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Case Report

Benign fibrous histiocytoma of the ethmoids in an infant

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Introduction

Benign fibrous histiocytoma (BFH) is a benign neoplasm of mesenchymal origin composed of histiocytes and fibroblasts.[1] It is commonly found as the cutaneous form in the sun exposed skin and rarely as a non-cutaneous lesion in the head and neck. We report the clinical and pathological aspects of a rare case of BFH of the ethmoid sinus in a nine-month-old male infant patient treated at our center.

Case report

A nine-month-old male infant patient was referred to our center with history of nasal blockage left side and protrusion of left eyeball of 2 months duration. The parents gave history of insidious onset, painless, progressive protrusion of the left eyeball of their son. The patient was a full term normal delivery of a non-consanguineous marriage with normal milestones. There was no history of nasal discharge, epistaxis or birth trauma. On clinical evaluation the patient had proptosis of left eye which was outwards and downward with restriction of ocular movements for adduction. Projection and perception of light was present in the left eye. On nasal endoscopy a large smooth swelling was seen arising from the lateral wall of nose obstructing the left nostril (Fig. 1A). Contrast enhanced CT scan revealed a 1.93 × 2.18 cm cystic lesion with well defined wall in the left ethmoids with erosion and scalloping of the lamina papyracea and pushing the left globe outwards. There was no evidence of erosion of the ethmoidal roof or extension into the anterior cranial fossa. The working diagnosis of congenital epidermal inclusion cyst

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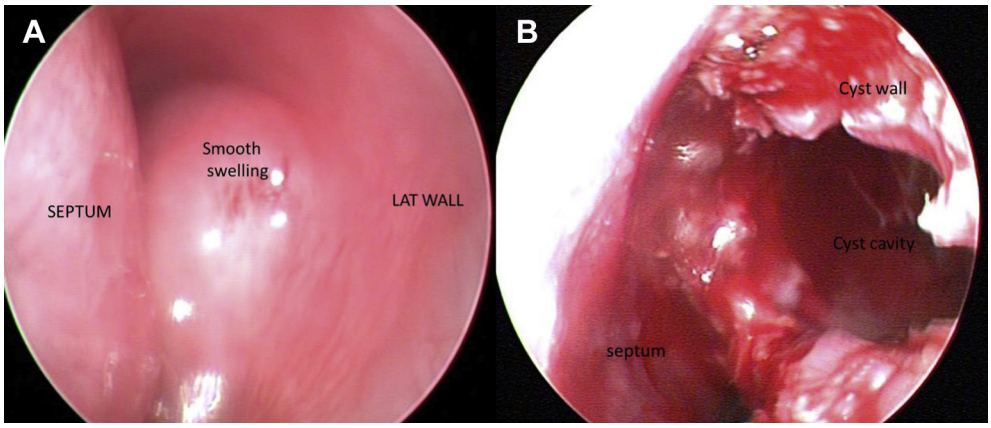

Fig. 1 – (A) Smooth swelling in the lateral nasal wall of left nasal cavity, (B) Endoscopic excision of cyst: cyst wall with thick walls visualized.

with pressure effects on the left orbit was reached and a plan of nasal endoscopic decompression of the cyst with biopsy was considered. On endoscopic excision, the cyst was found to be thick walled with clear fluid. The medial wall of cyst was removed and the specimen sent for histopathology (Fig. 1B). Postoperative period was uneventful with the patient having remarkable improvement in the left eye. The histopathology revealed diffuse areas containing fibroblast like spindle cells and histiocytes. The spindle shaped cells were focally arranged in streaming (storiform) pattern. There was no evidence of cellular pleomorphism, mitotic figures, nuclear atypia or presence of necrosis (Fig. 2). Immunohistochemistry was positive for Vimentin and negative for S100, CD 34, Desmin and SMA. The parents were counseled of the condition and were advised complete resection of the tumor, but they refused, saying that the patient had no complaints. The patient reported after 3 months with recurrence of the condition. On evaluation there was severe proptosis of left eye with swelling in the nasal cavity. CECT scan revealed 3 cm diameter predominantly cystic lesion with irregular specks of calcification in the left ethmoids displacing the optic nerve, left medial and superior rectus muscle causing proptosis with no intraocular extension (Fig. 3A). MRI revealed a well defined solid cystic lesion 4 cm × 3 cm × 4.5 cm (APx MLxCC) with multiple hyperintense cystic locules separated by thick septae epicentered in left ethmoid sinus, causing proptosis and stretching of optic nerve and extraocular muscles (Fig. 3B). The patient was taken up for open surgical procedure which included a lateral rhinotomy and medial maxillectomy (Fig. 4 A–D). Final histopathology showed BFH with clear margins. Postoperatively patient improved and is presently on regular review without recurrence since the past 2 years.

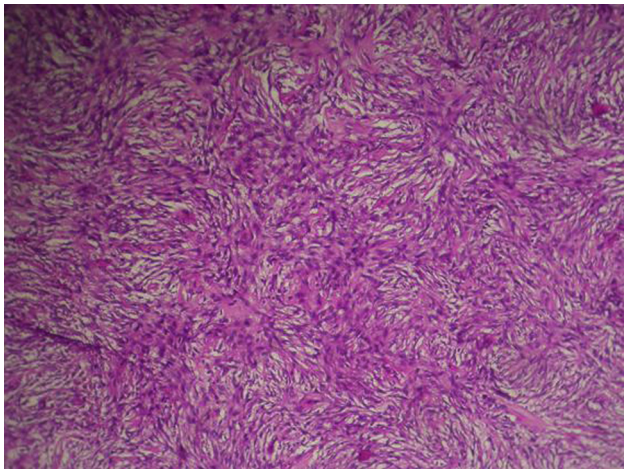

Fig. 2 – Spindle shaped fibroblasts in storiform pattern (H&E).

Discussion

Fibrous histiocytic tumors comprise a group of lesions with certain overlapping morphologic features but with variable origin and biologic behavior. Fibrous histiocytoma, a soft tissue tumor also referred to as fibrous xanthoma, dermatofibroma, xanthogranuloma and fibroxanthoma was first described as a separate clinical entity in the 1960.[2,3] BFH is a common benign neoplasm found in the sun exposed dermis and superficial subcutaneous tissue of the extremities, but is also found less frequently in the deep soft tissue and occasionally in parenchymal organs.[2–4] Non-cutaneous BFH represents approximately 1% of all benign Fibrous Histiocytic lesions,[2] and most frequently occurs in the soft tissues in the lower extremities (50%), less frequently in the upper extremities (20%), retroperitoneum (20%).[4] BFH in the deep soft tissues of head and neck is rare with a few cases reported in the orbit, oral cavity, mandible, paranasal sinus,[2,3,5] larynx and trachea. BFH affects adult patients in the fourth and fifth decade with no specific sex predilection. In the nose and paranasal sinus, twelve cases have been described in the English literature of which four were of the ethmoid sinus.[5] To the best of our knowledge, this is the fifth reported case of benign fibrous histiocytoma involving the ethmoid sinus.[6]

The etiology of BFH is unclear and its biologic nature is reported as both neoplastic and reactive. The patients with cutaneous BFH usually have a history of sun exposure, trauma, or chronic infection, suggesting that it is a reactive disease.[7] Various theories of histogenesis include origin from primitive undifferentiated mesenchymal cells by electron