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Primary bone diffuse B cell lymphoma of the thoracic spine: a rare entity

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ABSTRACT

Primary bone lymphoma is a rare entity, accounting for approximately 3-7% of malignant bone tumors, 5% of extra-nodal lymphomas and < 1% of all non-Hodgkin lymphomas. Primary spine localized lymphoma is 1.7% of all primary bone lymphomas. A 73 year-old female presented with a two month history of severe backache. T1W and T2W MRI showed a hypointense soft tissue component extending from the left posterior elements to Th7 corpus. The patient underwent Th6-7-8 total laminectomy. Th7-8 extradural mass was totally resected. Excisional biopsy was performed through Th7 left pedicle and vertebroplasty was applied. Significant reduction in pain was seen postoperatively. The patient was diagnosed with diffuse large B cell lymphoma. The patient received combined treatment with R-CHOP chemotherapy and local radiotherapy. Any other lesions were not detected after the examinations following 6 months period and it was accepted as primary bone lymphoma.

Keywords: Primary bone lymphoma, diffuse large B cell lymphoma, thoracic spine, excisional biopsy, vertebroplasty

Primary bone lymphoma (PBL) involving the spine is an extremely rare tumor and therefore may be missed in diagnosis. It could cause pain and neurological symptoms, and affect the quality of life negatively, especially in elderly and comorbidized patients [1]. Following a well-planned surgery, it is possible to control the tumor with chemotherapy and radiotherapy applications [1, 2].

CASE PRESENTATION

A 73 year old woman presented with a two month history of severe backache started without any trauma. She had primary hypertension, hypercolesterolemy, type 2 diabetes and chronic obstructive pulmonary disease. Localized tenderness and percussion pain were observed at Th6-8 levels. T1W and T2W MRI showed a hypointense soft tissue component extending from the left posterior elements to Th7 corpus (Fig. 1). The patient was considered primarily plasmacytoma or metastasis in differential diagnosis. No other focus or suspicious lymph nodes were observed in thorax CT, abdominal CT, dynamic abdominal MR, whole body bone sintigraphy, and all spinal MRI.

The patient was operated under general anesthesia. Th6-7-8 total laminectomy performed. The extradural lesion at Th7-8 totally excised. The tumor extending into the corpus was curetted through the T7 left pedicle, and vertebroplasty was applied (Fig. 2). As a result of histopathological and immunohistochemical analysis, lymphoid cells CD 20 (+), Bcl-2 (+), CD10 (+),

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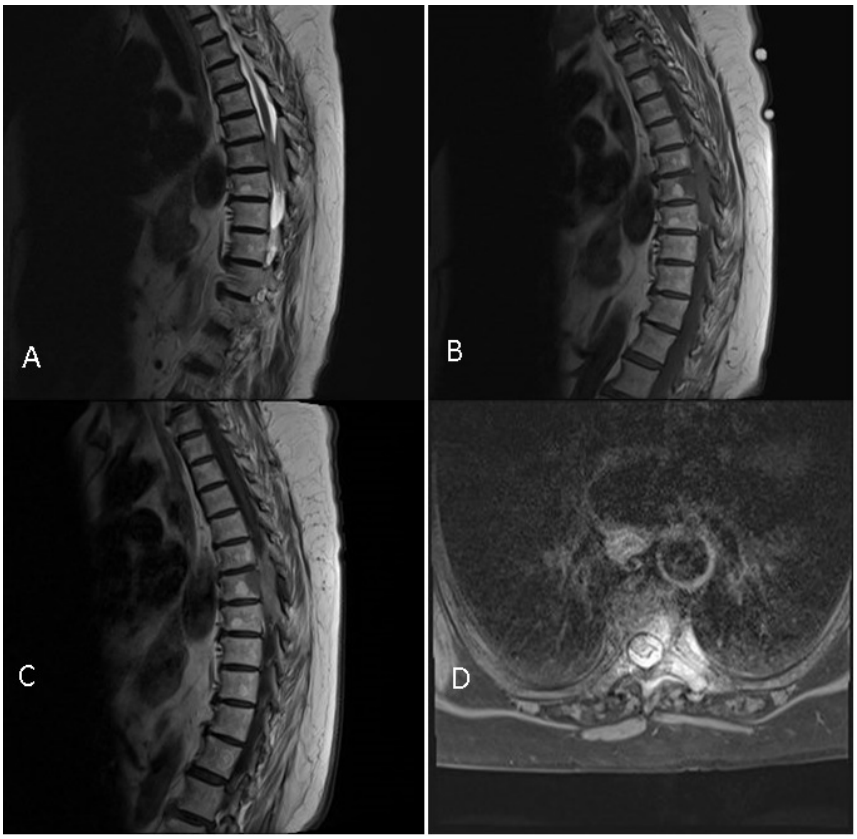

Fig. 1. **(A) Preoperative sagittal T1W, (B) sagittal T2W, (C) post-contrast sagittal and (D) post-contrast axial MR images of the patient showing a soft tissue component with T1W and T2W hypointense signal characteristics, and extending from the left posterior elements and Th6-7 and Th7-8 foramens, posterior to the Th7 vertebral corpus.**

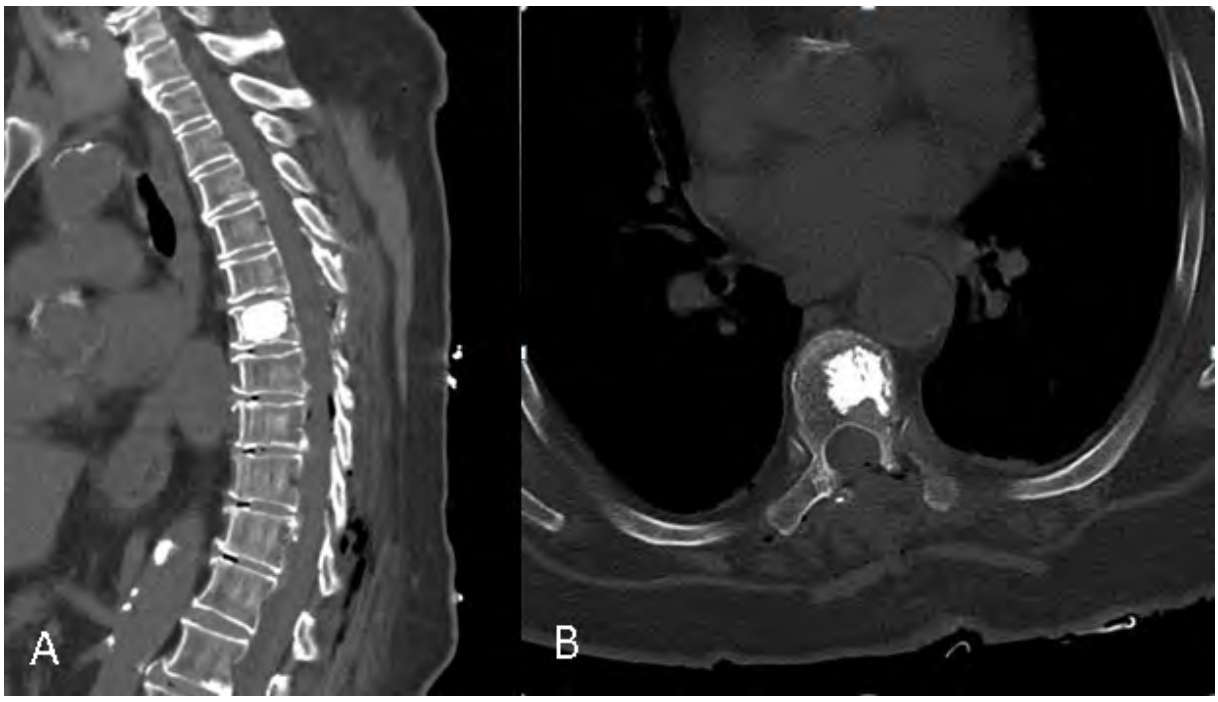

Fig. 2. **(A) sagittal and (B) axial early postoperative control thoracal CT showing the mass was totally excised and the polymethylmethacrylate (PMMA) applied to the 7. thoracic vertebra.**

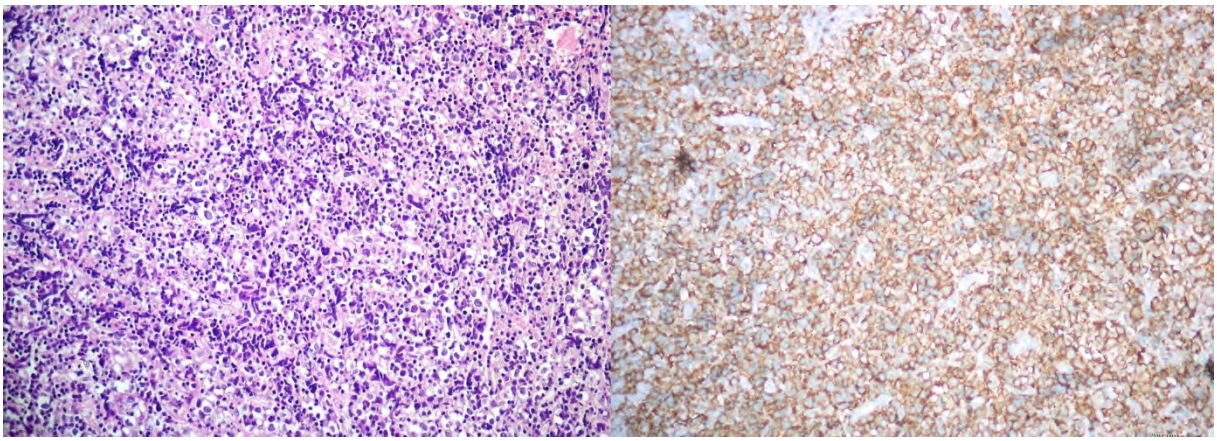

Fig. 3. Histopathology: (A) Diffuse infiltration of large atypical lymphoid cells (HE ×200) and (B) diffuse CD20 positivity in atypical lymphoid cells (CD20 ×200).

PAX-5 (+), Bcl-6 30% (+), Mum-1 (-). Ki67 proliferation index detected around 70%. It was thought "Germinal Center Originated" B cell phenotype with Hans algorithm. It was accepted as diffuse large B-cell lymphoma (Fig. 3). The patient received combined treatment with four cycles of R-CHOP chemotherapy (rituximab, cyclophosphamide, doxorubicin, vincristine, prednisone), and radiotherapy in a dose of 46 Gy in 15 sessions.

The patient was diagnosed with primary bone lymphoma, with no local or systemic extra involvement at the post-op control within six months. The patient has no complaints or neurological deficits 15 months postoperatively. No local recurrence was observed in control spinal MRI (Fig. 4).

DISCUSSION

Primary bone lymphoma is a rare entity; and constitutes 3-7% of all malignant bone tumors, less than 1% of all non-hodgkin lymphomas and 5% of all extranodal lymphomas [3,4]. Primary bone lymphomas are mostly reported in the femur or pelvis (50%) and upper extremities long bones (20%). The remaining 30% cases include rare localizations such as costa,

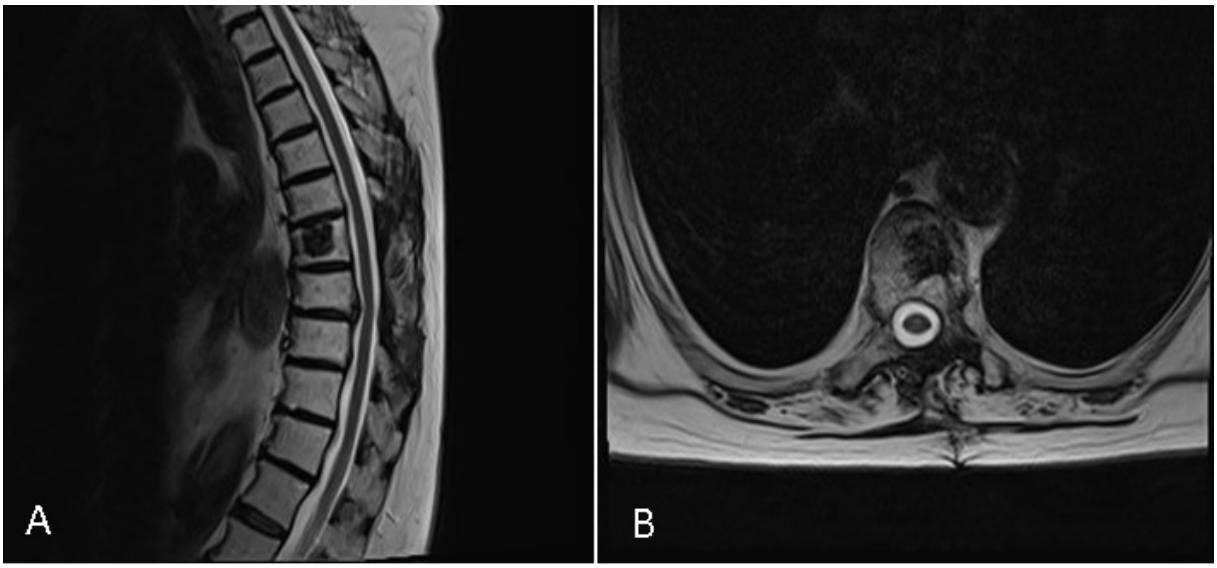

Fig. 4. (A) T2W sagittal and (B) T2W axial control MRI sections at the postoperative 15th month, showing the mass was completely excised and no local recurrence was seen.

mandible or scapula. 1.7% of all primary bone lymphomas involve the spine, mostly thoracic or lumbar spine [5]. World Health Organization classification of neoplasms of the hematopoietic and lymphoid tissues in 2022 characterized PBL as an independent disease [2].

Following four criteria should be met in the detection of primary bone lymphoma today: (1) primary involvement site of the bone; (2) no evidence of extra-bone lesion; (3) no evidence of any other extra-bone lesion six months after bone lesion is diagnosed; and (4) the diagnosis is confirmed by both pathological morphology and immunochemistry [2].

The most common histological type (90%) of primary bone lymphomas is diffuse large B-cell lymphoma [4]. It is moderately higher in men, most common age is 45-60, it peaks around the age of 50 [6]. Pain is the most common symptom (80-95%). Pathological fractures (15-20%), hypoesthesia, paraplegia, bladder and anal sphincter dysfunction could be determined [7].

MRI provides clear information about soft tissue spread, and spinal cord compression. PET CT provides insight into disease spread and involvement sites [8]. Primary bone lymphoma as a rare entity should be kept in mind in the differential diagnosis of metastatic carcinoma, Ewing sarcoma, osteosarcoma, plasmacytoma and chronic osteomyelitis [9]. The definitive diagnosis of primary bone lymphoma is based on histopathological and immunohistochemical examinations, such as MUM1 positivity and BLC6 mutation compared to other extranodal lymphomas. MUM1 expression > 10%, low CD10 expression and non-germinal center origin are signs of poor prognosis. Patients initially presented with a pathological fractures were also associated with a poor prognosis [10].

Emergency surgery is indicated for spinal cord compression presenting with neurological deficits. Stabilization prevents instability and increases the quality of life, especially in multiple level pathological fractures. Percutaneous vertebroplasty preferred as a minimally invasive procedure for pain palliation in patients with only corpus involvement [1, 10]. With systemic chemotherapy (R-CHOP protocol) and local radiotherapy following surgical resection, five year survival range is from 62% to 88% [3, 4, 11].

CONCLUSION

Primary bone lymphoma involving the spine is an extremely rare entity. Histopathological and immunohistochemical examination is important in the differential diagnosis of diseases such as metastasis, ostomyelitis, plasmacytoma, and osteosarcoma.. Treatment options include percutaneous vertebroplasty, resection with open surgery, and resection + stabilization following systemic chemotherapy and local radiotherapy.

Authors' Contribution

Study Conception: EA, HBB, HY, AT, AY; Study Design: EA, HBB, HY, AT, AY; Supervision: EA, HBB, HY, AT, AY; Funding: EA, HBB, HY, AT, AY; Materials: EA, HBB, HY, AT, AY; Data Collection and/or Processing: EA, HBB, HY, AT, AY; Statistical Analysis and/or Data Interpretation: EA, HBB, HY, AT, AY; Literature Review: EA, HBB, HY, AT, AY; Manuscript Preparation: EA, HBB, HY, AT, AY and Critical Review: EA, HBB, HY, AT, AY.

Conflict of interest

The authors disclosed no conflict of interest during the preparation or publication of this manuscript.

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