

Can Extra-Axial Hypodense Lesion be Sub-Dural Hematoma or Meningioma or Non Hodgkins Lymphoma?

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Case Report

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Abstract: A previously healthy 40 year old female admitted in NeuroSurgical department with complaints of, intermittent headache for 1 week, altered sensorium for 2 days and has 3 episodes of convulsions, loss of consciousness, right facio brachial paresis. Computed tomography scan of Brain showed hypodense extra-axial lesion of left fronto-temporo-parietal region with midline shift of 12mm and was reported as chronic Sub-dural Hematoma associated with mass affect. MRI of Brain showed an extra-axial Sub-dural Supratentorial large left fronto-temporo-parietal crescentic shaped mass with surrounding edema which is well defined and merged with adjacent normal dura. She underwent left fronto-temporo-parietal craniotomy and excision of tumor done. Histopathological examination and immunohistochemistry showed tumor tissue suggestive of Non Hodgkin's lymphoma. Post operatively, patient was referred to Regional Institute of Oncology for the needful Radiotherapy and Chemotherapy.

Keywords: Chronic Sub-dural Hematoma, Enplaque Meningioma, Primary CNS Lymphomas(PCNSL).

INTRODUCTION

Involvement of the Central Nervous System (CNS) with Non-Hodgkin's Lymphoma (NHL) falls into one of the three categories: Primary CNS Lymphomas (PCNSL), Disseminated Lymphoma with CNS involvement, and Primary Dural Lymphoma. Primary CNS lymphomas are typically diffuse large B-cell lymphomas and are found in the Brain parenchyma and other CNS structures [1]. Primary Central Nervous System Lymphoma (PCNSL) is defined as the involvement of brain, Leptomeninges, cerebrospinal fluid, eyes or spinal cord by an extranodal Non-Hodgkin Lymphoma (NHL) without evidence of a systemic lymphoma at the time of diagnosis.

It represents 0.8 to 6.6% of all primary CNS tumors [2]. Although the majority of the patients are immunocompetent, immunosuppression for autoimmune diseases and after organ transplantation, Acquired Immunodeficiency Syndrome(AIDS), Human Immunodeficiency Virus infection (HIV), and congenital immunodeficiencies are the major predisposing factors. Approximately 95% of PCNSL are diffuse large B-cell lymphoma (DLBCL) and the remaining cases are low grade B cell lymphoma of follicular, lymphoplasmacytic, and mucosa-associated lymphoid tissue types, Burkitt lymphoma or rarely T cell lymphoma [3,4].

CASE REPORT

A previously healthy 40 year old female admitted in NeuroSurgical department with complaints of, intermittent headache for 1 week, altered sensorium for 2 days and has 3 episodes of convulsions, loss of consciousness. She had right facio brachial paresis. She

was diagnosed as hypertensive 1month ago. She has no other significant past medical illness. On Neurological examination she was sleepy, arousable and disoriented but obeying few verbal commands with Glasgow Coma Scale of E4V2M6 with right facial palsy and grade 3/5 power in right upper limb. Her Computed tomography scan of Brain (figure 1.) showed hypodense extra-axial lesion of left fronto-temporo-parietal region with midline shift of 12mm and was reported as chronic Sub-dural Hematoma associated with mass affect. Within few hours multiplanar MRI of Brain (Figure2.) was performed and it showed an extra-axial Sub-dural Supratentorial large left fronto-temporo-parietal crescentic shaped mass with surrounding edema which is well defined and merged with adjacent normal dura. Correlating the CT and MRI findings, a presumptive pre-operative Radiological diagnosis of Sub-dural Hematoma or Enplaque Meningioma was made.



Fig-1



Fig-2

She underwent left fronto-temporo-parietal craniotomy. Intraoperatively thickened dura with a firm pinkish grey fleshy tumor underlying the dura was found firmly adherent to the dura and brain parenchyma. The tumor was readily separable from dura and brain parenchyma. There was no subdural hematoma or calvarial involvement. The tumor was dissected out from the brain parenchyma and excised along with the overlying adherent dura and duraplasty was done for the dural defect. The excised specimen was sent for histopathological examination and immunohistochemistry.

Histological examination

The surgical specimen consisted of multiple greyish brown fleshy plaque-like tissues measuring approximately 5.5 X 4 X 2 Cm in aggregate and membranous flat bit of 5.5 X 3.2 X 0.2 cm and cut surface is solid and firm, homogenous and greyish white. Paraffin-embedded sections showed a fibro collagenous and tumor cells arranged diffusely, composed of small round cells with irregular nuclear contours and coarse chromatin (Figure 3). There are many congested blood vessels seen within the tumor tissue suggestive of Non Hodgkin's lymphoma (Figure 4).

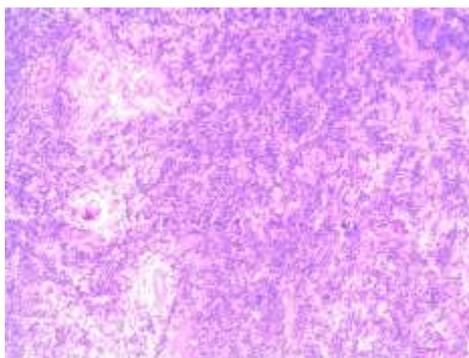


Fig-3: Small round cells with irregular nuclear contours and coarse chromatin

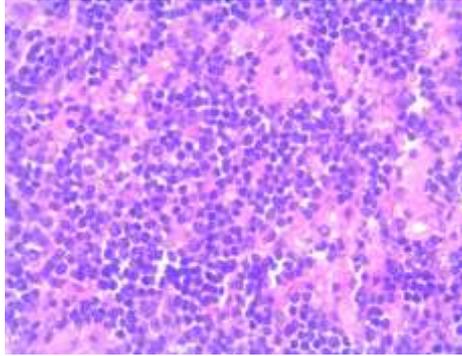


Fig-4: Congested blood vessels with Small round cells with irregular nuclear contours and coarse chromatin

Immunohistochemistry was performed after the Histopathological examination, which revealed CD20+ (Figure 5,6), CYCLIN D1+(Figure 6), Bcl-2

+(Figure 7), Ki 67 +(Figure 8) suggestive of grade I Follicular Lymphoma.

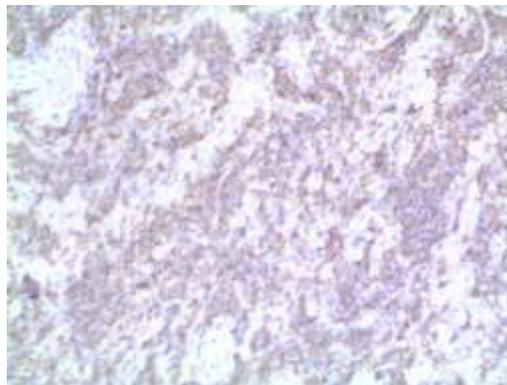


Fig-5: HPE Low power CD 20

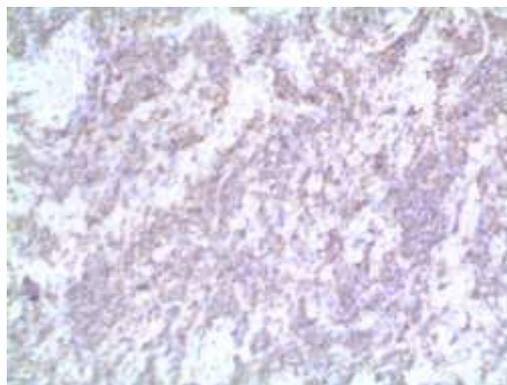


Fig-6: HPE High power CD 20

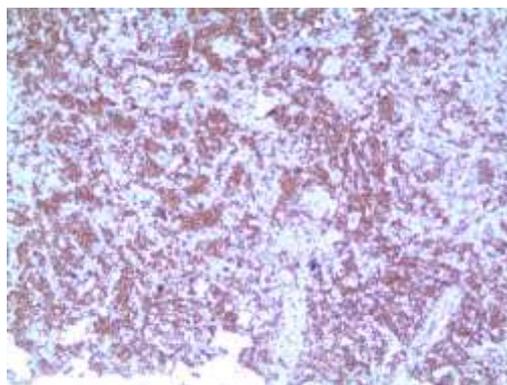


Fig-7: Bcl-2 Low power

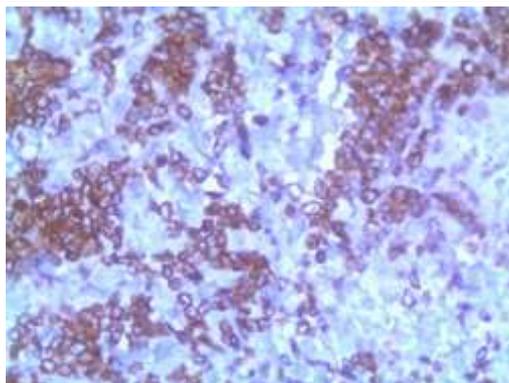


Fig-8: Bcl-2 High power

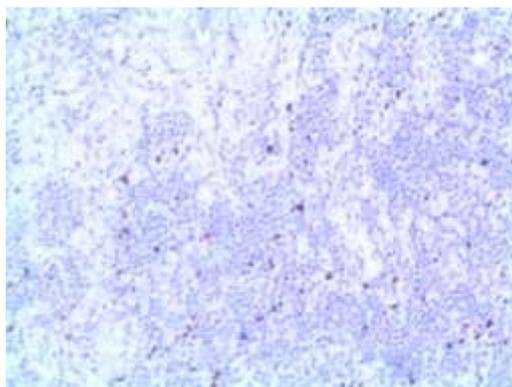


Fig-9: Ki 67 + Low power

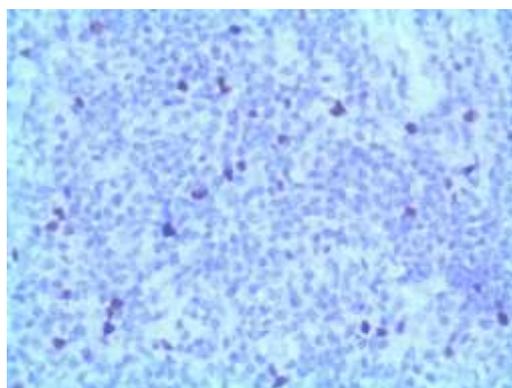


Fig-10: Ki 67 + High power

Post operatively the patient underwent extensive work up for extra cranial primary Lymphoma. Clinical examination, CT scan of chest and abdomen were unremarkable. Laboratory investigations and Bone Marrow biopsy, blood investigations for immune status, HIV, EBV were negative. The patient was referred to oncologist.

DISCUSSION

Approximately 95% of Primary CNS Lymphomas are diffuse large B cell lymphoma (DLBCL) and the remaining cases are low grade B cell lymphomas of follicular, lymphoplasmacytic, and mucosa-associated lymphoid tissue types, Burkitt lymphoma and rarely T cell lymphoma [3, 4]. Ninety percent of non-HIV-associated PCNSL is diffuse large

B-cell (DLBCL) type, and the remaining 10% are poorly characterized low-grade lymphomas, Burkitt's lymphomas, or T-cell Lymphomas [5]. There are several hypotheses which aim to explain the pathogenesis of PCNSL. Since the central nervous system lacks lymphoid tissue or lymphatic vessels, PCNSL may be caused by the monoclonal proliferation of continuously trafficking T-cells or B-cells in CNS, or the specific tropism of neoplastic T or B lymphocytes for CNS. Malignant transformation of T or B cells after a benign inflammatory process within the CNS may also be the origin of PCNSL. Additionally, neoplastic lymphocytes eradicated by the intact peripheral immune system may escape to the CNS [3].

PCNSL likely arises from late germinal center or post germinal center lymphoid cells and may localises to the CNS because of a poorly understood neurotropism [6].

CONCLUSION

In summary, we report an unusual presentation of a primary malignant B-cell Dural Lymphoma, a rare subtype of PCNSL without systemic involvement, who's CT scan, mimicked chronic subdural hematoma. Surprising points of this case include the very short clinical history of the onset of the patient's symptoms, rapid neurological deterioration, confusing imaging findings and the final diagnosis of a high-grade PCNSL. In spite of vast clinical search and an improved research in depth on etiology of this disease, still clinicians face difficulty in diagnosing this type of presentation in acute setting. Though MR imaging differ between immunocompetent and immunocompromised in most cases, the difference in presentation, neuroimaging and its final diagnosis makes this disease a challenging task to clinicians by its exact match with other entities like sub dural hematoma or Enplaque meningioma. So clinicians have to be vigilant to include PCNSL as a differential diagnosis, as noticed in our case.

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