

Primary Dural Lymphoma-A Case Report

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Abstract: Primary dural lymphoma is one of the rare manifestation of extranodal nonHodgkins lymphoma. These tumours are slow growing and does not show any evidence of systemic involvement. On MRI dural lymphoma mimics other dural based neoplasms like en plaque meningiomas. we report a case of primary dural lymphoma in a 37 year old female presenting as extradural mass which was surgically removed.

Keywords: Primary dural lymphoma, Dural based mass, Enhancement.

I. Introduction

Primary lymphoma of CNS is a Non-Hodgkins lymphoma that occurs in brain in the absence of systemic involvement (1). Primary Dural Lymphoma (PDL) is a subtype of primary CNS lymphoma which presents as a single or multiple extraaxial masses like meningioma. Though dural lymphomas are found along cerebral convexities, they are also seen in falx, tentorium, sella, parasellar region and spine (2). Dural lymphomas have to be differentiated from similar dural based lesions like solitary fibrous tumour, leiomyosarcoma, dural metastasis, neurosarcoidosis etc. In the present study we report a case of primary dural lymphoma in a 37 year old female who presented with altered behavior and seizures.

II. Case Report

A 37 year old female was brought to the hospital with progressive altered behavior of ten days duration associated with focal seizures, urinary incontinence, right upper limb weakness. There was no history of head injury, vomitings, hypertension, diabetes. She had a past history of Intermittent headache since one year. On clinical examination patient was conscious, irritable, right hemiplegia and right facial palsy were present. Routine lab tests were within normal limits. She was nonreactive for HIV I, HIV II and Hbs Ag.

Contrast enhanced CT brain showed large enhancing, iso attenuating extradural mass along the left frontoparietal convexity (Fig 1). No evidence of focal calcifications or erosion of adjacent bone. MRI brain with contrast revealed a large extradural mass which is hypointense on T1 weighted image, mildly hyperintense on T2 weighted and Flair imaging with no restriction on Diffusion or blooming on GRE (Fig 2). Post contrast there was profuse enhancement of the mass (Fig 3a, 3b). There was mild ipsilateral ventricle compression with midline displacement. Adjacent brain showed grade II perilesional edema with indistinct brain-tumour interface. On surgery greyish, nonsuckable tumour located adherent to dura was found in left frontoparietal region. The tumour was excised in total. HPE showed fibrocollagenous tissue, diffusely arranged small round cells with irregular nuclear contours and coarse chromatin. There are many congested blood vessels seen within the tumour, features suggestive of Non-Hodgkins lymphoma. Immunohistochemistry was positive for CD20, cyclinD1, ki 67 (Fig. 4, 5, 6, 7).

III. Discussion

There are many neoplastic and a non-neoplastic lesion that involves the dura of which meningioma is the most common entity. The list includes hemangiopericytoma, lymphoma, Solitary fibrous tumour, metastasis, Rosai-Dorfman disease, Epstein bar virus associated smooth muscle tumours, melanocytic neoplasms, Erdheim Chester disease, tuberculosis, neurosarcoidosis etc. (3,4).

Primary dural lymphoma is a subentity of primary leptomeningeal lymphoma which is a subtype of primary CNS lymphoma. The incidence of PDL is less than 1 % of all CNS lymphomas. Majority of dural based lymphomas are Mucosa associated lymphoid tissue (MALT) Lymphomas. Though the CNS does not have lymphoid tissue, the origin of dural based Malt lymphoma may be from 1) Meningeal seeding from an undiagnosed systemic MALT lymphoma. 2) Inflammatory conditions involving the dura can result in accumulation of polyclonal lymphocytes from which MALT lymphoma can occur (5, 6).

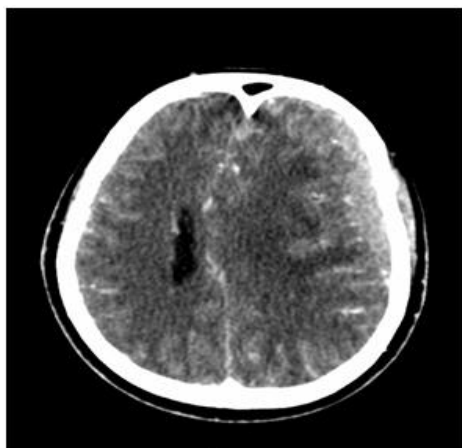
Primary dural lymphomas are usually reported in middle aged females with slow progression of symptoms like headache, seizures and meningeal signs. Sunil et al (7) reported a case of primary diffuse large B cell lymphoma in a 14 year old boy. The tumour can be single or multiple and closely mimics meningiomas. On imaging they are hyperattenuating on CT, iso to hypo intense on T2 weighted MR imaging with profuse enhancement on contrast images. Adjacent brain parenchyma shows vasogenic edema. There will be indistinct brain-tumour interface which is suggestive of PDL than Meningioma (8). The presence of vasogenic edema and parenchymal brain invasion with indistinct brain tumour interface suggests PDL than meningioma (9). PDL has to be differentiated from many benign and malignant lesions involving dura as some of the imaging features are similar. Hemangiopericytomas are more common in young males; they frequently cause adjacent bone erosion. On imaging they are almost always solitary, attached to dura with a narrow base. They are typically isointense to grey matter on T1W and T2W images with prominent flow voids. Hemorrhage, necrosis, cystic areas, lobulated contours are more common in anaplastic form. MR spectroscopy shows a high myo-inositol peak at 3.56 ppm and lack of

alanine .A characteristic histologic feature is presence of prominent network of reticulinfibers. Immunohistochemistry shows CD34 positive, 70 kD protein complex negative.

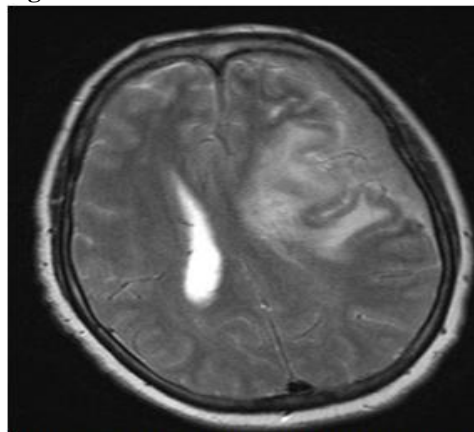
Metastasis to dura is usually from breast, lung, prostate. They are frequently solitary. On imaging they appear as linear dural thickening or nodular lesions. Histological studiesreflects the primary neoplasm involvement. Immunohistochemistry evaluation shows strong pancytokeratin immunoreactivity with negative EMA. Solitary fibrous tumor are benign spindle cell tumours, they rarely involves central nervous system. The common sites are along the falx, occipital and spinal dura. Imaging features that suggest the diagnosis include presence of intratumoral calcification, heterogenous signal intensity on T2weighted images with profuse contrast enhancement and flow voids. On MR spectroscopy lipid and lactate peaks and elevated myoinositol (3.5ppm) seen. The pathological features include presence of spindle cells with intercellular collagen, diffuse CD34 positive and EMA negative. Rosai-Dorfman disease is a rare benign histiocytosis, usually affects children and young adults. The common sites affected are parasagittal, petroclival and suprasellar regions. These lesions are either solitary or multiple. Isointense on T1weighted images and iso to hypointense on T2 weighted Images and are relatively hypovascular on angiography. Histological studies show polymorphous infiltrate of lymphoplasmacytic cells and histiocytes with immunoreactivity for S100 protein and CD68.EBV associated smooth muscle tumours (Leiomyoma, Leiomyosarcoma) have to be considered in differential diagnosis of dural based tumours in HIV positive patients. Lesions are hypo to isointense on T1W images and iso to hyperintense on T2 W images with profuse contrast enhancement.

Erdheim Chester Disease is a non Langerhans cell histiocytosis that can rarely involve CNS. The meningeal subtype is described as nodular thickening along the dura with diffuse pachymeningeal thickening. The lesions are iso-hypo intense on T1weighted and T2weighted images. They are often multiple. Histological examination reveals large histiocytes with a background of fibrosis. Immunohistochemistry reveals CD68 positive, CD1a negative. Melanocytic neoplasms are rare cause of dural masses which are typically hyperintense on T1W images, iso to hypo intense on T2W images. Pathologic features are presence of melanin pigment with in cytoplasm of cells. IHC positive for S100, Melan A, HMB-45. In Neurosarcoidosis, the dural involvement will be in the form of diffuse thickening or focal and mass like. They are isointense on T1W and hypointense on T2W images. In the present case based on MRI findings like grade II vasogenic edema in adjacent brain and indistinct brain-tumour interface, a diagnosis of PDL was made which was confirmed on histopathology. The only significant finding in clinical history was acute onset of neurological deficit which was unusual. Surgery followed by chemotherapy provides complete treatment in these patients(10,11).

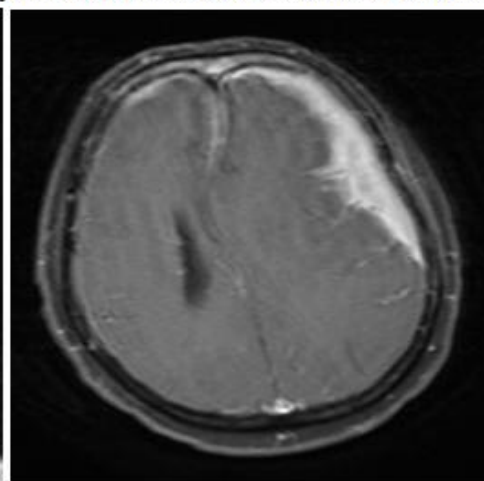
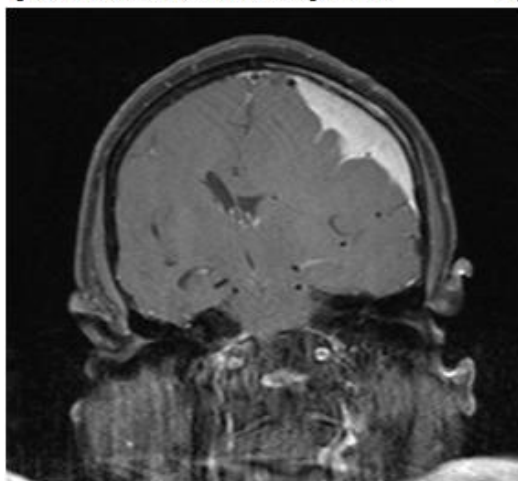
IV. Figures



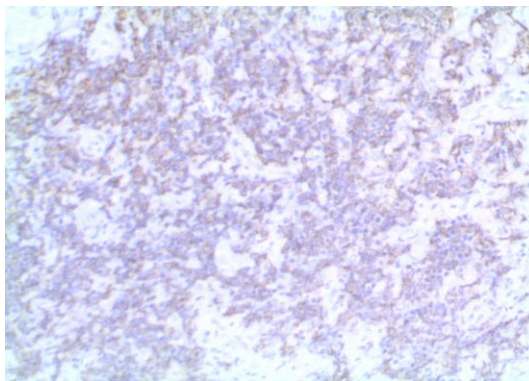
"Fig1": contrast CT shows extradural mass with ipsilateral lateral ventricle compression.



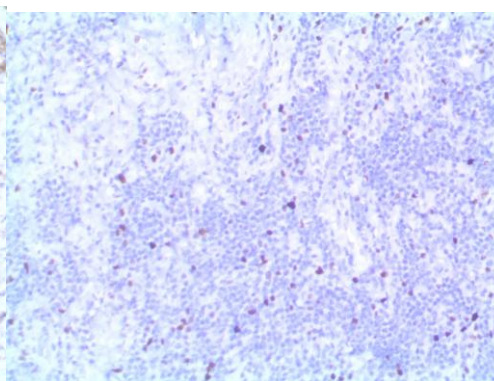
"Fig 2"-Axial MR T2 weighted image shows hyperintense extradural mass with edema of adjacent brain and indistinct brain tumour interface.



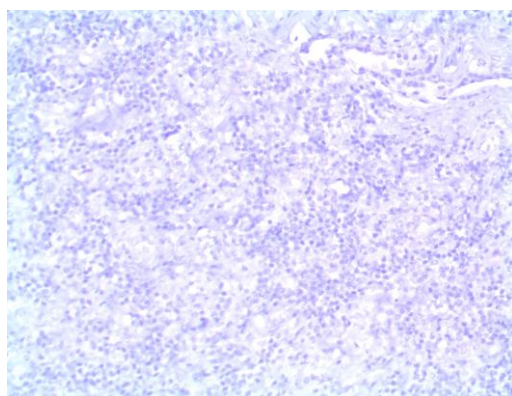
"Fig 3a & 3b" showing profuse enhancement of the extradural mass.



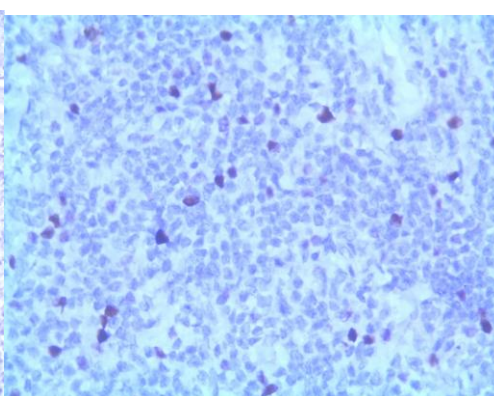
“Fig 4” : CD20



“Fig 6” : Ki 67 low power.



“Fig 5” : CYCLIN D1



“Fig 7” : Ki 67 high power.

V. Conclusion

Enhancing extradural masses in adult population has wider differential diagnosis. Thorough evaluation with MRI with contrast followed by surgical excision with HPE supported by proper immunohistochemistry has to be performed to make accurate diagnosis which is necessary for further management.

References

- [1]. Edelman,Hesselink,Zlatkin,Cruess.chapter 40 supratentorial adult brain tumours in Clinical Magnetic Resonance imaging. volume2. Third edition, 2006, 1097-1136.
- [2]. I.S.Haldorsena,A.Espelanda,E.-M.Larsson:Central nervous system lymphoma:characteristic findings on Traditional and advanced imaging, AJNR 2011,32:984-992.
- [3]. Alice Boyd smith,Iren Horkanyne-Szakaly,Fason W.Schroeder,Elisabeth F.Rushing;Mass lesions of the Dura:Beyond Meningioma-Radiologic - Pathologic correlation, Radiographics 2014,34:295-312.Neurologic/Head and neck imaging.
- [4]. Danai Chourmouzi,StamatiaPotsi,Anestis Moutzouoglou,Elisavet Papadopoulou,Kostas Drevelegas,ThomasZaraboukas,and Antonios Drevelegas:Dural lesions mimicking meningiomas:A pictorial essay, World J Radiol. Mar28,4(3),2012,75-82.
- [5]. Ferguson SD,Musleh W,Gurbuxani S,Shafizadeh SF,Lesniak MS.Intracranial mucosa-associated lymphoid tissue (MALT) lymphoma, J clin Neurosci 17(5),2010,666-669.
- [6]. Kumar S,kumar D,Kaldjian EP,Bauserman S,Raffeld M,Jaffe ES.Primary low grade B cell lymphoma of the dura:a mucosa associated lymphoid tissue type lymphoma, AmJ surg Pathol 21(1),1997,81-87.
- [7]. Sunil Munakomi,Bhattarai B,Srinivas B,Cherian I. Case Report:Primary dural based diffuse large B cell lymphoma in a 14 year old boy, F1000 Research,25 Mar 2015,4:78.
- [8]. Iwamoto FM,De Angelis LM,Abrey LE.Primary dural lymphomas:a clinicopathologic study of treatment and outcome in eight patients, Neurology 66(11),2006,1763-1765.
- [9]. Won Hee Lee,Bomi KIM,Moo Seong KIM;A Case of Primary Dural Lymphoma:Diffuse Large B-cell type, Turk Neurosurg vol:24,No:5,2014,799-803
- [10]. Angelo Borsarelli Carvalho Brito,Fabiano Reis,Carmino Antonio de Souza,Jose vassallo,Carmen Silvia,Passos Lima;Case report-Intracranial primary dural diffuse large B cell lymphoma successfully treated with chemotherapy, Int J Clin Exp Med 7(2),2014,456-460.