Primary Anaplastic Large Cell Lymphoma of Central Nervous System

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ABSTRACT

Central nervous system (CNS) involvement is extremely rare in anaplastic large cell lymphoma (ALCL). Primary ALCL of CNS on radiology is often misdiagnosed as tuberculosis. We report a fatal case of primary ALCL of CNS in a 17 year old male. He came with history of headache and left partial seizures. MRI showed a well-circumscribed lesion in the right fronto-parietal lobe eroding the skull bone. Biopsy showed large pleomorphic cells. Immunohistochemical stains showed positivity for CD30, CD43, EMA and ALK-1. In spite of radiotherapy and steroids, patient expired. Hence a high level of suspicion is essential for early diagnosis and for instituting appropriate treatment.

KEY WORDS: Anaplastic large cell lymphoma, central nervous system

Anaplastic large cell lymphoma (ALCL), also known as Ki-1, pleomorphic or histiocytoid lymphoma was reported first by Stein et al. in 1985.[1] Primary or secondary involvement of central nervous system (CNS) is extremely rare in ALCL.[2] Only 15–20 well-documented cases of primary CNS–ALCLs are found in the western literature.[2–4] To the best of our knowledge this would be the first case in Indian literature.

A 17-year-old male presented with headache, left upper limb monoparesis and left partial seizures since 5 months. MRI-scan showed a right fronto-parietal enhancing lesion. Diagnosis of tuberculosis was made and patient was started on antituberculosis treatment, carbamezapine and steroids. There was no improvement and on repeat MRI, a well-circumscribed 3 x 4 cm mass was noted in the right fronto-parietal region eroding the skull bone and forming a scalp swelling (Figure 1). The patient was HIV-antibody negative and his haemogram was within normal limits. Intraoperatively, the mass was attached to the dura. An excisional biopsy showed large pleomorphic cells with abundant eosinophilic to amphophilic cytoplasm and prominent nucleoli. Few eosinophils were also noted in the background. Necrosis was absent. Hence a differential diagnosis of granulocytic sarcoma or lymphoma was considered (Figure 2). On further evaluation there was absence of systemic lymphadenopathy or organomegaly. Cerebrospinal fluid (CSF) examination and bone marrow examination were not performed. Myeloperoxidase and chloracetate esterase stains were negative, whereas CD30, CD43, LCA, EMA and Alk-1 were positive on immunohistochemistry (Figure 3). Hence, a final diagnosis of primary ALCL of T-cell type of CNS was made. Patient was treated with radiotherapy and steroids. He developed a left supraclavicular lymphnode within three months. Fine needle aspiration cytology showed cytological features of ALCL. He was then started on chemotherapy (cyclophosphamide, adriamycin and vincristine). Inspite of these measures, patient expired after 1 month. Autopsy was not performed.
Figure 2: Large pleomorphic cells with mildly pleomorphic nuclei, abundant amphophilic cytoplasm, prominent nucleoli, multinucleate cells resembling RS cells and cohesive growth pattern

Figure 3: Immunohistochemistry results (clockwise): (A) Negative CAE, (B) Positive Alk-1, (C) Positive CD 43, (D) Positive EMA

Discussion

Primary central nervous system lymphomas (PCNSL) are rare entities forming 2–6% of brain tumours. We made the diagnosis of PCNSL as it fulfilled the criteria for PCNSL.1
- The patient presented with neurological symptoms.
- The diagnosis of lymphoma was made by biopsy.
- At the time of initial evaluation, no evidence of lymphoma was found at any other site.

Primary central nervous system ALCL is extremely rare and most of the cases have been reported in young adults. Analysis of cases of primary ALCL described in the literature, indicate a bimodal age distribution with 50% below 20 years and other peak is observed after fourth decade.2-4 It is found to be more frequent in females than in males. No association with HIV is found and most of them are immunocompetent individuals. Though most cases showed multifocal lesions, there was a predilection for frontoparietal region and almost all had lepto meningeal involvement. Around half of the cases were Alk-1 positive. Few cases out of these had been clinically misdiagnosed as infectious lesions, mostly mycobacterial infection and treated accordingly, as it was in our case.2,3

Primary central nervous system ALCL seems to behave more aggressively than nodal ALCL. Multifocality, tumor necrosis, monomorphic appearance of the cells and Alk-1 negativity were associated with bad prognosis. Most of the reported cases relapsed in 2–3 months and died due to tumour or treatment related causes. Our case, despite of having none of these features behaved aggressively and relapsed in the form of nodal involvement, which is not mentioned clearly in other cases.2–4

A differential diagnosis of blastic type of granulocytic sarcoma was considered due to site, age and horse-shoe shaped nuclei. However, a clue to the diagnosis would be the presence of eosinophillic precursors and presence of 2 to 3 nucleoli, which were not seen in our case.

To summarize, PCNS–ALCL is a very rare tumour and is seldom clinically diagnosed. It is often misdiagnosed as mycobacterial CNS infection on imaging techniques. Hence an early recognition of PCNSL–ALCL by biopsy is important in permitting a more tailored therapeutic approach, as they may have a very rapidly deteriorating clinical course.

References