CASE REPORT

Inflammatory myofibroblastic tumor parieto-occipital bone

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ABSTRACT

Inflammatory myofibroblastic tumor is a diverse group of lesions first described in lung and later on reported in various sites like stomach, bowel, spleen, bone. We report a case of inflammatory myofibroblastic tumor in a 30-year-old male who presented with a slowly progressive scalp swelling of two-year duration. Magnetic resonance imaging showed an intradiploic well enhancing lesion in parietal and occipital bone, isointense on T1 weighted images and hypo intense on T2 weighted images with dural enhancement. On histopathological examination, the lesion was composed of variable admixture of spindle cells with eosinophilic cytoplasm and inflammatory cells comprising of plasma cells and lymphocytes. The lesion was infiltrating the underlying dura. The spindle cells showed strong positivity for smooth muscle actin on immunohistochemistry. A final histopathologic diagnosis of inflammatory myofibroblastic tumor was rendered.

KEY WORDS: Bone, inflammatory myofibroblastic tumor, parietal bone

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INTRODUCTION

Inflammatory myofibroblastic tumor (IMT), also called earlier as inflammatory pseudo tumor, nodular lymphoid hyperplasia, plasma cell granuloma, are rare groups of lesions characterized histologically by acute and chronic inflammatory cells with variable degree of fibrous stroma.[1] Clinically and radiologically they may look like malignant tumors. First described in lungs; a large number of extra-pulmonary sites of IMT have been described in literature.[2,3] We present a case of IMT of parieto - occipital bone in a young adult male with involvement of the dura.

CASE REPORT

A 30-year-old male presented with history of swelling in the left parieto-occipital region since two years. The swelling was slowly progressive with associated pain since the last one year. There was no history of seizures, nausea or vomiting. On examination the swelling measured 8 x 5 cm, was globular and hard in consistency. No sensory or motor deficits were noted. Computed tomography (CT) scanning revealed an osteolytic contrast enhancing lesion destroying the outer table of skull [Figure 1a]. No intracranial extension was seen. Magnetic resonance imaging (MRI) showed a well defined intra-diploic contrast enhancing lesion measuring 5.9 × 1.5 × 5.5 cm in the left parietal and occipital bone, isointense on T_1 weighted images and hypo intense on T_2 weighted images with dural enhancement [Figure 1b]. The underlying brain parenchyma was normal. A radiological impression of eosinophilic granuloma was given. Ultrasonography of the abdomen was normal. CT of the thorax was normal. The patient was taken for surgery and an enbloc excision of left parieto-occipital bony lesion with excision of dura followed by duroplasty and cranioplasty was done. The lesion was sent for histopathological examination.

Grossly, the lesion measured 7 × 7 × 2 cm, adherent to the scalp bone. The cut section was gray white and firm in consistency. Five micron thick sections were cut and stained with hematoxylin and eosin. On light microscopy, the tumor was composed of diffuse sheets of oval to spindle shaped cells intersected by fibro-collagenous bands [Figure 2a]. The cells showed moderate to abundant granular eosinophilic cytoplasm with nuclei showing convolution at places with a fine chromatin pattern [Figure 2b]. In addition, numerous plasma cells, few lymphocytes with lymphoid follicle formation at places and nutrophils were seen intermixed intimately with the tumor cells [Figures 3a, 3b]. A few cells showed crystalline substances in the cytoplasm. On immunohistochemistry, the cells showed strong positivity for smooth muscle actin and vimentin [Figures 4a, 4b] and focal positivity for epithelial membrane antigen in the plasma cells. The tumor was infiltrating the dura and involving the skull bone [Figure 5]. No meningothelial cells were seen. Due to the non-availability of anaplastic lymphoma kinase -1 antibody, we could not perform further tests for myofibroblastic differentiation. On the basis of the histomorphological features and immunohistochemistry a final diagnosis of inflammatory myofibroblastic tumor, parieto-occipital bone with infiltration into the underlying dura.

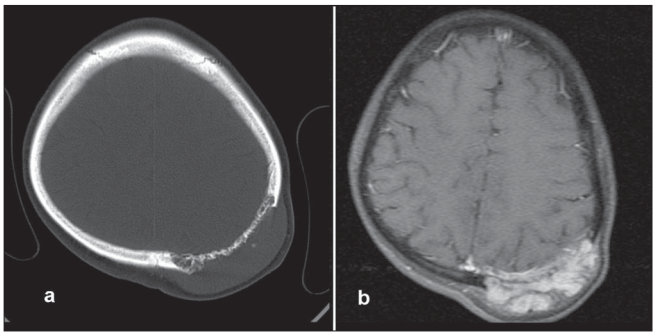

Figure 1: (a) Computed tomography axial section showing a destructive osteolytic lesion of the parietal bone with destruction of the outer table with a soft tissue mass overlying the defect, (b) Magnetic resonance imaging showing well defined intradiploic contrast enhancing mass in the parietal bone with extension into the occipital bone

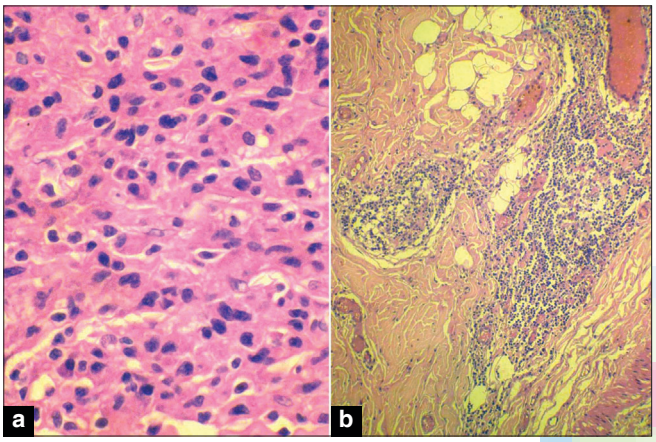

Figure 3: (a) Paraffin section plasma cells intermixed with the tumor cells (H and E, ×400), (b) Paraffin section showing lymphocytes with follicle formation at places (H and E, ×100)

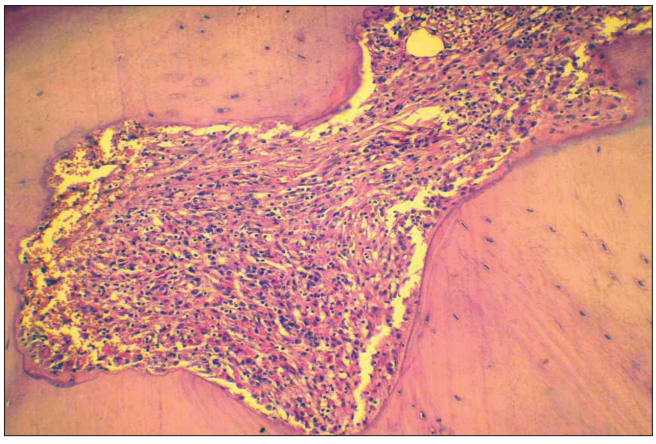

Figure 5: Paraffin section showing tumor involving the bone. (H and E, ×100)

DISCUSSION

Inflammatory myofibroblastic tumor is currently accepted to be a true neoplasm with a wide spectrum of biological behavior, varying from benign lesions to the rare tumors which

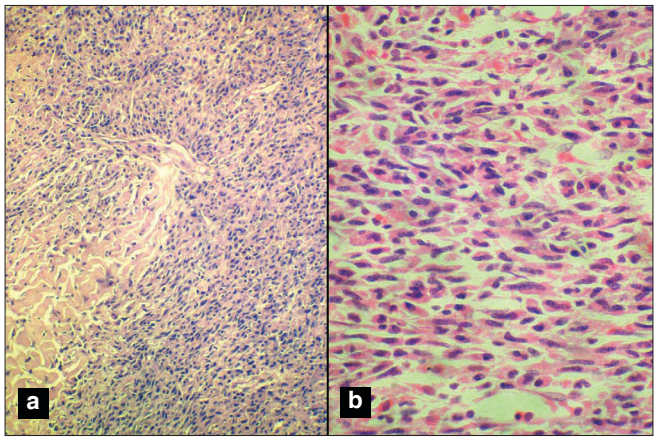

Figure 2: (a) Paraffin section showing diffuse sheets of oval to spindle cells intersected by fibrocollagenous bands (H and E, ×100), (b) Paraffin section showing spindle shaped cells with dense eosinophilic cytoplasm (H and E, ×400)

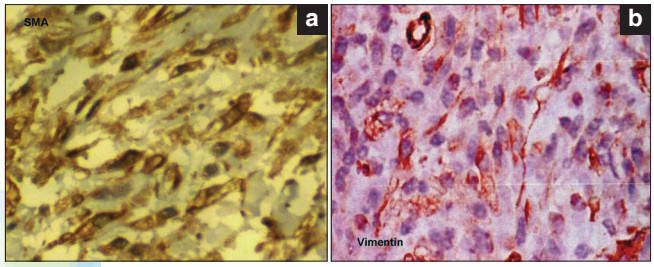

Figure 4: Immunohistochemistry showing tumor cells positive for (a) smooth muscle actin and (b) vimentin (Avidin Biotin Complex Immunoperoxidase, ×400)

are multifocal and prone to recurrence.[4] Some researchers suggest the role of cytokines, particularly interleukins - 6 in its pathogenesis.[5]

IMT involving the central nervous system are described in various locations like cranial, spinal, and orbital. Most of these were dural based and radiologically showed two patterns: isolated mass forming and an en plaque pattern.[6] IMT in bone is very unusual, and there are only two cases reported, involving the temporal bone.[7] Gasparotti *et al.*[8] described a case of IMT in temporal bone in a young black patient, which on radiological imaging revealed a homogenously enhancing soft tissue mass of the right mastoid with bone erosion of the tegmen and extensive dural thickening. The present case lacked these clinical symptoms and presented solely with scalp swelling. The present case did not have pachymeningitis, cranial neuropathy or uveitis as described in the other case report.[9] Most of the bone IMT are osteolytic in appearance and a destructive bone lesion expanding into the soft tissue are well described, as was found in the present case.[7] However, it must be noted that it is very difficult, both clinically and radiologically, to decide whether a bone tumor with infiltration into the soft tissue is a pseudo-tumor or a true neoplasm. In our case, the lesion was first thought to be neoplasm such as meningioma, eosinophilic granuloma, metastasis, or an

aggressive infectious process. In such instances, without a biopsy, differentiation between the above mentioned differential diagnosis is difficult. Tumors composed of myofibroblast and fibroblasts which pose significant challenge in differential diagnosis are aggressive fibromatosis, fibrosarcoma or other spindle cell malignancies. In fibromatosis, the spindle cells are more slender and there is more prominent collagenous tissue. The dense inflammatory component will be lacking in fibromatosis. Fibrosarcoma is a malignant tumor and will show herring bone pattern of arrangement of cells with mitotic activity. However, there is undoubtedly morphologic and clinical overlap between IMT and inflammatory fibrosarcoma. The question of distinction between the two rests on whether they are truly distinct or a part of neoplastic continuum of myofibroblastic proliferation with increasing cellular atypia and aggressiveness.

Our case showed histologic benign features like low cellularity, minimal nuclear atypia and no mitoses. Since it was a scalp tumor a differential diagnosis of eosinophilic granuloma, lymphoplasmacyte rich meningioma was also considered. However, no eosinophils or typical langerhans cell histiocytes with coffee bean nuclei were seen. No meningothelial clusters were seen and the spindle cells were negative for epithelial membrane antigen. MR imaging also showed extensive dural thickening and enhancement. Dural thickening and enhancement represent an unspecific reactive change to any adjacent dural lesion. They seem to be a common finding in IMT of the skull base, as reported by Han *et al.*[10] who observed intracranial dural involvement in all patients with fibrosing inflammatory pseudo tumors of the skull base.

The only effective treatment of IMT is complete surgical resection. Because of the higher recurrence rate, approaching 25% in extra pulmonary IMT, the patient has been kept on close follow-up.

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