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Radiology

CENTRAL NERVOUS SYSTEM INVOLVEMENT IN CLASSICAL HODGKIN'S LYMPHOMA: CASE REPORT AND REVIEW OF LITERATURE

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ABSTRACT Involvement of Central Nervous System by Hodgkin's Lymphoma is an extremely rare occurrence, with the latest figures showing involvement to be present in less than 0.02% cases. Extensive workup of the patient should be done to rule out any other ongoing etiology before reaching to the diagnosis of CNS Hodgkin's Lymphoma. The disease usually occurs in the advanced stages of the disease in relapse or remission phase. We report such a case of disseminated Hodkgin's Lymphoma in a young male who showed involvement of central nervous system in the form of leptomeningeal, pachymeningeal and calvarial deposits in the relapsing phase of the disease. Imaging played an utmost important role in reaching to the final diagnosis which is well highlighted in the reported case.

KEYWORDS :

INTRODUCTION:

Central Nervous System involvement in Hodgkin's Lymphoma is extremely rare, occurring in <0.02 % cases. It is so atypical that the patient should be investigated thoroughly to rule out any other ongoing etiology. Cerebral Hodgkin's disease is invariably secondary to systemic disease. Primary cerebral Hodgkin's disease is still controversial. We report a case of a young male with disseminated Hodgkin's Lymphoma who presented with seizures and was found to have involvement of central nervous system on imaging. The role of imaging in diagnosis of such a rare entity is reiterated through the presented case.

CASE REPORT:

A thirty-six-year-old male patient presented to our institute with two episodes of generalized tonic clonic seizures. He was a known case of disseminated Classical Hodgkin's lymphoma, Stage IV, IPS score 5; diagnosed on histopathological evaluation from axillary lymph node sample (Figure 1). He underwent 6 cycles of Adriamycin, Bleomycin, Vinblastine, and Dacarbazine (ABVD) chemotherapy. Within 10 days of completion of chemotherapy, he had developed the seizure episodes. The patient did not have any history of fever, headache or weakness. He was referred for MRI Brain for evaluation of the cause of seizures.

Contrast Enhanced MRI of brain (GE Discovery 750 W 3T MRI Scanner) was done which showed T1 isointense and T2/FLAIR hypointense contents in sulcal spaces in the left frontal region. Adjacent cerebral parenchyma showed grey-white matter edema with irregularity of grey-white matter interface. Susceptibility weighted images showed hypointensity in the involved sulcal spaces with negative signal on phase images indicating hemorrhage. The sulcal contents showed a focus of restricted diffusion on Diffusion Weighted images and enhancement on Post Contrast images (Figure 2). Adjacent to the enhancing sulcal contents, another small lesion was seen along the mid sagittal dura. The lesion was hypointense on T2 weighted images and showed post contrast enhancement. Note was made of a subcentimetric lesion in left parietal bone which was hyperintense on T2 weighted sequence and also showed post contrast enhancement (Figure 3). Non contrast CT scan (Siemens Somatom Flash 128 Slice CT Scanner) of the brain was done for the patient which showed sulcal and white matter hypodensity in the left frontal lobe (Figure 4).

Subsequently, Lumbar puncture was done for the patient which showed CSF pleocytosis (40 cells/mm3) with Lymphocyte predominance (75%). Extensive workup of the patient showed no evidence of infection or any other malignancy.

Considering the diagnosis of disseminated Classical Hodgkin's Lymphoma, typical leptomeningeal involvement in brain and the suggestive CSF picture, a final diagnosis of Central Nervous System involvement of Hodgkin's Lymphoma was made in the form of leptomeningeal, pachymeningeal and calvarial deposits. The patient was managed with supportive care and appropriate antiepileptics and was kept on regular follow up.

DISCUSSION:

Involvement of central nervous system by Hodgkin's Lymphoma is a rare occurrence that most commonly occurs as secondary manifestation of the systemic disease. Rarely it may present as Paraneoplastic Syndrome in Hodgkin's Lymphoma. Primary CNS Hodgkin's Lymphoma is yet a doubtful entity with less than 25 cases described in Literature.

Secondary CNS Hodgkin's Lymphoma is a rare but an established complication of Systemic Hodgkin's Lymphoma. It occurs as a late complication in advanced stages of the disease. It most commonly presents in the relapsing phase of the disease and may also occur in patients who are in remission phase.– It carries a poor prognosis and may be a cause of deterioration or mortality of the patient. Immunodeficiency, family history of Hodgkin's Lymphoma and infection with Epstein Barr Virus have been postulated as the predisposing factors.– Our patient also showed secondary involvement of CNS in the relapsing phase of disseminated Hodgkin's Lymphoma when he had completed his six cycles of chemotherapy.

The most common histological subtypes of CNS Hodgkin's Lymphoma include Mixed Cellularity and Nodular Sclerosing varieties. However, it is not definitely agreed that whether the WHO classification of systemic Hodgkin's Lymphoma can be applied to CNS Hodgkin's Lymphoma.– Our patient on the contrary had Classical Hodgkin's Lymphoma as diagnosed by the histopathological evaluation of his axillary lymph node biopsy sample.

CNS Hodgkin's Lymphoma more frequently involves the spine than brain. Various mechanisms have been postulated for cerebral involvement which include direct extension through the bony skull or dura; meningeal involvement and metastasis; and hematogenous dissemination. Leptomeningeal dissemination is one of the major postulated mechanisms according to which Hodgkin's Lymphoma shows spread to the dura, then to the subdural space followed by penetration through the arachnoid matter, to finally produce deposits in the subarachnoid space. Some authors suggest that hematogenous dissemination is the most common mode of spread of Hodgkin's Lymphoma as there have been cases of cerebral involvement without any contiguous extra-axial deposit.

The most common sites of involvement include cerebral cortex followed by leptomeninges and dura; corpus callosum and pituitary gland.– Majority of the lesions are supratentorial in location. On CT, the lesions can appear as hyperdense or isodense to the grey matter with or without associated perilesional edema. On MRI, the lesions appear hypo to isointense and show post contrast enhancement. Contrast enhancement (CE) makes the lesions more conspicuous with CEMRI allowing better delineation than CECT.

Cross-sectional imaging is required for raising the suspicion of involvement of Central nervous system by Hodgkin's Lymphoma. Multiplanar MR imaging with administration of contrast is considered

better than CT as it can accurately depict tumor deposits in epidural, subdural space as well the brain parenchyma.

The paraneoplastic syndromes affecting the CNS in Hodgkin's Lymphoma most commonly present as cerebellar degeneration. Imaging findings may be normal at presentation but follow up scans may demonstrate cerebellar atrophy. Other sites of involvement include basal ganglia, brainstem and supratentorial cerebral parenchyma. Other modes of presentation include ataxia, chorea, stiff person syndrome, acute inflammatory demyelinating polyradiculopathy and myasthenia gravis.– The disease process is usually progressive; however, cases of spontaneous resolution have been reported as well.

Certain neurological manifestations may occur as a complication of treatment of Hodgkin's Lymphoma which more commonly involve the peripheral nervous system as compared to the central nervous system. The complications occurring secondary to Radiotherapy include "Dropped Head Syndrome", "Brachial Plexopathy" and "Episodic Neurological Dysfunctions" like flashing lights, language dysfunction, segmental motor or sensory dysfunctions etc. The neurological complications occurring secondary to the standard chemotherapy regime "ABVD" include transient ischemic attacks or cerebral infarcts which may be due to cardiac emboli secondary to dilated cardiomyopathy caused by Doxorubicin or cerebral infarcts caused by Bleomycin. Vinblastine may lead to sensory peripheral neuropathy.–

The prognosis of CNS Hodgkin's Lymphoma is poor with median survival of approximately 46 months.– However, attempt should always be made towards its cure. The appropriate treatment modality for Hodgkin's Lymphoma has not been established. Variety of treatment modalities have been tried, including radiotherapy, whole brain irradiation or combination chemotherapy with variable results.– A series reported a durable response to radiation and/or chemotherapy. The relapsed disease limited to the brain or CNS involvement when the initial diagnosis of Hodgkin's Lymphoma is made is associated with better prognosis. Central Nervous System prophylaxis is not recommended in Hodgkin's Lymphoma in routine.–

CONCLUSION:

Only a handful of cases of intracranial involvement in Classical Hodgkin's lymphoma have been reported. Imaging provides important clues to reach to the diagnosis of this rare entity. However, the guarded prognosis and atypical patterns of CNS lymphoma including leptomeningeal, dural and skull metastasis require further studies to establish the characteristic diagnostic features and an appropriate management protocol.

LEGENDS

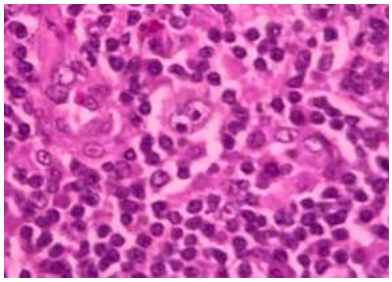

Figure 1: Histopathological image from right axillary lymph node biopsy sample of the patient shows features of Classical Hodgkin's Lymphoma with Castleman's like areas. The cells were immune-positive for CD15 and Cd30.

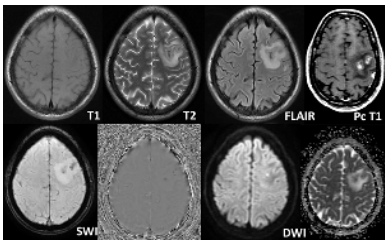

Figure 2: Axial MR images of brain of the patient showing T1 isointense and T2/FLAIR hypointense contents in sulcal spaces along the left frontal convexity. Adjacent cerebral parenchyma shows grey-white matter edema with irregularity of grey-white matter interface. Post contrast T1 weighted images show enhancement of the sulcal contents. Susceptibility weighted images show hypointensity in the involved sulcal spaces with negative signal on phase images. A focus of restricted diffusion is seen in the sulcal contents on Diffusion Weighted images.

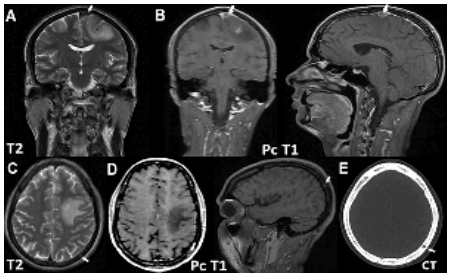

Figure 3: A. Coronal T2 weighted images of brain of the patient show a small hypointense lesion along the mid sagittal dura adjacent to the enhancing sulcal contents (arrow). B. Coronal and Sagittal Post contrast T1 weighted (Pc T1) images show enhancement in the small dural deposit. C and D. Axial T2 weighted image and corresponding axial and coronal post contrast images of brain show a subcentimetric lesion in left parietal bone which is hyperintense on T2 weighted sequence and shows post contrast enhancement (arrows). E. NCCT brain (Bone Window) of the patient makes the lesion more conspicuous.

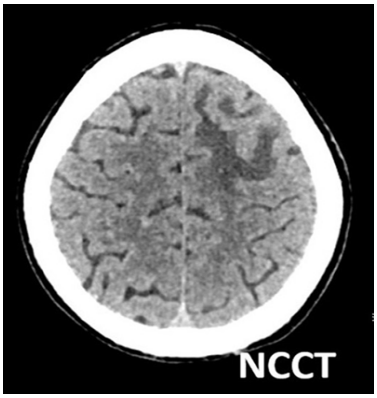

Figure 4: Axial Non contrast CT image of brain of the patient shows sulcal hypodensity in the left frontal region.

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