

Nasal Chondromesenchymal Hamartoma- A Rare Case Report

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Abstract

Case Report

Nasal hamartomas are very rare lesions occurring in the early childhood. We present a case of an eight year old female patient with nasal blockade and facial deformity, with radiological suspicion of a soft tissue density lesion in right nasal cavity and right Ethmoid and frontal sinuses. Histopathology revealed a rare benign hamartoma - Nasal Chondromesenchymal hamartoma.

Keywords: Nasal hamartoma, Mesenchymal hamartoma.

Key Message: Nasal chondromesenchymal hamartoma, though rare is interesting entity. Histopathology diagnosis is the key confirmatory tool for management.

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INTRODUCTION

Nasal Chondromesenchymal Hamartoma (NCMH) is a rare benign tumour like growth seen in the nasal and paranasal cavities. McDermott et al., suggested the term 'nasal chondromesenchymal hamartoma' as a distinct pathologic entity that showed the characteristic histologic features of mixed stromal and chondroid tissue in various proportions¹. Though seen in all age groups it is predominantly seen in infants and more common in males [1, 2]. Its incidence is very rare with only over 60 cases reported worldwide. Though benign, it is a locally aggressive condition which can lead to destruction of adjacent tissues [3].

CASE HISTORY

Eight year old female child presented with history of Nasal blockade of four months duration. Also the child developed deformity of the right side of the face. A clinical examination revealed a mass completely occluding the right nasal cavity. CT imaging identified soft tissue density lesion with hyper densities within the Right nasal cavity, Ethmoidal air cells & right Frontal sinus with expansion of nasal cavity, ethmoidal air cells with thinning of bony walls & displacing the right eye globe laterally, with erosion of base of skull in the anterior cranial fossa (Fig-1a). A differential diagnosis

of Polyp or a mass lesion was opined. Intra operatively the lesion was found completely filling the right nasal cavity and extending into ethmoidal and frontal sinuses. It was a firm, pale, globular mass. The mass was removed in toto and sent for histopathology examination. Gross examination revealed multiple grey white, fleshy, soft tissue bits, largest bit measuring 3x2.5x1cm, smallest 0.8x0.5x0.3 cm (Fig 1b-c). The tissue was routinely fixed, processed and stained with Hematoxylin and Eosin staining.

On light microscopic examination, the sections showed solid and cystic areas. There is lining of respiratory epithelium with a submucosal tumour showing with dense cellular areas and loose myxoid areas. There are irregular, small islands of hyaline and fibrocartilage (Fig-2a-b) amidst sheets of benign looking plump spindle-cells (Fig-2c-d) with rare mitosis and focal osteoclastic type of giant cells and calcifications (Fig-3a-d). Many dilated congested vascular spaces were seen. An additional immunohistochemical staining for Ki67 show low proliferative index (1%). Based on the histomorphological findings a diagnosis of Nasal Chondromesenchymal Hamartoma (NCMH) was established.

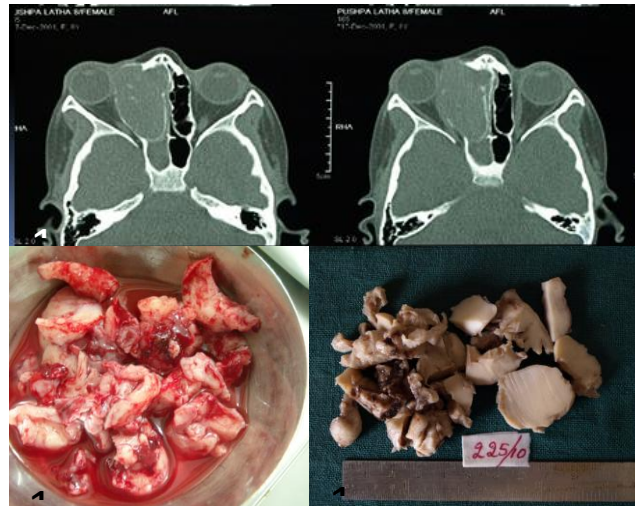


Fig 1a CT: a mass lesion in right nasal cavity. 1b & 1c Unfixed & Fixed gross specimen

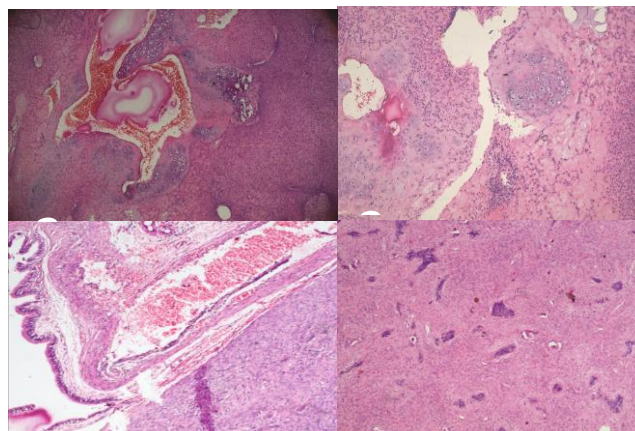


Fig 2a,b: Scanner & low power view of cartilaginous islands. 2c: Blood filled cystic spaces. 2d: Solid component composed of stromal cells

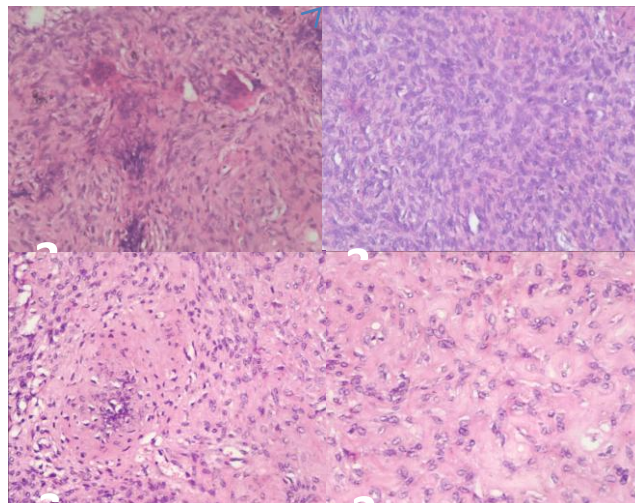


Fig-3a: High power view of foci with Osteoclastic type of giant cells. 3b-d: High power view of areas showing bland spindle cells with collagenous matrix

DISCUSSION

Hamartomas are non tumourous malformation of tissues, composed of abnormal organization of native structures. Nasal Hamartomas are rare and are either mesenchymal or epithelial in nature. Mesenchymal hamartomas are more common and are variously known

as chondroid hamartoma, chondromesenchymal, angiomatous, lipomatous etc. based on the overgrowth of specific mesenchymal element [2]. Epithelial hamartomas are very rare and composed of glandular and epithelial component along with mesenchymal component [3].

The term Nasal Chondromesenchymal hamartoma (NCMH) was first coined by McDermott *et al.*, in 1998 after studying a series of cases [1]. It was classically described as distinct pathologic entity that showed the characteristic histologic features of mixed stromal and chondroid tissue in various proportions, reminiscent of the mesenchymal hamartoma of the chest wall. It is thought to be a congenital lesion with, most cases reported during infancy [4] most commonly seen in children less than 3 months. The mean age of occurrence being 9.6 yrs [9]. The most common site is with in nasal cavities and rarely in paranasal sinuses. The cause of disease is not yet known. A strong genetic predisposition has been suggested by co-existence of Pleuropulmonary blastoma [1] and this was later confirmed by Jhonson *et al.*, [5]. Vatsa *et al.*, in a cohort study found specific germline mutations of DICER1 gene to be associated with these malformations [8]. There are studies showing association of NCMH with Chronic sinusitis especially when diagnosed in adulthood [7, 8].

Till date only 66 cases of hamartomas have been reported and clinical manifestations depend on the nature and amount of obstruction like nasal obstruction, feeding difficulties, middle ear effusions, epistaxis etc [10]. The histopathological examination of tissues is key to diagnosis as radiological findings are not confirmatory. The benign nature can only be established by microscopic findings of benign cartilaginous and mesenchymal stromal component. A thorough sampling of tissue is needed to exclude areas with malignant transformation. The IHC markers which can be useful are smooth muscle actin (SMA), S-100, Vimentin, KP-1 and Leu which are positive markers and negative markers include cytokeratin, Epithelial Membrane Antigen (EMA) and desmin [3]. One of the close differentials include Chondrosarcomas, though known to be rare in sinonasal tracts. Histologically NCMH has islands of well demarcated mature and immature hyaline cartilage cartilage and benign looking fibroblasts with no or very rare mitosis and atypia. These cases have a good prognosis with low recurrence rates [3].

In our report, an eight year old female child who presented with nasal obstruction was diagnosed on CT findings as mass lesion of nasal cavity. The histology revealed the hamartomatous nature of lesion with chondroid, mesenchymal and vascular component. The post-operative period was uneventful and the patient did not have any recurrence even three years after the excision.

The main treatment modality is surgical. Though open surgery is the treatment of choice, many recent studies showed equal success with minimal invasive endoscopic approaches. The main aim is to

completely remove the lesion to avoid recurrence [11]. In conclusion, awareness about this interesting though rare entity, is essential to provide proper and complete treatment.

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