Case Report

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Pathology Section

Benign Fibrous Histiocytoma: An Uncommon Presentation

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ABSTRACT

Intracranial fibrous histiocytomas are rare; Benign Fibrous Histiocytoma (BFH) being uncommon than its malignant counterpart. BFH comprises fibroblasts and histiocytes without any nuclear pleomorphism or atypia. We present a case of a 42-year-old male who had swelling over the occipital region for the past five years, which progressively increased in size. He developed headache, dizziness, and gait disturbance over the last six months. Computed tomographic scan revealed a posterior fossa space-occupying lesion. Fine-needle aspiration cytology from the swelling revealed spindled fibroblasts along with histiocytes and multinucleated giant cells. Later, histopathology showed presence of spindle-shaped cells in storiform pattern admixed with histiocytes and giant cells. The giant cells and histiocytes were immunopositive for CD68 and spindled cells were positive for vimentin, but immunonegative for CD34, epithelial membrane antigen, CD1a and S100. The final diagnosis was intracranial BFH. We present this case because of its extreme rarity and unusual location.

Keywords: Intracranial, Meninges, Middle-aged man

CASE REPORT

A 42-year-old male presented to the neurosurgery outpatient department with a firm swelling over the occipital region of the scalp, measuring about 6 cm × 5 cm [Table/Fig-1a]. Initially, the swelling was small (1.5 cm × 1 cm). It gradually increased in size over the last five years. The patient had no neurological symptoms until the last 6 months, when he suddenly developed headache, dizziness and disturbance of gait.

The patient underwent computed tomographic scan of brain which revealed an intracranial well-circumscribed expansile posterior fossa space-occupying lesion extending to suboccipital region destroying the occipital bone. The lesion was of heterogeneous density with moderate enhancement and it measured 65 (AP) × 54 (Tr) × 65 (V) mm^3 [Table/Fig-1b].

Fine-needle aspiration cytology of the lesion yielded a blood-mixed particulate material. Cytological smears were moderately cellular and comprised dispersed population of oval to spindle shaped fibroblasts along with histiocytes admixed with multinucleated giant cells. Overall, cytomorphological features were suggestive of a fibrohistiocytic lesion [Table/Fig-1c, d].

Subsequently, the tumour was excised and it was sent for histopathologial examination. Grossly, multiple friable grayish-white solid tissue pieces were received, altogether measuring 6 cm × 5 cm × 2 cm [Table/Fig-1e]. Sections from routine formalin fixed paraffin embedded tissue stained with hematoxylin and eosin revealed ovoid to spindle shaped fibroblastic cells in fascicles as well as storiform pattern. Histiocytes and multinucleated giant cells were also noted [Table/Fig-1f].

Immunohistochemistry was done using antibodies against vimentin, CD68, Progesterone Receptor (PR) [Table/Fig-1g], CD34, CD1a, and epithelial membrane antigen. The spindled stromal cells were positive for vimentin [Table/Fig-1h] but negative for all the other markers. The histiocytes and multinucleated giant cells were positive for CD68 [Table/Fig-1i]. Reticulin stain revealed no increase in reticulin fibrosis [Table/Fig-1j].

Clinicoradiological correlation along with cytology, histopathology, and immunohistochemistry helped to establish the final diagnosis of primary intracranial BFH.

After one year, on follow-up, the patient did not have any neurological symptom and was ambulant.

DISCUSSION

BFH occurs predominantly in the soft tissues and bones [1]. Our case is one of the rare cases of BFH which had an intracranial location. BFH in the central nervous system is thought to arise from the brain parenchyma, dura, spine, or may extend intracranially from the skull base or the pterygopalatine fossa [2-4]. In this case, the tumor originated from the dura, had destroyed the overlying occipital bone.

Most cases of intracranial BFH that have been reported in literature affected the paediatric population, commonly below 24 months of age [5,6]. Hence, our case is also unique by the fact that the patient was 42-year-old at presentation.

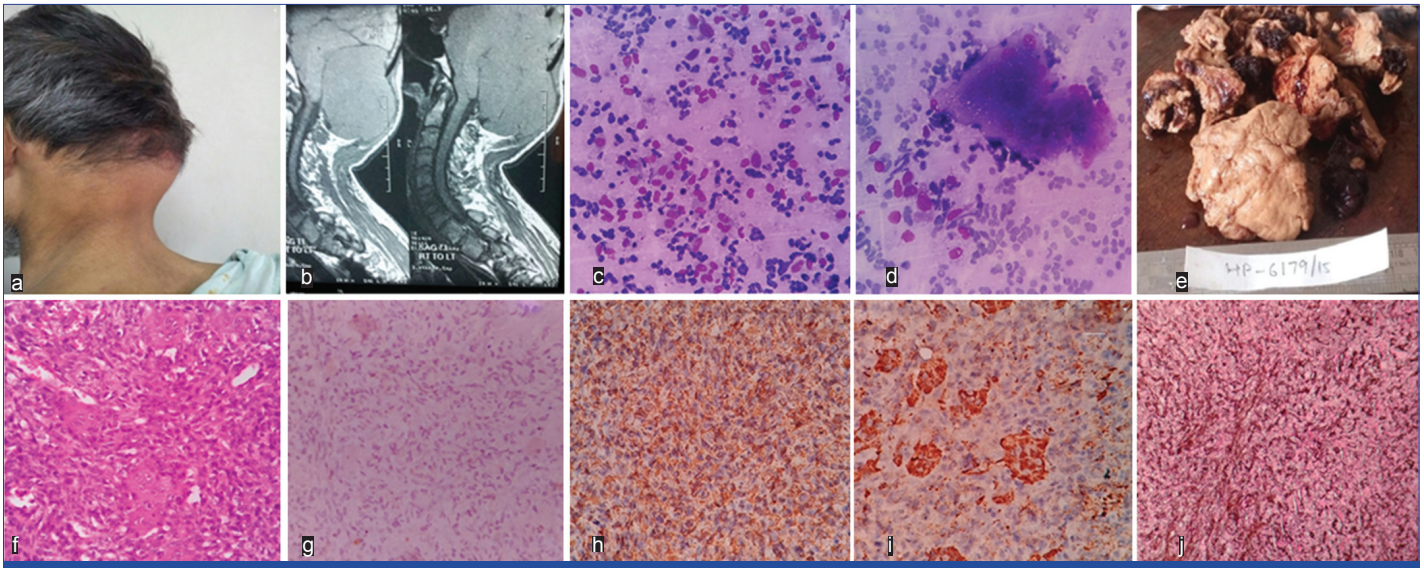
The origin of intracranial fibrohistiocytic tumours is still under debate [5,7]. Perivascular pial sheath and primitive mesenchymal cells are probable cells of origin of this rare tumour [5].

Histologically, BFH comprises of predominantly two types cells - atypical fibroblasts and histiocytes. The cells are arranged in storiform pattern as well as interlacing fascicles [8]. In the present case, histology of the excised intracranial tumour revealed bland fibroblasts which lacked any nuclear atypia and histiocytes arranged in fascicles and storiform pattern along with scattered multinucleated giant cells. Hence, the histology was consistent with BFH to a considerable extent.

Immunohistochemical and histochemical studies have established the fact that BFH is positive for vimentin, CD68 and reticulin but immunonegative for glial fibrillary acidic protein and S100 [2,5,7,9]. In the current case, immunonegativity for CD1a and PR helped to exclude close mimickers of BFH, namely Langerhans cell histiocytosis and meningioma, respectively. Immunopositivity for vimentin and CD68 reinforced the histological diagnosis of BFH in this case.

These tumours characteristically lack nuclear pleomorphism and necrosis and have low mitotic activity [8,10] as in the present case. This reflects their benign biological behaviours. However, these tumours are notorious for recurrence.

Surgical resection remains the mainstay of treatment for patients with primary intracranial BFH. Excision of these tumours relieves the intracranial pressure and improves the overall clinical outcome in such patients. Fritz MA et al., have recommended the role of adjuvant radiation and chemotherapy in cases associated with bone invasion [3]. However, the present case was treated with surgical resection alone and the patient is asymptomatic after one year of follow-up.



[Table/Fig-1]: (a) Firm swelling over occipital region; (b) Computed tomography scan of brain: A large intracranial well-circumscribed expansile posterior fossa space-occupying lesion extending to suboccipital region destroying occipital bone; heterogeneous density and moderate enhancement measuring 65 [AP] × 54 [Tr] × 65 [V] mm; (c) Fine-needle aspiration cytology (FNAC): Moderate cellular smear showing oval-to-spindle-shaped fibroblasts along with histiocytes mostly in dispersed pattern (Leishman and Giemsa, 400×); (d) FNAC: Smear shows multinucleated giant cells (Leishman and Giemsa, 40×); (e) Gross features: Grayish-white solid tissue pieces altogether measuring 6 cm × 5 cm × 2 cm; (f) Histopathology: Bland-appearing fibroblastic cells and histiocytes arranged in interlacing fascicles and storiform pattern admixed with multinucleated giant cells (H and E, 10×); (g) Immunohistochemistry: Spindled stromal cells negative for progesterone receptor (DAB chromogen, 40×); (h) Immunohistochemistry: Spindled stromal cells positive for vimentin. (DAB chromogen, 40×); (i) Immunohistochemistry: Histiocytes and multinucleated giant cells positive for CD68 (DAB chromogen, 40×); (j) Reticulin stain showing no increase in reticulin fibrosis (reticulin, 10×).

The present case is, therefore, distinct in the following aspects: Location of the lesion, age at first presentation and asymptomatic on follow-up.

CONCLUSION

To conclude, primary intracranial BFH is an uncommon entity particularly in adults. Histopathological examination and immunohistochemistry help in establishing a definitive diagnosis. These tumours have a benign course and most patients have a long survival following surgical excision of the tumour. However, intracranial BFH recurs more frequently than its dermal counterpart. Hence, prompt diagnosis and meticulous follow-up must be instituted to improve survival rates in these patients. This further emphasizes the fact that there should be awareness and substantial knowledge among clinicians and pathologists regarding this rare entity for early recognition and prognostication of such cases. Since this lesion has an indolent clinical course, limited surgical resection can be curative, thereby lowering the morbidity and mortality rates.

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