Nasal Chondromesenchymal Hamartoma: A Rare Benign Lesion in Adult Female

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ABSTRACT

Nasal chondromesenchymal hamartoma (NCMH) is a rare benign lesion of the sinonasal tract in children and adolescent with orbital involvement. NCMH is histologically composed of nodules of cartilage with cellular density variation and maturation of the chondrocytes, a myxoid to spindle cell stroma, focal osteoclast-like giant cells in the stroma, and erythrocyte- filled spaces. This lesion may present with a destructive pattern on imaging, highly suggestive of malignancy. Total endoscopic resection is the choice of treatment nowadays, however incomplete excision could result in tumor recurrence but rare.

Keywords: Nasal chondromesenchymal hamartoma

INTRODUCTION

Nasal chondromesenchymal hamartoma (NCMH) is an extremely rare, benign sinonasal tract tumor. In the English literature, 47 cases have been documented, with the vast majority of these presentations occurring in newborns and young children, frequently under the age of one. NCMHs have a mixed morphological structure that is mostly composed of mesenchymal and cartilaginous components. NCMH patients report with symptoms that vary depending on whether the tumor is located in the nasal cavity or paranasal sinuses and whether it compresses local structures. Nasal blockage, vision impairment, and face and dental pain are among the symptoms. There have only been six cases of adult NCMH presentation to date[1-3].

The nasal chondrome Senchymal hamartoma (NCMH) is an uncommon benign tumor of the sinonasal tract in children that can invade the orbit and skull base. This tumor is characterized by a heterogeneous mixture of spindle cells, collagen fibers, and irregular islands of osseous and chondroid tissue[4].

We discovered 56 reported instances of NCMH in single reports and small series after reviewing the literature. The most prevalent symptoms are difficulty breast-feeding and nasal blockage, and the clinical presentation is non-specific. Because the vast majority of NCMH patients are discovered at a young age, the lesion is thought to be congenital[5,6].

Endoscopic excision of NCMH is possible. Although incomplete excision may result in tumor recurrence, only a few studies have reported NCMH relapse.

NCMHs (nasal chondromesenchymal hamartomas) are extremely rare tumors. In the international literature, only 21 cases have been reported. Polypoid lesions commonly appear shortly after birth. They are benign tumors, and total excision is the preferred treatment. NCMH has also been referred to as "chondroid hamartoma," "mesenchymoma," "nasal hamartoma," and other names in the literature. The term "nasal chondromesenchymal hamartoma" was first proposed in 1998 as a distinct pathologic entity. The tumor is made up of a variety of mesenchymal components[7,8]. The properties of histopathology, computed tomography (CT), and magnetic resonance imaging (MRI) have been thoroughly documented. Only three cases of digital subtraction angiography have been described in the radiology literature[9,10]. Here we presented A 42 year old Saudi female patient known case of hypertension came to our outpatient clinic complaining of slowly growing midline mass for 2 years. It is associated with nasal obstruction, loss of smelling and pain.

CASE PRESENTATION

A 42 years old Saudi female patient known case of hypertension came to our outpatient clinic complaining of slowly growing midline mass for 2 years. It is associated with nasal obstruction, loss of smelling and pain.

History: No history of nasal bleeding or discharge, no other ENT complaints was indicated. On examination on midline nasal mass, hard, non tender noted (Figure 1). On anterior rhinoscopy bilateral nasal obstruction and hypertrophic inferior turbinates. CT PNS with contrast (Figure 2) findings left maxillary antrum shows soft tissue with convex inner border representing left maxillary antral polyps or retention cyst, the ethmoidal cells show opacification of the third minus variance show opacification of bony rales. The bony walls of paranasal sinuses are intact. The osteomeatal complex are free bilaterally. Under general anesthesia examination done showed thickened nasal septum, multiple layers of cartilaginous septum and thickened cartilage of nasal dorsum. Multiple biopsies taking from septum and dorsum mass.

Histopathology: Histopathology sections show Islands of cartilage showing focal degenerative change and mesenchymal tissue forming storiform pattern and infiltrated by chronic inflammatory cells.

Histopathological examination revealed cartilage and aneurysmal bone coated in stratified squamous epithelium with keratinization in the nodule. A haematoxylin and eosin stain was used to make a histopathological diagnosis. These findings were compatible with a nasal chondromesenchymal hamartoma diagnosis. Further nose, septal cartilage, right and left dorsum, incisional biopsies:- Most consistent with chondromesenchymal hamartoma.

Follow-up and Outcome: The patient was followed up in clinic and was discharged after 2 years having exhibited no symptoms of recurrence. A telephone interview was also conducted 4 years after the operation, and the patient reported no recurrence of the nasal tumour.

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He affirmed that he had no post-operative issues and was pleased with the procedure's outcome.

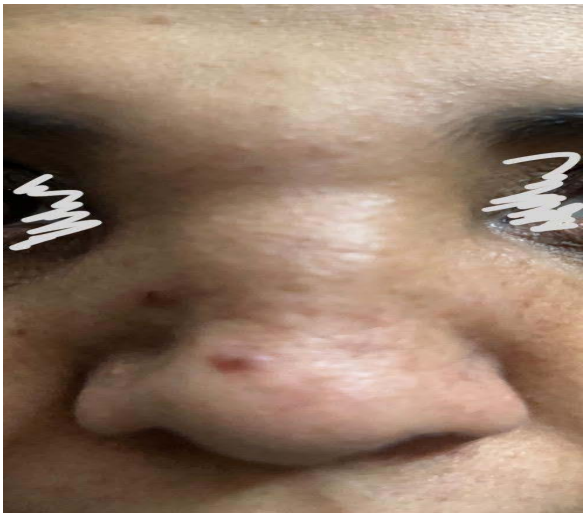

Figure 1: Patient with midline nasal mass

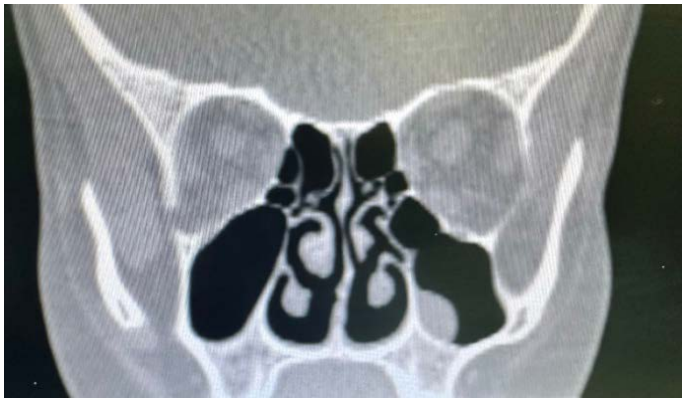

Figure 2: CT PNS w/contrast _left maxillary antral polyp or retension cyst, ethmoidal cells opacification

DISCUSSION

McDermott et al. was the first to recognize NCMH as a distinct clinic-pathological entity in 1998 and it is also known as ‘‘chondroid hamartoma’’, ‘‘mesenchymoma’’, ‘‘nasal hamartoma’’, etc. NCMHs are predominantly benign lesions that are locally destructive and because of their aggressive appearance can be mistaken for a malignant tumor. NCMH is slow growing lesion with delayed onset of symptoms[11].

An NCMH has been given several names, including chondroid hamartoma, mesenchymoma, and nasal hamartoma. Only 25 examples have been mentioned in the English literature to yet. NCMH pathophysiology is currently unclear. Although it was formerly assumed to be developmental or congenital, it now appears implausible due to reported occurrences affecting people who had an uneventful childhood. It is possible that the tumor is caused by an underlying genetic predisposition combined with the appropriate stimulus. This stimulation could be environmental in nature, and it could be related to chronic inflammation or hormones[12,13].

An NCMH is distinguished by a diversity of mesenchymal components with a focally lobular morphology. The most visible components are irregular islands of adult and immature hyaline cartilage, with a few binucleated chondrocytes. The cartilage islands are clearly distinguished from the surrounding stromal tissues, which have a myxoid background and are made up of a generally bland and compact spindle cell population with varied cellularity. There have been no reports of aberrant mitotic figures or malignant features. Reactive bone, small thick-walled arteries, cystic development, and erythrocyte-filled gaps, on the other hand, have been reported[14,15].

Differential diagnoses include hemangioma, angiofibroma, antrochoanal polyp, nasoethmoidal encephalocele, or meningoencephalocele, as well as other rare benign pediatric mass lesions. CT and MRI scans are used in radiological diagnostics. Head CT and MRI concentrating on the sinonasal tract can identify the place of origin, true size of the lesion, and interaction with key structures. The CT modality reveals diverse soft-tissue masses with primarily solid and cystic components. The microscopic appearance of NCMH is characterized by uneven islands of adult and immature hyaline cartilage with sporadic chondrocytes. The cartilage islands have well-defined borders with the surrounding stromal tissues, which have a myxoid backdrop[16,17].

Complete excision is the preferred treatment, and good outcomes are frequently obtained. NCMH is susceptible to endoscopic resection. Nowadays, total endoscopic resection should be regarded the primary treatment option in patients of NCMH. Incomplete excision may result in tumor recurrence; nevertheless, only a few cases of NCMH return have been described. As a result, there is no need for adjuvant radiotherapy or chemotherapy. At the same time, some evidence suggests that total resection cannot completely remove the chance of recurrence[14,15].

CONCLUSION

The majority of NCMH patients are children and infants under the age of one, but there have been a few adult cases of presentation. In the majority of instances, surgical excision is the therapy of choice, with low recurrence rates. CT and MR imaging characteristics of an NCMH, a rare benign hamartoma, in patients. Because it frequently mimics cancer on imaging, understanding this entity is critical for avoiding potentially damaging therapy.

The endoscopic endonasal technique is the gold standard in surgical management. In the event of an incomplete resection, rigorous follow-up MRI investigations are advised.

This case and analysis demonstrated that NCMH might be mistaken for other benign and malignant lesions, and that surgeons and clinicians should be aware of unusual illnesses that cause nasal masses.

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Competing Interest: None

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REFERENCES

1. Moher D, Liberati A, Tetzlaff J, et al. Preferred reporting items for systematic reviews and meta-analyses: the PRISMA statement. J Clin Epidemiol 2009;62(10):1006-12.
2. Schultz KA, Yang J, Doros L, et al. pleuropulmonary blastoma familial tumor predisposition syndrome: a unique constellation of neoplastic conditions. Pathology Case Rev 2014;19(2):90-100.
3. Kang Jun HYO, Gueng HA, Young MK, et al. Nasal Chondromesenchymal Hamartoma a case report. Korean J Pathol 2007;41:258-62.
4. Cho YC, Sung IY, Son JH, et al. Nasal chondromesenchymal hamartoma: report of a case presenting with intraoral signs. J Oral Maxillofac Surg 2013;71(1):72-6.
5. Roland NJ, Khine MM, Clarke R, et al. A rare congenital intranasal polyp: mesenchymal chondrosarcoma of the nasal region. J Laryngol Otol 1992;106(12):1081-3.
6. Silkiss RZ, Mudvari SS, Shetlar D, et al. Ophthalmologic presentation of nasal chondromesenchymal hamartoma in an infant. Ophthal Plast Reconstr Surg 2007;23(3):243-4.
7. Kim DW, Low W, Billman G, et al. Chondroid hamartoma presenting as a neonatal nasal mass. Int J Pediatr Otorhinolaryngol 1999;47(3):253-9.
8. Hsueh C, Hsueh S, Gonzalez-Crussi F, et al. Nasal chondromesenchymal hamartoma in children: report of 2 cases with review of the literature. Arch Pathol Lab Med 2001;125(3):400-3.
9. Shet T, Borges A, Nair C, et al. Two unusual lesions in the nasal cavity of infants—a nasal chondromesenchymal hamartoma and an aneurysmal bone cyst like lesion. More closely related than we think? Int J Pediatr Otorhinolaryngol 2004;68(3):359-64.
10. Kim B, Park S, Min HS, et al. Nasal chondromesenchymal hamartoma of infancy clinically mimicking meningoencephalocele. Pediatr Neurosurg 2004;40(3):136-40.
11. Alrawi M, McDermott M, Orr D, et al. Nasal chondromesynchymal hamartoma presenting in an adolescent. Int J Pediatr Otorhinolaryngol 2003;67(6):669-72.
12. McDermott MB, Ponder TB, Dehner LP. Nasal chondromesenchymal hamartoma: an upper respiratory tract analogue of the chest wall mesenchymal hamartoma. Am J Surg Pathol 1998;22(4):425-33.
13. Chandra M, Sharma N, Venkatahalam VP. Nasal chondromesenchymal hamartoma: a case report and review of literature. JK Pract 2014;19(5):53-9.
14. Stewart DR, Messinger Y, Williams GM, et al. Nasal chondromesenchymal hamartomas arise secondary to germline and somatic mutations of DICER1 in the pleuropulmonary blastoma tumor predisposition disorder. Hum Genet 2014;133(11):1443-50.
15. Norman ES, Bergman S, Trupiano JK. Nasal chondromesenchymal hamartoma: report of a case and review of the literature. Pediatr Dev Pathol 2004;7(5):517-20.
16. Yao-Lee A, Ryan M, Rajaram V. Nasal chondromesenchymal hamartoma: correlation of typical MR, CT and pathological findings. Pediatr Radiol 2011;41(5):675-7.
17. Kato K, Ijiri R, Tanaka Y, et al. Nasal chondromesenchymal hamartoma of infancy: the first Japanese case report. Pathol Int 1999;49(8):731-6.