Case Report

Non-Hodgkin Lymphoma of Cauda Equina: A Diagnostic Conundrum: Case Report

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Abstract

Primary central nervous system lymphoma (PCNSL) is uncommon with scarce cases having involvement of the spinal cord. Cauda equina is unique in its location and shows very rare involvement by diseases pathologies. When the same occur, they pose a lot of diagnostic difficulties as the location is difficult to access with overlapping radiologic abnormalities. It is an unusual location for lymphomas to occur with only few cases reported in literature. The cauda equina lymphomas may mimic other entities which occur at that site. Histopathology is the gold standard for the same. Here, we report an unusual case of cauda equina lymphoma mimicking a myxopapillary ependymoma in a 50-year-old male.

Keywords: Angiocentric, CD20, immunohistochemistry, lymphoma, MRI

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Introduction

Lymphomas are malignant lesions of the lymphoid tissue. They can arise at extranodal locations including the central nervous system. Primary CNS lymphoma (PCNSL), a subtype of extranodal lymphoma, is rare with scarce cases having involvement of the spinal cord.[1] Involvement of the cauda equina region is exceedingly uncommon with limited literature on the same.[2] Here, we report an unusual case of primary cauda equina non-Hodgkin lymphoma (NHL) mimicking as myxopapillary ependymoma after taking consent for the same.

Case Report

A 50-year-old man, presented with midback pain, progressive paresis, paresthesia of bilateral lower limbs, increased frequency and incomplete evacuation for four months. On examination, vitals were normal. An examinationof the central nervous system showed a GCS of 15. There was motor and sensory loss in bilateral lower limbs with altered bowel and bladder habits for 4 months. Power in lower and upper limbs was 0 and 5, respectively. Sensory level was absent below the level of umbilicus. Deep tendon reflexes were absent. Bilateral plantar reflexes were non-reactive. Details of the workup done in outside hospital were not available. However, there was no history of any comorbidities or any significant medical history or past history of any other surgeries. No significant family medical history was found. On contrast MRI, multifocal patchy enhancing soft tissue lesion involving lower dorsal cord, conus and cauda equina from D10 to L4 level associated with diffuse spinal cord signal changes and syrinx formation from lower border of D10 upto conus was seen [Figure 1a]. Post-operative soft tissue changes with atrophy and edema of bilateral lumbar, paraspinal, gluteal and iliopsoas muscle were seen. Hence, possibility of myxopapillary ependymoma was considered.

To assess the tumour, an exploration laminectomy at L2 and D11/D12 was done and biopsy sample was taken. Excised tumour biopsy revealed multiple bits of necrotic tissue with viable areas showing abrupt transition between glial tissue and highly cellular zones [Figure 1b and c]. The latter showed large tumour cells arranged in diffuse infiltrating pattern with angiocentricity. These cells had pleomorphic enlarged nuclei,

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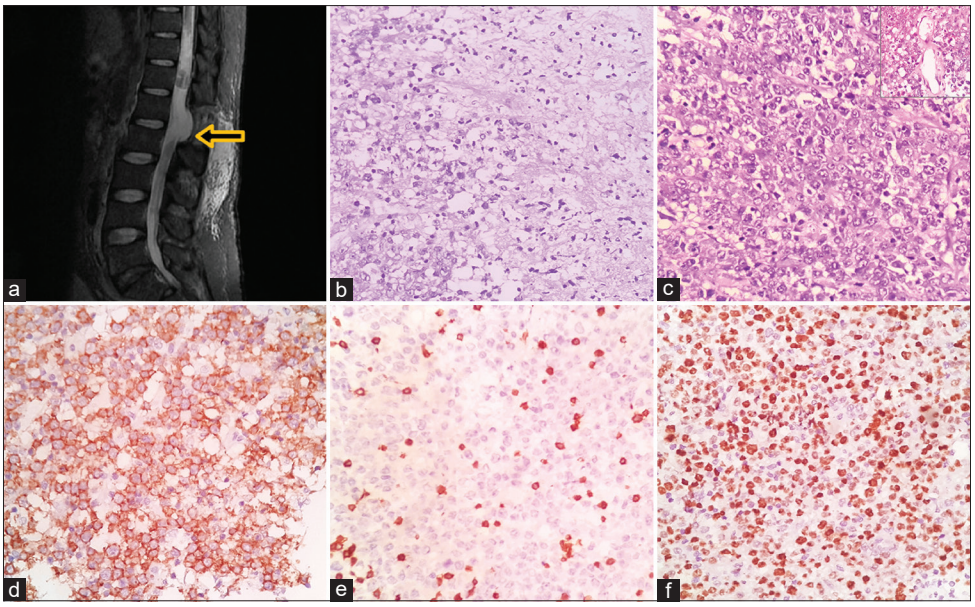

Figure 1: (a)Contrast MRI: multifocal patchy enhancing soft tissue lesion involving lower dorsal cord, conus and cauda equina from D10 to L4. (b) Section from the viable area showing abrupt transition between glial tissue and cellular tumour zones (H and E, 100×). (c) Tumour showing large pleomorphic cells arranged in diffuse infiltrating pattern with angiocentricity (inset) with enlarged nuclei and scanty cytoplasm, with prominent nucleoli. (H and E, 400×). (d–f) Immunohistochemistry showing diffuse strong membranous positivity for CD20 (d), scattered positivity for CD3(e) and high Ki-67 labelling (f) (400×)

many showing prominent nucleoli and scanty cytoplasm. Brisk mitoses were also noted. No papillary areas or areas of myxoid change suggestive of myxopapillary ependymoma were seen. Immunohistochemistry showed diffuse positivity for CD 20 in the large tumour cells, scattered CD3 positive reactive lymphoid cells and more than 80% Ki-67 staining [Figure 1d-f]. A diagnosis of high-grade NHL of Bcell type was considered. In correlation with the morphology, possibility of a diffuse large B cell lymphoma (DLBCL) was suggested. A detailed haematological evaluation was performed. Complete blood count, peripheral blood and bone marrow examination revealed no evidence of lymphomatous infiltration. Clinico-radiological workup showed no other focus of lymphadenopathy. Hence a diagnosis of primary CNS lymphoma; CELconsistent with DLBCL was confirmed. The patient was referred to oncology centre to plan for chemotherapy.

Discussion

Lymphomas can be nodal or extranodal and primary CNS lymphoma (PCNSL) is a variety of the latter.[1] Extranodal lymphomas are uncommon, and in the PCNSL, they can involve any part of the CNS ranging from the brain parenchyma, meninges, spinal cord or eye ball without any form of systemic disease.[2] Among these sites, the spinal cord shows the least common involvement site in the CNS, of which cauda equina is the lowest portion and is the rarest site. The cauda equina region is rarely affected by pathologies and may be affected by a number of lesions which may range from non-neoplastic inflammatory conditions like arachnoiditis and infective conditions like tuberculosis and cryptococcosis to overt malignancies including metastasis.[1] Only 26 cases of cauda equina lymphoma (CEL) have been documented in the English literature to the best of our knowledge.[3-6] There is limited information on the clinical presentation, imaging findings and histopathology of the same.

The exceptionally rare cases of CEL have been documented in patients aged from 11 to 79 years with slight male predominance.[4] Our case was an elderly male within documented range. The clinical features of these lymphomas are non-specific and basically stem from either infiltration or compression of the nerve roots in the vicinity.[2] In the present case also the clinical features were non-specific and pertaining to cauda equina compression and infiltration. Though, in this particular case, only a short duration of the presenting symptoms was seen, the clinical symptoms that may range from 0.5 to 36 months have been noted.[4] Gadolinium-enhanced MRI helps in diagnosis of these entities. The majority of the cases documented in literature have shown involvement uptoL1–L4 level, with increase in root volume as a consistent finding.[2] However, in the present case there was an exceptional involvement of the spine unusually reaching upto D10.

As histopathological examination and immunohistochemical confirmation has been the gold standard, the diagnostic material used for confirming these lesions in the literature varied from tumour excision/resection, thoracic laminectomy or biopsy specimens and less frequently clues have been picked up from the CSF cytology. In the majority of the cases, routine CSF examination have shown abnormality in the majority of CNSL at the time of initial presentation.[2] As this patient had

been worked up outside prior to referral, the CSF examination had already been performed there, the same was not repeated in this case. The CSF examination in the cases of cauda equina lesions provides great clues to the underlying pathology. Biochemical derangements in CSF range from elevated protein with normal to lowered glucose in the CSF. Pleocytosis with elevated leucocyte count is a frequent finding.[4] Though biopsy is essential for diagnosis, in inflammatory and metastatic disease biopsies may be obviated with the help of a preliminary CSF examination.[1]

Tumour tissue remains the best and most used modality of diagnosis in conjunction with an appropriate panel of immunohistochemical markers.[4] Here, exploration laminectomy specimen was studied. More than 90% of these tumours found in the literature are histologically and immunohistochemically NHLs of B cell type; particularly diffuse large Bcell lymphoma (DLBCL), consistently positive for Bcell markers and immunoglobulin light chains. Rare cases of lymphoblastic lymphomas, Tcell lymphomas and NK/Tcell lymphomas have also been documented.[2,4]In histology, the most important feature which helps in suspecting the diagnosis is the sheets of tumour cells, high N: Cratio, scant cytoplasm, coarse clumped chromatin and prominent nucleoli. The tumour cells show angiocentric pattern of arrangements and frequent mitosis. Also the use of immunohistochemistry for the LCA positivity and CD3, CD20 and Ki 67 staining helps in confirming the lymphoid origin in these cases and enables the definitive categorisation. The present case was found to have features compatible with DLBCL.

The major limitation experienced in this case was the lack of details of the previous hospitalisation and paucity of CSF sample for cytologic evaluation. Also other limitations include the limited tissue for IHC and lack of much literature on the present entity.Various treatment modalities have been used from only steroid therapy to multimodality therapy with or without radiotherapy.[4]Methotrexate is the drug of choice and chemoradiation is superior to any one modality alone.[1] Thirteen out of 21 patients, in literature, have survived in a follow-up period of 3 to 81 months.[4] As emphasised in this case, it is essential to do a detailed clinico-haematological workup to rule out a secondary NHL.[5]

Conclusion

The present case stands unique as it highlights the rare occurrence of cauda equina lymphoma in an elderly male with no systemic manifestations and radiologically masquerading a myxopapillary ependymoma. The classic histomorphology of large cell NHL with immunohistochemical expression of CD20 and high Ki67 labelling helped in confirming the diagnosis. A high index of suspicion, detailed clinico-radiological and haematological, histomorphological and immunohistochemical assessment with other ancillary techniques can help in timely diagnosis and optimal management.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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