the lesion and provide material for pathological diagnosis) should be followed by chemotherapy and radiotherapy (for localized presentation) for subclinical metastasis. Radiotherapy should be given after chemotherapy as it causes less normal tissue damage (if given before chemotherapy). If done otherwise, it may decrease the bone marrow reserve and ultimately limiting the total dose of chemotherapy. They also enumerated that whatever functional gain (motor and sphincter control) was obtained it was due to surgical intervention. There were no further improvements noticed after chemo or radiotherapy.

Though the radiotherapy but very rarely chemotherapy, are used alone the results were sub-optimal [13, 14]. Moreover, Perry *et al.* [3] noticed that if either of the modality was used alone then there was discordantly recurrence and mortality [1]. In our case, we scheduled the all three modalities. Prognosis of PENHL is nasty and its survival rate is dissimilar in different studies. Average patient survival rate varies between six to nine months [15]. Flanagan *et al.* found that 36% cases survived for 2 years in his series [16]. Immunocompromised patients had worst outcome while the old age, paraplegia, higher LDH level and sphincter involvement are associated with bad results [17].

CONCLUSION

Primary epidural Non-Hodgkin's lymphoma is a rarer presentation of the NHL. Orthopaedician needs awareness for clinical presentation of this entity to include it as a differential diagnosis in sub-acute presentation of backache with neurological involvement. Surgical decompression followed by chemotherapy

and radiotherapy should be executed in all cases for better recovery and improved survival.

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