CASE REPORT

Long term outcome of chondromesenchymal hamartoma of sinonasal cavity: a rare entity

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Abstract

Nasal chondromesenchymal hamartoma in the sinonasal cavity is an unusual entity mostly found in young infants and children. We present the case of a nasal chondromesenchymal hamartoma in a young female. NCMH has a favourable outcome that shows neither recurrence nor any malignant behaviour.

Keywords: Nasal Chondromesenchymal Hamartoma, Malignant, Recurrence.

DOI: 10.47391/JPMA.4198

Submission completion date: 13-11-2021
Acceptance date: 13-10-2022

Introduction

Nasal chondromesenchymal hamartomas are uncommon tumours that present as a polypoidal mass in infancy and childhood.[1] They are a mixture of mesenchymal tissues with different proportions of stromal and chrondroid elements. It is composed of nodules of cartilage with dense cellular variation and mature chondrocytes, focal giant like osteoclastic cells in the stroma and spaces filled with erythrocytes.[2] It mimics hamartoma of chest wall due to similar histological findings[3].

Patients with NCMH present with complaints that are based on location of the mass or lesion. In the nasal cavity or paranasal sinuses, it causes compression of local structures with symptoms ranging from nasal obstruction to dental pain. Some patients also complain of visual impairment and facial pain.[3] It can also present as an intranasal mass or facial swelling in infants and children and very rarely in adults.[4-6]

We report a case of a young female who presented with right-sided nasal blockage for a year and nasal endoscopy showed a polypoidal mass on the floor of nasal cavity involving osteomeatal complex. Her biopsy revealed NCMH for which she underwent Endoscopic removal of the tumour. This case perhaps is the first reported case in our country and according to literature fourth reported case presented in an adult female.

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Case Report

A young female of age 25 years presented in the outpatient department of Liaquat National Hospital, Karachi – a tertiary healthcare center, in January 2019 with complaint of right nasal blockage for the past one year. Patient reported a progressive increase in severity of blockage with postnasal dripping and rhinorrhea.

She also noted swelling of her right eye and lacrimation for a period of one month but had no visual disturbances or associated headache and anosmia. Extraocular movements were normal and vision was intact. Anterior rhinoscopy showed polypoidal swelling behind the inferior turbinate of right nasal cavity and deviated nasal septum on the opposite side with reduced right side patency. Fibreoptic nasal endoscopy was done which showed a polypoidal mass present on the floor of nasal cavity extending to right osteomeatal complex with bony septum deviating towards the left side. Nasopharynx and left nasal cavity were normal.

Computed tomography scan showed a large, polypoidal, expansile mass involving the right nasal cavity. It measured 5.8 x 6.5 cm in size with opacification of right maxillary sinus and erosion of adjacent nasal turbinates and causing deviation of nasal septum towards left (Figure 1). The mass was causing significant pressure

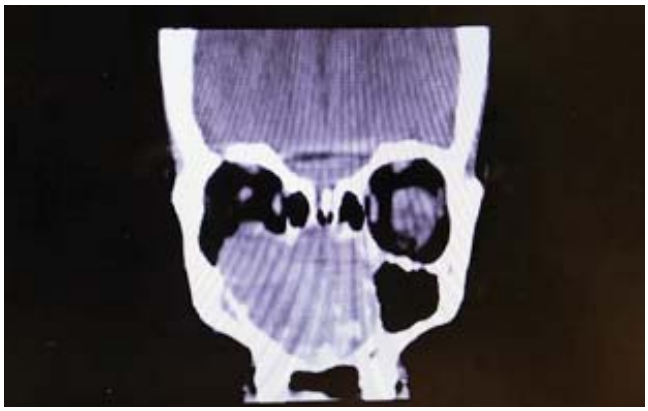

Figure-1: Coronal and axial view showing heterogenous mass in right nasal cavit and right maxillary sinus causing deviation of nasal septum towards left.

erosion and ballooning of bones of the nasal cavity extending superiorly into ethmoidal sinuses. Laterally it was also causing pressure erosion of medial wall of right maxillary sinus and superior bulge on the floor of right orbit.

Biopsy revealed features that were consistent with NCMH and endoscopic removal of the tumour under general anaesthesia was planned. The patient underwent endoscopic removal of tumour from right maxillary sinus with medial maxillectomy. Endoscopy showed necrotic tissue filling the right nasal cavity and right maxillary sinus eroding its medial wall. Floor of orbit and lamina papyracea were absent and posterior part of septum was also absent. Right uncinectomy was done and the tumour was removed from anterior and lateral wall of right maxillary sinus, floor of nasal cavity and was also peeled off from the posterior part of the septum. A small part of the tumour adherent to the eyeball was removed inferiorly and sent for histopathology. The nasal cavity was then packed with non-absorbable Merocel after achieving good haemostasis. Histopathological examination revealed multiple fragments of tissue exhibiting a lesion composed of cartilaginous tissue along with focal area containing spindle cells. Background stroma appeared loose and focally myxoid with scattered blood filled cystic spaces. The findings were in favour of the diagnosis of chondromyxoid hamartoma (Figure 2). Postoperatively, the patient remained stable and was discharged the next day. She was followed in outpatient clinic at regular intervals for up to 2 years. No recurrence was found at a 2-year follow-up.

Discussion

The classification of head and neck tumours according to (WHO) World Health Organization has accepted NCMH as a new entity of benign tumours of bone and cartilage because of its distinct characteristics morphologically as well as clinically[1].

There is only one case reported in literature with malignant transformation of NCMH in adults[1]. Recurrence after surgery has been reported in patients with incomplete surgical removal or microscopic deposits of a residual tumour. Radiation and chemotherapy have been given for lesions that are not completely resected however limited clinical experience has made it difficult to conclude how to treat residual or unresectable tumours.[1]

The gold standard treatment for nasal chondromesenchymal hamartoma is its surgical excision. If the lesion is confined to the nose or sinuses it is removed with the help of an endoscope. If the tumour is extending intracranially or intraorbitally, an external approach is considered[2].

Mason in his study has revealed that a nasal chondromesenchymal hamartoma can present at any age from birth to 69 years with a mean of 9.6 years.[3]

The ratio of male to female is 2.2:1. The common sites are nose and sinuses from where it can extend to orbit and through the cribriform plate to cranium due to its aggressive behaviour.[3] The presenting symptoms are nasal obstruction, mass in nasal cavity, or eye symptoms.

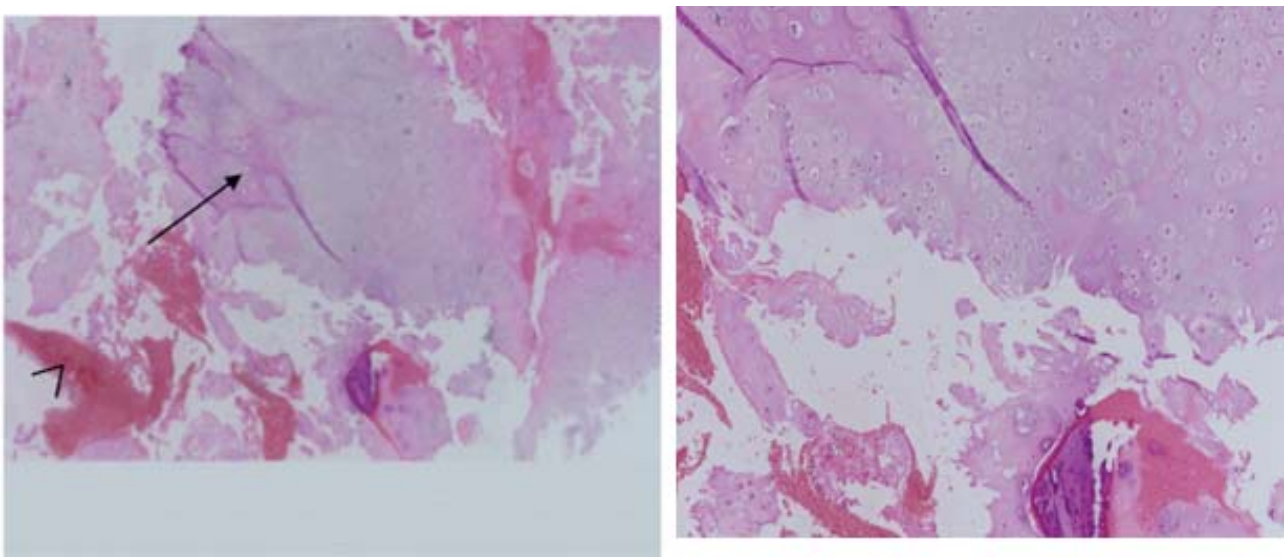

Figure-2: Well demarcated mature cartilaginous tissue (black arrow). Blood filled cystic spaces can also be appreciated (Arrow head) resembling aneurysmal bone cyst-like formations. Stroma appears loose and mxyoid.

Signs include strabismus, hypertelorism, extropia, enophthalmus, proptosis and ophthlmoplegia[3].

It is either a non-neoplastic malformation or an inborn developmental error. McDermott et al. in 1998 was the one who noticed NCMH as a different entity both clinically and pathologically which has given different names in the past as ''chondroid hamartoma'', ''mesenchymoma'' ''nasal hamartoma''.[4]

Chondromesenchymal hamartomas are benign predominantly but because of their aggressive behaviour and locally destructive nature, they are being mistaken as malignant lesions. They are slow growing tumours with delayed presentation[4]. Around 60 cases have been reported up till now[5]

The diagnosis of chondromesenchymal hamartoma is confirmed from histopathologic findings. However, CT and MRI are reliable imaging choices for diagnosis to know the site of the tumour, involvement of adjacent structures, and to determine the extent of mass and help in planning surgery[6,7].

It is an overgrowth of multiple aberrant cells, indigenous to the site in which it occurs. They can be epithelial, mesenchymal, or mixed. The hamartoma that are present in the sinonasal region contain spindle cells, collagen fibres and heterogeneous islands of osseous and chondroid tissue.[8]

Chondromesenchymal hamartoma due to germline or somatic mutations of DICER1 gene has an association with the pleuropulmonary blastoma tumour disorder in children[9]. Our patient being the 61st, out of total reported cases in which few patients were adults. It is difficult to distinguish the origin of chondromesenchymal hamartoma in adult population, but it is hypothesised that it derives from embryologic cell rests[10].

Symptoms associated with chondromesenchymal hamartoma in infants and children are respiratory and feeding difficulties, bleeding per nose, watery nasal discharge, visual impairment, and middle ear infection. Orbital involvement of the tumour can result in enophthalmos, proptosis, or restricted eye movement[8]. In our patient symptoms were rhinorrhea, nasal blockage, postnasal dripping and right cheek swelling. Ophthalmic signs included asymmetry of both eye globes and epiphora.

According to histopathology, these lesions are similar to other mesenchymal hamartomas, and consist of chondroid tissue with areas of calcification, and mesenchymal tissues such as spindle cells and myxoid stroma.

Wang et al in his study said that calcification is a main diagnostic feature which can distinguish it on imaging from other nasal masses. In his study, CT scan of the lesion showed a well-defined enhancing soft tissue mass with hypointensity on T1 images and hyperintensity on T2-weighted MRI images, with enhancement in a heterogeneous fashion; 50% showed calcification or cystic areas within the lesion.[7] In our study there were punctate areas of calcification and pressure erosion and ballooning of maxillary sinus, nasal cavities and hard palate. In one of the case series of chondromesenchymal hamartoma, bony remodeling and erosion were also reported to be present in ethmoid sinus with intracranial extension in 53% cases[6] 50% had internal calcifications, 40% had cystic components, and 67% showed enhancement.[7]

Conclusion

Nasal chondromesenchymal hamartoma is a unique paediatric tumour which is rarely seen in adults. We report the 4th case of hamartoma in an adult female who presented with complaints of nasal blockage, right eye swelling and lacrimation. Endoscopic medial maxillectomy and complete surgical resection was done. We followed the patient for a total period of 2 years to see the long term outcome of this benign tumour and signs and symptoms of recurrence. NCMH has a good prognosis if resected completely. It can mimic symptoms of chronic rhinosinustis hence histopathology reporting helped in reaching the final diagnosis.

Informed Consent: Written informed consent was obtained from the patient for publishing her case report..

Disclaimer: None.

Conflict of Interest: The authors declared no potential conflicts of interest with respect to the research, authorship, and publication of this article.

Source of Funding: The authors declared that this study has received no financial support.

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