Case Report

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Inflammatory Myofibroblastic Tumor of Central Nervous System: A Case Report

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Abstract

Since many year there has been a debate regarding the disease entity of inflammatory myofibroblastic tumor (IMT) in the central nervous system (IMT-CNS) because of its rarity and high frequency of recurrence. IMT-CNS is an important differential diagnosis among tumor-like intracranial lesions, and total resection is required. A 4-year-old female presented with a rare inflammatory myofibroblastic tumor (IMT) manifesting as a recurrent headache, vomiting, and one episode of GTCS. Computed tomography Scan Brain and contrast revealed an isointense extra-axial lesion involving the left parieto-occipital region with intense homogenous contrast enhancement. The tumor was grayish-white, firm to hard moderately vascular and was resected en bloc with a clear margin. Histological examination revealed multiple spindle cells with plasma cells and lymphocytes scattered among these spindle cells. The spindle cells were diffusely immunopositive for vimentin and negative for smooth muscle actin, epithelial membrane antigen, S100 negative, and CD34 negative so a diagnosis of benign spindle cell lesion inflammatory myofibroblastic tumor was rendered.

Key words: Central nervous system, Inflammation, Tumor

INTRODUCTION

Inflammatory myofibroblastic tumor (IMT) is a rare lesion consisting of myofibroblastic spindle cells and inflammatory cells that occurs primarily in soft tissue and the viscera of children and young adults.[1,2] IMT has been described under many different names, such as inflammatory pseudotumor and plasma cell granuloma. It usually follows a benign clinical course and is most common seen in the lung, omentum, and mesentery. Diagnosis depends on histological examination because the radiological and clinical findings are non-specific. IMT is a rare disease with unknown etiology characterized by non-neoplastic polyclonal proliferation of mature plasma cells and other mononuclear cells. IMT in the central nervous tends to arise from meningeal structures. The disease entity has a high frequency of recurrence and malignant transformation compared with IMT not affecting the central nervous system (CNS).[3]

Here, we describe a case of cerebral intraparenchyma-tous IMT in an adult, which seemed to exhibit the benign and inflammatory characteristics of IMT-CNS or inflammatory pseudotumor.

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CASE REPORT

The 4-year-old female was admitted to our emergency department with one episode of generalized tonic-clonic seizures. She was also suffering from recurrent headaches and occasional vomiting for the past 3 months. Neurological examination revealed GCS of 14/15 and right hemiparesis MRC grade 4/5. Fundus examination revealed bilateral papilledema. Results of blood examination were normal. Computed tomography scan brain (Figure 1) revealed an isointense extra-axial lesion involving the left parieto-occipital region with intense homogenous contrast enhancement, causing squashing of ipsilateral ventricle and hydrocephalus. Grossly, it was grayish-white, firm to hard moderately vascular, and arising from left anterior petrous

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dura mater (Figure 2). Morphological appearance was like meningioma. The lesion was resected en bloc.

Histopathology revealed multiple spindle cells in patternless arrangement with plasma cells and lymphocytes scattered among these spindle cells. There was no pleomorphism, mitoses or necrosis. The spindle cells were diffusely immunopositive for vimentin and negative for smooth muscle actin, epithelial membrane antigen, S100 negative, and CD34 negative so a diagnosis of benign spindle cell lesion IMT was rendered.

DISCUSSION

IMT is a rare tumor that can exceptionally be found in the CNS. IMT is a distinctive myofibroblastic spindle cell lesion accompanied by an inflammatory infiltrate of plasma cells, lymphocytes, and eosinophils.[4] Three subtypes have been described: (1) Compact spindle cells with inflammatory cells, (2) inflammatory areas resembling nodular fasciitis, and (3) dense plate-like collagen resembling scar.

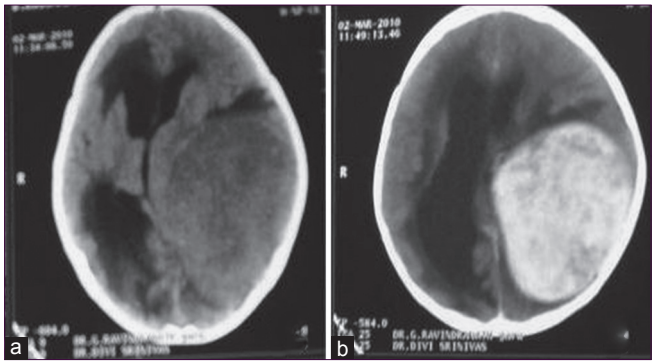

Figure 1: (a and b) Computed tomography scan brain showing an isointense extra-axial lesion involving the left parieto-occipital region with intense homogenous contrast enhancement, causing effacement of ipsilateral ventricle and hydrocephalus

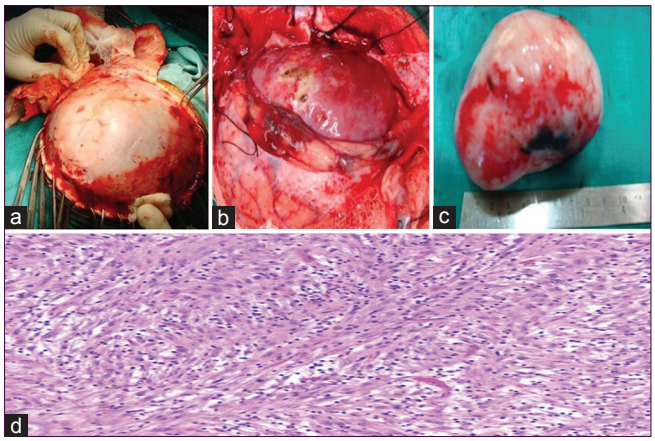

Figure 2: (a-d) Grayish-white, firm to hard moderately vascular tumor. Furthermore, seen in the histopathological picture (d) multiple spindle cells in patternless arrangement with plasma cells and lymphocytes scattered among these spindle cells without any pleomorphism or necrosis

IMT-CNS is rare, with only approximately 100 reported cases.[3] IMTs of the CNS can be classified into two histopathological types: A form rich in spindle myofibroblasts mixed with few inflammatory cells, also called the FHC variant, and the PCG-like type composed mainly of plasma cells and lymphocytic infiltration. Recent case series proposed that the two types are different in terms of tumor aggressiveness. Localization of IMT can be divided into 5 types: Intraparenchymatous, meningeal, mixed intraparenchymatous and meningeal, intraventricular, and extending to the cranial cavity/sphenoid sinus.[3] The etiology of IMT-CNS is unknown, but 60% of IMT-CNS tumors have arisen from the dural/meningeal structures and only 12% from intraparenchymatous lesions.[3] The present case had attachment to dural/meningeal structures, and was classified as the meningeal type.

The neuroimaging findings and clinical course of IMT-CNS are non-specific, such that differential diagnosis is important in the case of meningeal lesions, especially with meningioma or sarcoma.[5] Earlier cases likely included an indiscriminate mixture of neoplastic and non-neoplastic process. Of patients with extrapulmonary IMT, 15% had one or more recurrences over a period of 1-24 months (mean 6 months),[1] but patients with IMT-CNS had recurrence rate after incomplete resection of 40% within 2 years.[3] Two of the three young patients with IMT-CNS extending from the meningeal structures into the brain tissue showed local recurrence after resection of the tumor and histological investigations revealed transformation into a semi-malignant fibrohistiocytic tumor in one patient.[3] Therefore, whether IMT-CNS is a different entity from IMT non-CNS remains controversial. Therefore, patients need close follow-up. The FHC variant often contains clonal rearrangements in chromosome band 2p23 that constitutively activate the *ALK* gene.[6] *ALK* is a tyrosine kinase receptor that is normally expressed in the developing CNS. In IMTs located outside of the CNS, investigators have reported several fusion genes that render *ALK* oncogenic. *ALK* rearrangements with specific fusion genes have been reported in other types of cancer such as anaplastic large cell lymphoma, non-small cell lung cancer, and renal medullary carcinoma.

IMT of the CNS that express *ALK* can have an aggressive course despite gross total resection. The *ALK* expression in IMT of the CNS is specific to the FHC variant. Compared with IMT of the CNS that do not express *ALK*, the reported recurrence rate of *ALK*-positive tumors tend to be higher. Further research with longer follow-up is needed to clarify the natural history of this rare tumor.

CONCLUSION

Total resection IMTs should be achieved as theses tumors recur often rapidly. Confirmation of the FHC variant by histopathology warrants searching for *ALK* expression. Such patients can be offered adjuvant therapy such as radiotherapy or novel *ALK* inhibitors.

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