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Case Report

Primary Hodgkin's Disease with Involvement of the Brain in an Immunocompetent Individual - A Rare Yet Deadly Occurrence

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Abstract

Hodgkin's disease is usually a disease involving the peripheral lymphatic system with strong predilection to the cervical lymph nodes. Diagnosis is made by a combination of histopathology (where the Reed-Sternberg giant cell is pathognomonic) as well as by lymphocytic markers such as CD20 and CD30. Hodgkin's disease rarely involved the brain and non-lymphoid organs. We present a known case of Hodgkin's disease with a lesion in the frontal lobe which presented as new onset seizures in a 29-year-old patient. The lesion was successfully removed and on analysis was seen to be a part of Hodgkin's disease. The rarity of CNS involvement coupled with a recurrent pattern of this disease (the CNS involvement seen in the recurrence rather than the primary presentation) makes this a rare and interesting report.

Keywords: Hodgkin's Disease; Brain; Immunocompetent; Deadly Occurrence

Introduction

Hodgkin's lymphoma (HL) most commonly presents with progressive, painless enlargement of peripheral lymph nodes, especially around the cervical region or less frequently mediastinal or abdominal lymph nodes and progresses to other groups of lymph nodes and eventually to non-lymphoid organs [1]. HL involves the central nervous system (CNS) in 0.2-0.5% of cases either by contiguous or hematogenous spread. CNS involvement is more common in patients with widespread relapsed disease or at initial diagnosis in immunocompromised patients but can also be seen at initial diagnosis in immunocompetent patients [1,2]. We present a report of a patient with diagnosed Hodgkin's lymphoma with a lesion in the right frontal lobe of the brain which once resected was shown to be a manifestation of the same disease [1-3]. A 10-year review of literature is presented to highlight the rarity of the disease and present treatment algorhythms to best manage this rare presentation.

Case Report

The patient a 29-year-old male was diagnosed to have Hodgkin's disease in 2019 when he developed painless progressive lymphadenopathy in the deep cervical lymph nodes. A biopsy of the lymph nodes showed a greyish yellow infiltrative mass which consisted of a pleomorphic population of lymphocytes, plasma cells and histocytes along with a few giant mononucleate cells resembling the Reed Sternberg cell. IHC analysis was positive for CD20 and CD30 as well as positive for pay5 with a high MIB index. A diagnosis of Hodgkin's disease was thus made and chemotherapy was started. He completed the course in early 2020 and was asymptomatic since.

The current series of events began in august 2020 with 2 episodes of focal seizures localized to the right upper limb which was associated with frothing from the mouth, up rolling of eyes, tongue bite, clenching of the mandible with forced expiration through the

mouth producing a hissing sound. The ictus lasted for approximately 10 seconds. There was no premonition or aura before the start of the seizure and after the ictus there was confusion lasting for an hour. The patient was rushed to hospital and started with anticonvulsants and gradually regained consciousness after 1 hour. He was then investigated with a contrast MRI of the brain. His scan showed a 3.8 x 3.5 x 2.7cm sized intra-axial, ill defined, haemorrhagic mass lesion in the left frontal lobe. The lesion was hypointense on T1 sequences and heterogeneously hyperintense on T2 and FLAIR sequences. There was significant perilesional oedema in the left frontal region extending onto the rostrum of the corpus callosum causing impending subfalcine herniation and a midline shift of 6mm to the right (Figure 1). Along with this lesion, an MRI of the spine done to rule out metastatic spread. This showed tumor infiltration of the pedicles and vertebral bodies of L1 vertebra and the left L5 pedicle, along with the sacrum causing expansion of the bone compressing the neural foraminae as well as extending onto the right Sacro-iliac joint. There were no symptoms however, which pertained to the lumbar vertebral lesions.

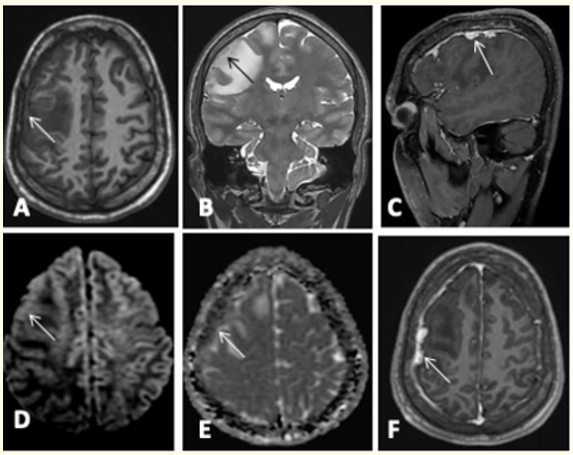

Figure 1: Showing a multi focal, discontinuous, nodular pachymeningeal thickening which is no intense on T1 and hypo intense on T2 W image (arrows in A and B) with perifocal vasogenic edema is noted along the right frontal convexity. Corresponding areas shows restricted diffusion on DW and ADC images (D and E). Nodular pachymeningeal thickening demonstrate enhancement with contrast (C and F). There is also adjacent smooth pachymeningeal enhancement noted along the right fronto-parietal convexity.

After discussing the lesion with relatives and obtaining consent the patient was subjected to surgical removal of the lesion. Surgery was proposed for 3 reasons. Firstly, surgery was needed to remove the offending lesion from the brain thereby reduce the perilesional oedema and thereby preventing the impending herniation of the cingulate gyrus. Secondly to obtain a histopathological and immunohistochemical analysis of the tumor tissue, thus allowing for the planning of adjuvant therapy for the patient, and finally to alleviate the cause of seizures which was the primary presenting symptom of the patient.

Using Neuronavigation guidance a small minicraniotomy was made along the site of the lesion. The mass was seen penetrating the dura and densely attached to the surrounding brain. The mass was rubbery, and vascular making it difficult to remove (Figure 2). The lesion was removed en masse and sent for analysis. Post-surgery no further episodes of seizures occurred and the patient was discharged to review in the outpatient clinic with the biopsy report to plan further management of the lesion.

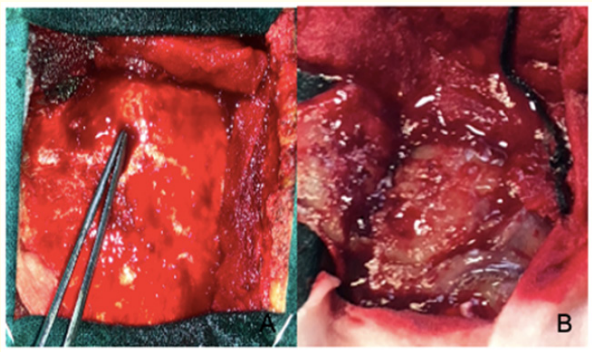

Figure 2: Showing intraoperative photographs of the lesion in question. The dural nodule is seen in (A) while the whole lesion is seen in (B) after the dura has been reflected off the brain. Note a clear plane of cleavage between the lesion and brain is not seen.

The biopsy showed, a diffuse infiltration of lymphocytes on a background of fibro collagenous stroma with some macrophages and plasma cells. Amidst these were a few large, atypical cells dark staining nucleosomes and prominent nucleoli and scant cytosol. Perivascular cuffing of the lymphocytes was also present. Immunohistochemical markers were applied to the sample. The large, atypical cells were positive for CD20, CD30, MUM1 and Bcl6 with

a dim positivity for PA X 5. The CD3 and CD 15 were negative for the large cells which showed increased mitotic activity as demonstrated by the Ki-67 index. Based on this morphology a diagnosis of large B cell lymphoma with the possibility of Hodgkin's was given (Figure 3).

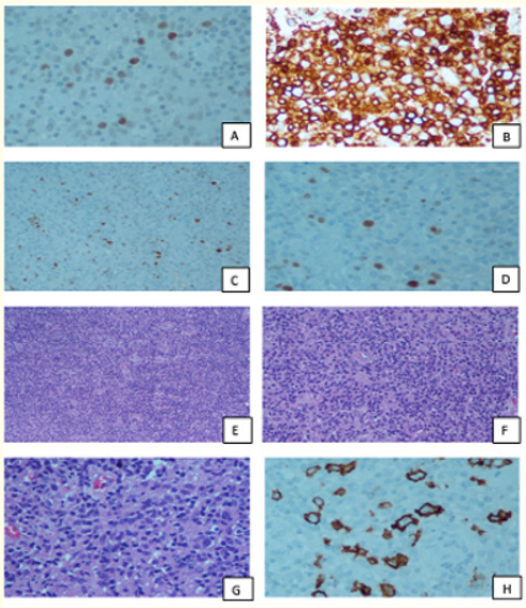

Figure 3: Showing large cells that show positivity for PAX5 (× 40X) (A), negative for CD15, CD30, CD3 and CD109 in (B), Scattered large cells show positivity for MUM1 (in 20X) in (C), and scattered large cells show positivity for MUM1 (in 20X) in (D). In the H and E sections showing a few scattered large cells (E), a few scattered atypical lymphoid cells with high endothelial venules (× 20X) (F), A few atypical lymphoid cells against a background showing mature lymphocytes and macrophages (in 40X) (G) and the large, atypical cells that show CD20 positivity (in 40X) in (H).

The patient was immediately evaluated and started on chemotherapy for the same. He endured the entire course of chemotherapy and is doing well as seen in the last follow up 3 months ago.

Discussion

Aetiology, treatment, and prognosis of intracranial HL have not been established. Common presentations include cranial nerve palsies, motor and/or sensory deficits, headaches, papilledema, coma, and seizures [1-5]. The most common presenting feature of intracranial Hodgkin's disease is cranial nerve palsy [4-7]. Brain parenchymal involvement shows a predilection for supratentorial lesions, this being the most common intracranial site of involvement. Mixed cellularity histology is the most frequent subtype of Hodgkin's disease among these patients, although the case reported by Vetter., *et al.* was of nodular lymphocyte predominance [8]. Our patient showed a mixed cellularity like picture as well.

The distinction between HL and HL-like PLD (proteinaceous lymphadenopathy) is often challenging and perhaps arbitrary [7-9]. Atypical cells in such PLD cases usually are B cells expressing CD20, CD30, CD45, and EBV antigens, the latent membrane protein (LMP-1), and/or the Epstein-Barr virus encoded RNA 1 (EBER-1) [8-10]. Median survival following local (10-20 Grey) and whole-brain (30-40 Grey) radiotherapy has been reported in the range of 8 months to 2 years.

Systemic chemotherapy in combination with radiotherapy has been advocated, and treatment protocols have included ABVD, COPP (cyclophosphamide, vincristine, procarbazine, prednisone), MOPP (nitrogen mustard, vincristine, procarbazine, prednisone), and/or intrathecal methotrexate [11,12]. Long-term disease-free survival has been reported following combined-modality therapy [13].

Conclusion

Although rare, the presentation of intracranial Hodgkins disease is possible. Proper investigation protocols coupled with modern neurosurgical technique can lead to admirable results in the short and long haul.

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