Nasal Chondromesenchymal Hamartoma: A Rare Case Report

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1. Case Report

A 9 months old male child presented to the outpatient department with difficulty in breathing and nasal obstruction since 7 months. The patient also presented with recent onset feeding difficulties. No significant antenatal or perinatal history was present. The patient had no other significant medical history.

On nasal endoscopy, a smooth well circumscibed mass was seen completely obscuring the left nasal cavity and causing deviation of lateral nasal wall. Ophthalmologic examination was normal.

Laboratory investigations were within normal limits.

On CECT PNS, a large expansile non-enhancing soft tissue density lesion measuring 41x25x30mm (APxTRxCC) with multiple coarse calcifications within was seen occupying the left nasal cavity. The lesion was causing severe bowing of the nasal septum towards right causing severe obliteration of right nasal cavity. The lesion extended into the region of left posterior ethmoidal air cells with minimal extension into the nasopharynx. The cribriform plate appeared to be eroded at few places. The lesion was also causing bowing and remodelling of left lamina papyracea and inferior wall of left orbit, however no intra-orbital extension of the lesion was present. Scalloping of alveolar recesses of left upper incisor teeth, bilateral pterygopalatine and infratemporal fossae was noted.

On MRI PNS (Plain+Contrast) revealed an ovoid encapsulated mass occupying the left nasal cavity. It showed homogeneous high signal on T2 weighted images. Few T2 hypointensities were noted within it suggestive of calcifications. No evidence of intra-oral, intra-orbital or intra-cranial extension of the lesion was noted.

A biopsy from the mass sent for histopathology examination revealed lobules of mature cartilage along with fibromyxoid
stroma infiltrating into the surrounding tissue. Few of the bits were lined by ciliated pseudostratified columnar epithelium. Surrounding tissue showed moderate lymphocytic inflammatory infiltrate. On histopathology, diagnosis of nasal chondromesenchymal hamartoma was confirmed.

Complete excision of the tumor was obtained.

2. Discussion

Hamartoma refers to a mass of disorganized overgrowth of mature specialized cells and tissue indigenous to the organ in which it occurs. Nasal hamartomas may be composed predominantly of mesenchymal or epithelial tissue. The mesenchymal hamartomas can be chondroid or chondromesenchymal. Nasal chondromesenchymal hamartoma (NCMH) is a rare benign tumor that occurs predominantly in infants. They usually present as polypoidal lesions shortly after birth. Most of the cases occur in infancy, however few cases have been reported in adolescents and even in adult patients.

The presentation of NCMH depends on the size and site of tumor and also on the involvement of adjacent structures. Infants usually present with respiratory and feeding difficulties, rhinorrhea, epistaxis and otitis media. The symptoms also depend on whether the lesion has intraorbital and intra-cranial extension.

Contrast enhanced CT and MRI are the basic preoperative modalities. Detailed characteristics of the lesion like solid cystic components, calcifications and enhancement are indentified on CT and MRI. They also provide information about the involvement of paranasal sinuses, bony remodelling, intracranial and intraorbital extension. There is a broad list of differential diagnosis which include benign paediatric nasal masses like nasal glioma, ossifying fibroma, giant cell reparative granuloma and aneurysmal bone cyst. NCMH can also mimic malignant paediatric tumors like rhabdomyosarcoma, esthesioneuroblastoma and chondrosarcoma.

The standard gold for diagnosis of NCMH is histopathology. The most characteristic appearance is of irregular islands of mature and immature hyaline cartilage with occasional binucleated chondrocytes. Immunohistochemical staining demonstrate vimentin and SMA (smooth muscle actin) in the stromal mesenchymal components and S-100 positivity in both the spindle areas and mature cartilage.

Management of NCMH is complete excision of the mass. There are no recurrences as long as the mass is completely excised. When the mass is localised to nasal cavity, endoscopic resection is preferred. With intracranial or intraorbital extension external approach like mid-facial degloving is recommended.

References
