ABSTRACT

Nasal Chondromesenchymal Hamartoma (NCMH) was first described by McDermott et al., in 1998. It is a very uncommon benign mass arising in the sinonasal area. Forty-eight cases of NCMH have been reported including 6 cases of adult presentation. It mostly occurs in younger age group with median age of 7 years. In this article, we report a rare case of Nasal Chondromesenchymal hamartoma (NCMH) in 17-years-old male who presented to us with persistent headache. The tumor was surgically excised in toto by using lateral rhinotomy approach with no recurrence.

KEY WORDS: Nasal Chondromesenchymal Hamartoma, Sinonasal, Lateral Rhinotomy.

HOW TO CITE THIS:

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INTRODUCTION

Hamartomas are very rare sinonasal lesions, usually identified in children. Nasal Chondromesenchymal Hamartoma (NCMH) is a term coined by McDermott et al., in 1998. It is a very uncommon benign sinonasal pathology of unknown etiology with characteristic clinicopathological features. 48 cases have been reported in literature. NCMH’s have a mixed morphology including mesenchymal and cartilaginous components. It usually occurs in children with a median age of 7 years. Although usually asymptomatic, symptoms when occur depends on the tumor size and location and commonly include nasal symptoms like nasal obstruction, rhinorrhea, epistaxis, anosmia, headache; ophthalmic symptoms like epiphora, diplopia, proptosis, decreased vision, hypotropia and intra-oral symptoms.

We report a rare case of NCMH of a 17-year-male, the first case from Pakistan, with a large mass in the left nasal cavity with telecanthus which presented to us due to persistent headache.

CASE REPORT

A 17-years-old male, presented in ENT outpatients with swelling over the nasal bridge and nasal obstruction since birth which increased insidiously causing facial disfigurement, till 6 months before presentation, when patient developed persistent throbbing headache. Examination revealed a bony swelling on nasal bridge and medial to the left orbit with tele-canthus (Figure - 1). Anterior rhinoscopy, revealed a firm mass on the left side, pushing the septum towards the right. The differential diagnosis considered were osteoma, chondroma and fibrous dysplasia.

CT scan revealed a well-defined rounded osseous and soft tissue density lesion involving the left ethmoid sinuses and left nasal cavity measuring 4.8×4.0×4.3 cm with irregular ossification (Figure - 2). It had a broad-based attachment to the ethmoid with expansion of the left nasal cavity with septal deviation towards the right and medial wall of left maxilla and orbit towards left.

Intranasal punch biopsy revealed a Chondroma. Surgical removal under general anesthesia through a lateral rhinotomy approach with extended Moure’s incision revealed a well-defined mass with attachment to ethmoids including cribriform plate, which was freed to remove the mass in toto. It was 6x5x4 cm, well encapsulated, hard and giving a gritty feel (Figure - 3). The Cavity was packed trans-nasally and wound closed in layers. Post operatively patient suffered from conjunctivitis, excessive crusting and epiphora in the first month, which gradually improved. Follow up was maintained for 2 years with no recurrence.

Histo-pathology (Figure - 4) revealed well delineated nodules of hyaline cartilage within the stroma showing variable cellularity with giant cells of osteoclastic appearance. Stroma also showed erythrocytes filled spaces. No significant pleomorphism and mitotic activity seen, however some nodules were ossified. Lining epithelium was normal respiratory epithelium. Thus, the diagnosis of Nasal Chondromesenchymal Hamartoma was made.
DISCUSSION

NCMH is uncommon, slow growing, benign sinonasal pathology with 48 reported cases including 6 cases of adult presentation. This is the first case of NCMH being reported from Pakistan. This lesion is mostly reported in younger children (median age 7 years). This is locally destructive with only one reported case of malignant transformation. These lesions have delayed presentation being slow growing and sometimes asymptomatic in infancy, which was true in our case. It grew insidiously, over 17 years, and ultimately presented with persistent headache.

Presentation depends on size and location of the tumor. Usually it is asymptomatic, but may present with nasal obstruction, rhinorrhea, epistaxis, anosmia, headache, epiphora, diplopia, proptosis, decreased vision, hypotropia, and intra-oral symptoms.

NCMH have a mixed morphology. Mesenchymal hamartomas being more common. Aetiology and origin of NCMH is not well understood. It may be caused by an underlying genetic predisposition. Some consider it a variant of chest wall mesenchymal hamartoma, others link it to blastoma of pleuropulmonary (PPB) origin. Microscopy reveals a variety of
mesenchymal components in follicles including irregular islands of mature and immature hyaline cartilaginous components with bi-nucleated chondrocytes. Stroma shows well delineated cartilaginous islands with myxoid background with a population of relatively bland and compact spindle cells with variable cellularity. Immune reactivity may show positive for smooth muscle actin, KP-1, Leu-7, S-100, vimentin, and negative for epithelial membrane antigen, cytokeratin, and desmin. CT Scan, reveals an un-capsulated lesion with some cystic areas and calcifications. This was true in our case. On MRI, it is homogeneously iso-intense to the cerebral cortex on T1 images and heterogeneously hyper-intense on T2. This is attributed to abundance of myxoid stroma with less number of cells. Complete Surgical Excision is the mainstay of treatment and approach depends on the lesion's site and size. Endoscopic surgery can be useful for tumors restricted to the nasal cavity, however, infiltrative nature, makes it difficult to get a tumor free margin. Tumor recurrence occurs, especially in cases with inadequate resections or where small deposits of tumor are left behind. We were successful in doing a complete resection of tumor without any recurrence. Radiotherapy and combination chemotherapy are other modes of treatment reported by Shet et al.

Therefore, Careful detailed preoperative assessment including imaging is essential to see the extent of lesion and status of the neighboring structures. Magnetic resonance imaging may be required in aggressive cases with suspicion of malignancy and to see involvement of important neighboring structures. Complete Surgical Excision is the mainstay of treatment.

CONCLUSION

NCMH may be rarely encountered when dealing with masses in the sinonasal area and should be considered in differential diagnosis.

Contribution of author:
Ahmed J: Manuscript writing
Saqulain G: Literature Review
Shahzad J: Data Collection, Histopathology slides

Disclaimer: None.
Conflict of Interest: None.
Source of Funding: None.

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