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SNI: Spine

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Case Report

Primary anaplastic lymphoma kinase-negative anaplastic large cell lymphoma of cervical spine presenting with quadriplegia: A case report and literature review

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ABSTRACT

Background: An anaplastic large cell lymphoma (ALCL) involving the cervical spine and leading to quadriplegia is very rare.

Case Description: A 48-year-old immunocompetent male presented with quadriplegia that warranted an anterior cervical corpectomy/fusion. He was previously being presumptively treated for cervical disease attributed to tuberculosis. The histopathology and immunohistochemistry revealed an ALCL that was anaplastic lymphoma kinase (ALK) negative. The patient had a favorable response to surgery followed by CHOEP (cyclophosphamide, doxorubicin, vincristine, etoposide, and prednisolone) chemotherapy.

Conclusion: ALK-negative ALCL presenting with quadriplegia due to primary involvement of cervical spine is extremely rare, but must be diagnosed and appropriately managed.

Keywords: Anaplastic large cell lymphoma, Anaplastic lymphoma kinase negative, Cervical spine, Quadriplegia

INTRODUCTION

Primary lymphoma of the spine is very rare.[7] An anaplastic large cell lymphoma (ALCL) is even less frequently encountered. Here, we report a case of anaplastic lymphoma kinase (ALK)-negative large cell lymphoma (ALCL) involving the cervical spine.

CASE REPORT

A 48-year-old male patient presented with 1 year's duration of progressive right-sided neck pain, a right C6 radiculopathy, and eventually a right > left spastic quadriparesis. The MRI revealed a destructive lesion involving the C3-C5 vertebra with an epidural collection. The original diagnosis was Potts disease (radiological diagnosis alone), and anti-tubercular drugs were utilized for 3 months. He finally developed a spastic quadriplegia with bladder involvement, leading to a

contrast enhanced cervical MRI (CEMRI) and holospinal – CT. When these studies confirmed a destructive C3-5 lesion with an epidural collection, unchanged from prior study, the patient underwent an anterior C3, C4 (complete), and C5 (partial) corpectomy/fusion using an expandable cage [Figure 1a-d].

Pathology

The histopathology showed diffuse sheets of large atypical oval to polygonal cells admixed with lymphocytes, eosinophils, neutrophils, plasma cells, and histiocytes. The tumor cells showed oval to horseshoe shaped nuclei, coarse chromatin, prominent nucleoli, and moderate amount of pale eosinophilic cytoplasm.

Immunohistochemistry

Immunohistochemistry showed tumor cells to be positive for CD3, CD4, CD7, CD8, CD30, and CD43 and negative for ALK-1, CD15, CD68, CD20, CD79a, CK, CD1A, and S-100; Ki-67 index of tumor cells was 20% [Figures 2 and 3].

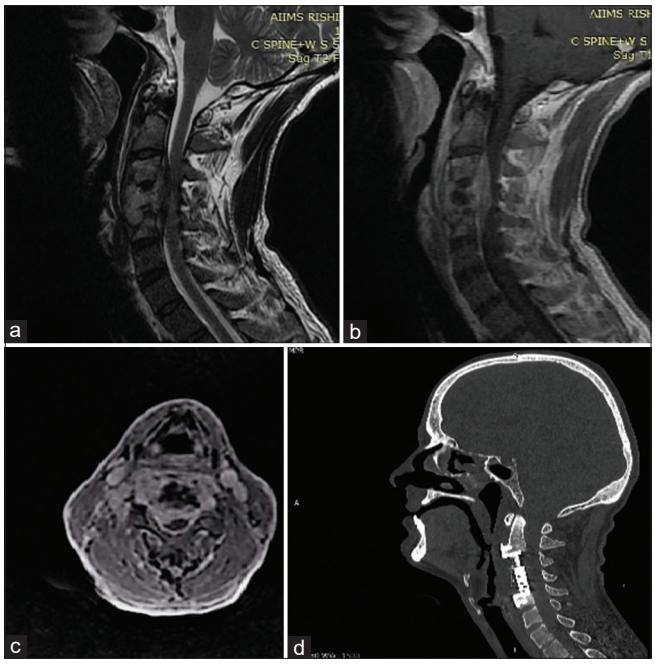

Figure 1: (a) T2 sagittal MRI cervical spine showing complete destruction C4 with partial destruction of C3 and C5 body with anterior epidural collection with compression of cervical spinal cord between C3 and C5. (b) T1 gadolinium contrast-enhanced sagittal MRI cervical spine showing contrast-enhanced epidural lesion anterior to C3-C5. (c) Axial contrast-enhanced cervical MRI at C4 showing canal compromise. (d) Postoperative NCCT cervical spine shows adequate decompression between C3 and C5 with expandable titanium cage *in situ*.

Staging of tumor

For staging, a CECT scan of the neck, thorax, abdomen, and pelvis, with bone marrow biopsy, CSF study with cytospin, and 99mTc-MDP bone scan were performed. The bone scan showed focal involvement at the operative site but there was no evidence of marrow involvement with lymphoma. The final diagnosis was primary ALK-negative ALCL of cervical spine.

Postoperative course

In the postoperative period, he was treated with CHOEP (cyclophosphamide, doxorubicin, vincristine, etoposide, and

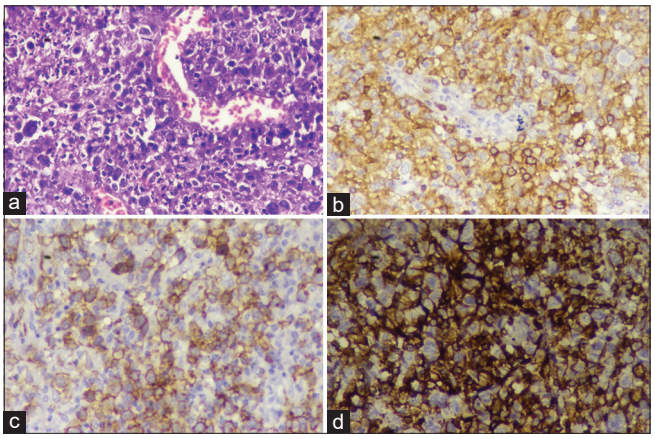

Figure 2: Photomicrographs showing (a) large atypical cells scattered around and at places focally aggregated; the atypical cells have abundant cytoplasm, wreath-like nuclei, embryo-shaped nuclei with open chromatin and multiple nucleoli; background shows mixed lymphoid population and eosinophils (H&E, ×400). Immunohistochemistry showing (b and c) diffuse, strong positivity for CD4 and CD30, respectively, in the atypical cells, and (d) diffuse positivity of LCA in large atypical cells.

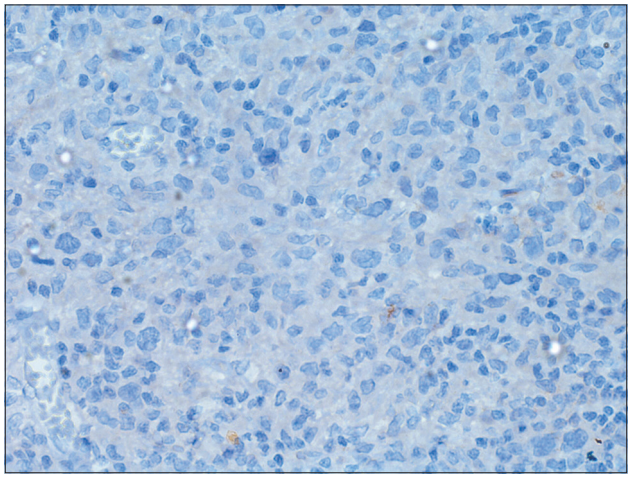

Figure 3: Negative anaplastic lymphoma kinase-1 staining in the atypical cells (Immunoperoxidase, ×400).

prednisolone) chemotherapy. He has received three cycles of CHOEP chemotherapy. A total of 6 cycles of CHOEP chemotherapy are planned. Postoperatively, he exhibited only mild residual spasticity.

DISCUSSION

Anaplastic large T-cell lymphoma (ALCL) is a CD30-expressing subtype of peripheral T-cell lymphomas; it accounts for less than 5% of all non-Hodgkin lymphomas.[7] ALCL has three distinct subtypes: ALK-positive systemic ALCL, ALK-negative systemic ALCL, and primary cutaneous ALCL [Table 1]. The majority of ALCL patients present with an advanced disease frequently associated with disseminated nodal, extra nodal, and prominent systemic symptoms. ALK-negative ALCL is generally associated with a significantly worse clinical prognosis versus ALK-positive ALCL.

Chemotherapy

Six cycles of multiagent chemotherapy, with or without involved site radiation therapy, are preferred for the treatment of ALK-negative ALCL. The first-line chemotherapy regimen consists of brentuximab vedotin + CHP (cyclophosphamide, doxorubicin, and prednisolone) [Table 2]. Another recommended regimen includes CHOEP. With these regimens, a trend toward an improved event-free survival has been noted in patients under 60 years of age with low-risk IPI scores (lymphoma international prognostic index) (IPI <1).[7]

Table 1: Types of ALCL.

1	ALK-positive systemic ALCL
2	ALK-negative systemic ALCL
3	Primary cutaneous ALCL

Table 2: Recommended chemotherapy regimens for ALCL.

1. Brentuximab vedotin + CHP (cyclophosphamide, doxorubicin and prednisolone)
2. CHOEP (cyclophosphamide, doxorubicin etoposide, vincristine, and prednisolone)
3. CHOP (cyclophosphamide, doxorubicin, vincristine, and prednisolone)
4. EPOCH (etoposide, prednisolone, vincristine, cyclophosphamide, and doxorubicin)

Vertebral involvement

Primary vertebral involvement by ALCL is very rare[1-3].Only a few cases of ALCL affecting spine have been reported [Table 3]. CT scan, MRI, and PET-CT's aid in diagnosis and staging.[4,5] The gold standard for confirmation is biopsy of the affected site.[6,7] Surgery is reserved for patients with unstable spinal fractures and those at risk for developing myelopathy/cord compression; the goal, therefore, is to restore/maintain function and/or alleviate pain. Postoperatively, patients should receive chemotherapy without delay. Here, our patient presented with quadriplegia due to primary bone lymphoma arising from multiple cervical vertebral bodies, which was treated with a C3-C5 anterior corpectomy and fusion, followed by pathological/immunohistochemical diagnosis of an ALK-negative ALCL; he responded favorably to CHOEP chemotherapy.

CONCLUSION

In a patient presenting with quadriplegia, an ALK-negative anaplastic large cell cervical spine lymphoma was diagnosed following an anterior corpectomy/fusion from C3-C5; this followed by appropriate chemotherapy resulted in significant neurological improvement.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Nil.

Table 3: Reported cases of ALCL affecting spine.

Author, year	Age/sex (year/male)	Spinal involvement	Other system involvement	Immune status	ALK	Treatment/response
Kumar *et al.*, 2010[4]	16	Dorsolumbar spine	Retroperitoneal lymphadenopathy, bone marrow, liver involvement	HIV reactive	Not mentioned	Surgery+CHOP chemotherapy/ unfavorable
Novello *et al.*, 2013[5]	21	Thoracic intradural, extramedullary involvement	Para-iliac lymphadenopathy	Immune competent	ALK positive	8 cycle CHOP/favorable
Smith *et al.*, 2010[6]	23	Cervical spine	Constitutional symptoms	Immune competent	ALK positive	6 cycles chemotherapy (CHOP)/favorable
Present case	48	Cervical spine	No	Immune competent	ALK negative	Surgery + Chemotherapy (CHOEP)/favorable

Conflicts of interest

There are no conflicts of interest.

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