

## Nasal chondromesenchymal hamartoma in an adolescent: a report of case and review of literature

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### Abstract

**Interaction:** Nasal chondromesenchymal hamartoma (NCMH) is an extremely uncommon primary benign cartilaginous growth of the nasal and paranasal sinuses. It has been reported almost exclusively in infancy. We report a NCMH in a 23-year-old patient who presented with anosmia, right gaze diplopia and proptosis. CT scan of paranasal sinuses revealed frontal sinus mass extending to the right orbit, ethmoid cells and nose. After an initial inconclusive incisional biopsy, the patient underwent a complete radical resection; and the defect was reconstructed with osteoplastic flap of frontal sinus. Histopathological examination confirmed NCMH, which, we believe, probably had been present and undetected for many years. This report greatly extends the age of NCMH, as part of the differential diagnosis of cartilaginous lesions of the nose and paranasal sinuses.

**Key words:** Hamartoma , Nasal cavity, Transnasal biopsy, Cartilaginous lesions.

### Introduction

Hamartomas of the nasal cavity are rare and usually identified in children. Nasal chondromesenchymal hamartoma (NCMH) is a new disease entity recently proposed by McDermott et al(1).

NCMH demonstrates characteristic clinicopathological feature and is presumed to be an upper respiratory tract analog of chest wall mesenchymal hamartoma(2). The median of age range is up to 7 years, mostly infants are less than 3 months (1,3).

We report a 23-year-old man presenting with a massive destructive tumor in the nasal cavity.

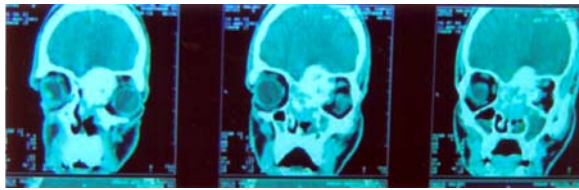
### Case report

A 23-year-old patient presented with anosmia, right gaze diplopia and proptosis with decreased visual acuity to about 5/10 on the right side (Fig.1)



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He had a history of intermittent epistaxis since two years ago, progressing gradually to frequent epistaxis even after minor trauma. A transnasal biopsy was inconclusive. CT scan of paranasal sinuses showed frontal sinus mass extending to right orbit, ethmoid cells and nose, with destruction of orbital roof and medial wall of right orbit (Fig 2).



**Fig.2:** CT scan of paranasal

The patient underwent surgery through the osteoplastic flap of frontal sinus approach with a bicoronal incision and the tumor was resected completely from frontal sinus, orbit, nose and dura of anterior cranial fossa. The right frontal sinus, roof and medial wall of orbit was reconstructed by periosteal flap. The patient was discharged five days after surgery in good condition and improved visual acuity.

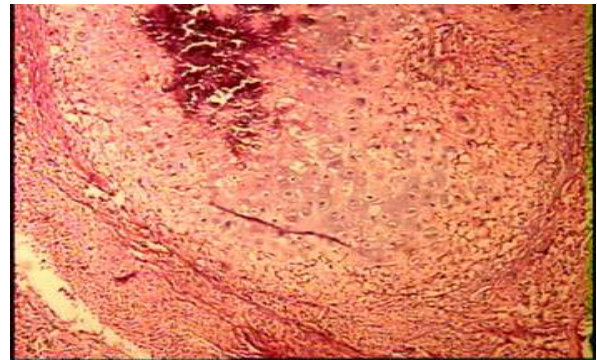
#### Methods and materials

Resected samples were fixed in 10% formalin, embedded in paraffin, sectioned in 4 micron thickness, stained with hematoxylin-eosin and examined by light microscopy.

#### Pathological findings

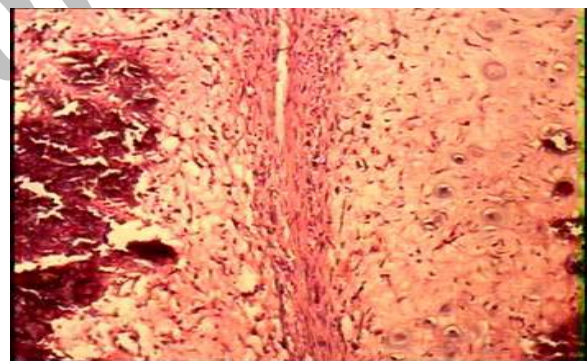
On gross examination, the samples removed from frontal sinus and the nasal cavity were fragmented fibrous tissue measuring up to 7×5×3 cm.

They were white and firm in consistency. Foci of calcification and bony tissue were included in the sample. Histologically, the tumor was composed of cellular, lobular cartilaginous nodules. The nodules appeared to merge and blend with the contiguous stroma, demonstrating features resembling immature cartilaginous tissue (Fig 3).



**Fig 3:** Low power photomicrograph showing a lobular proliferating cartilage

The nodules were occasionally hypercellular but lacked significant atypical feature. The stroma neighboring cartilage nodules were mainly composed of spindle cells with variable cellularity (Fig 4).



**Fig 4:** High power photomicrograph showing chondroid areas and primitive mesenchymal (40x)

Foci of calcification were observed in the entire lesion. It seems to be due to longstanding of the lesion.

#### Discussion

Nasal chondromesenchymal hamartoma is a new clinicopathological entity presented by McDermott *et al* in 1998(1). Their report was seven cases of infantile nasal tumor diagnosed before 3 months of age, except for one case diagnosed at the age of 7 year.

Extension to anterior fossa in contiguous fashion with characteristic radiographical features was observed in four of seven cases. The histological findings, including several mesenchymal elements, are similar to those of mesenchymal hamartoma of the chest wall (MHCW) in infancy (2). From the clinical data of the seven cases, the lesion was considered benign. To the best of our knowledge, no additional case has been reported afterwards, although, Keisuke et al reported the first Japanese case, Alrawi et al reported NCMH in a 16 year old, the oldest patient to have been reported(4,5).

The present case seems to be a typical example of "common type" NCMH with longstanding changes. We assume that the lesion had been present but unnoticed since birth. Its presentation in an adult is therefore, particularly unexpected. As with other hamartomas, NCMH may continue to grow as the child grows and perhaps the adolescent growth spurt was sufficient to bring it to the attention at an observant parent.

If the clinicopathological features, including age, localization, symptoms and histopathology are well recognized, it seems relatively easy to differentiate NCMH from other disease entities. Because cartilaginous tissue is a prominent component of NCMH, it is reasonable to consider cartilaginous tumors as differential diagnosis. They include chondromas, chondrosarcoma and chondroblastic osteosarcoma(6,7).

Chondrosarcoma is distinctly unusual in a pediatric age group (6), and the heterogeneous histologic appearance also argues against such an interpretation. Chondroblastic osteosarcoma may arise in facial bones and has been described in nasal bones by Rotand et al (7).

It was also considered here, especially in view of the destruction of nasal cartilage and the age of the patient. However, the circumscribed nature of the resected mass and presence of lamellar bone (as opposed to

malignant osteoid) within the mass did not support such a designation.

Meningioma with chondro osseous metaplasia is also a candidate for differential diagnosis.

However, onset in the congenital infantile period and chondro osseous metaplasia are extremely rare in meningioma of the upper respiratory tract (8-10). Furthermore, the variety of mesenchymal components observed in NCMH lessens the possibility of meningioma.

To this point, reported examples of NCMH have behaved in a benign fashion, even when incompletely excised(1). However it may be difficult to decide how to treat residual tumor or unresectable lesions. Local resection through functional endoscopic sinus surgery (FESS) is recommended when the tumor is localized in nasal cavity. No adjuvant therapy is necessary(1).

In conclusion, we report an example of a NCMH in a 23-year-old patient; the oldest patient reported to date to our knowledg. As typical examples of the lesion have only previously been reported in infants (the oldest patients have been 16-years-old (4), this case substantially extends the age range in which such lesions may be encountered. NCMH should, therefore, be considered in the differential diagnosis of chondroid proliferations of the nose and paranasal sinuses irrespective of age at presentation.

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### خلاصه

## هامارتوم کندرومزانشیمال بینی در بالغین: معرفی مورد و مروری بر مقالات

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**مقدمه:** هامارتوم کندرومزانشیمال بینی رشد غضروفی خوش خیم اولیه و بسیار ناشایع بینی و سینوس های پاراناژال می باشد. تاکنون تقریباً بدون استثناء در کودکان گزارش شده است. نویسندگان این مقاله یک مورد را در بیمار ۲۳ ساله که با عدم حس بویایی، دیپلولی فلجی به سمت راست و پرویتوز مراجعه کرده بود معرفی می کنند. CT اسکن سینوس های پاراناژال توده سینوس فرونتال با گسترش به اوربیت راست، سلول های اتموئید و بینی را آشکار کرد. بعد از یک بیوپسی انسزیونال بدون نتیجه، بیمار تحت رزکسیون کامل رادیکال قرار گرفت و نقص ایجاد شده با فلپ استئوبلاستیک سینوس فرونتال بازسازی شد. امتحان هیستوپاتولوژیک هامارتوم کندرومزانشیمال بینی را ثابت کرد و ما معتقدیم که احتمالاً برای چندین سال وجود داشته و تشخیص داده نشده است. این گزارش به طور واضح طیف سن این تومور را گسترش می دهد که بدین ترتیب در تشخیص افتراقی ضایعات غضروفی بینی و سینوس های پاراناژال قرار می گیرد.

**واژه های کلیدی:** هامارتوما، حفره بینی، بیوپسی ترانسناژال، ضایعات غضروفی