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Case Report

Benign fibrous histiocytoma of the ethmoids in an infant

Lt Col Satish Nair^{a,*}, Brig W.V.B.S. Ramalingam, (Retd)^b,
Brig Reena Bharadwaj^c, Col A.V. Ramesh^d, Akshat Malik^e,
Sharad Mohan^f

^a Classified Specialist (ENT), Army Hospital (R&R), Delhi Cantt, India

^b Professor & Consultant (ENT), BLK Super Speciality Hospital, Delhi, India

^c Professor & Consultant (Pathology), Dy DGAFMS (Pensions), O/o DGAFMS, Delhi 110001, India

^d Senior Advisor (ENT), Command Hospital (Central Command), Lucknow, India

^e ENT Surgeon, Tata Memorial Hospital, Mumbai, India

^f ENT Surgeon, Gold Field Institute of Medical Sciences & Research Faridabad, Haryana, India

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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

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

* Corresponding author. Tel.: +91 8130667567.

E-mail address: aachku@yahoo.com (S. Nair).

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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

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

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.

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

Fig. 3 – (A) CT scan showing loculated cystic lesion with thick walls in the left ethmoids pushing the left globe outwards. (B) MRI revealed a cystic lesion in the ethmoids with pressure on extraocular muscles and globe.

microscopy, histiocytic origin by tissue culture and fibroblastic origin by cell marker studies.^{3,4} Cytogenetic reports of clonal chromosomal abnormalities in BFH have supported its neoplastic origin.¹

Cutaneous BFH is usually asymptomatic and present as painless well demarcated solitary subcutaneous mass. In the non-cutaneous BFH, clinical features depend on the site involved and the pressure symptoms produced due to the rapidly enlarging tumor mass.³ Nasal obstruction, epistaxis, facial asymmetry, painless loosening of teeth and proptosis has been described in cases of BFH of the paranasal sinus.⁶

The diagnostic dilemma is due to the cellular origin and histopathological similarities with other sarcomas. The diagnosis of BFH is challenging and rests on characteristic histopathologic and immunohistochemical features.⁴ BFH is composed of a biphasic cell population of histiocytes and fibroblasts.¹ Histologically it is predominantly composed of fibroblastic cells arranged in a storiform pattern or interlacing fascicles, supported in a collagenous stroma and admixed with varying numbers of histiocytes, foam cells, siderophages and inflammatory cells. Immunohistochemical and electron microscopic studies are non-specific and provide confirmatory evidence of fibrous histiocytoma by excluding other

Fig. 4 – (A) Severe proptosis of left eye. (B) Open surgical excision: lateral rhinotomy approach with the visualization of the tumor. (C) Open surgical excision: complete excision of tumor. (D) Specimen of the tumor.

diagnoses.⁴ Tumor cells are usually positive for Vimentin and negative for lysozyme, desmin, S 100, keratin and Factor XIIIa.⁸ Due to the lack of specific markers for fibrohistiocytic lesions, the diagnosis of BFH is generally based on the absence of markers for cells of other lineages. Immunohistochemical staining and ultrastructural examination of the tumors and cell lines derived from them has revealed features of myoblastic and histiocytic differentiation as evidence of mesenchymal origin. Immunostaining for CD68 can be found in any tumor-containing lysosomal granules or phagolysosomes as in our case.⁴ Factor XIIIa has occasionally been reported for BFH. Among the various histological variants of BFH, 20% are found in the head and neck with close similarity to sarcoma and characteristics of rapid growth and higher recurrences.⁸ Malignant transformation and distant metastasis of BFH is rare.

CT scan and MRI are important radiological investigations, which help in defining the extent of lesion and involvement of adjacent structures. CT scan is useful in identifying the extent of lesion as well as the bony anatomy whereas MRI is superior in identifying the intracranial extension and involvement of brain parenchyma.

In a neonate, the differential diagnoses for various mass lesions of the paranasal sinus are encephalocele, epidermal/dermal inclusion cyst, hemangioma, glioma and lymphangioma.⁹

Due to their characteristics of local invasion and recurrence, the treatment of choice is wide local excision, which ensures a cure with no recurrence. Incomplete excision or enucleation results in significant recurrence as was seen in our case where initial marsupialization of cyst led to aggressive growth of the tumor which was later removed completely by open surgical procedure with no evidence of recurrence.

Conclusion

Benign fibrous histiocytoma is a rare benign tumor with characteristic histopathological features, which can be

managed surgically with complete cure. The aim of the report was to highlight BFH as a differential diagnosis of nasal mass in neonates and its excellent prognosis with excision with pathologically clear margins.

Conflicts of interest

All authors have none to declare.

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