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International Journal of Histopathological Interpretation

Case Report

NASAL CONDROMESENCHYMAL HAMARTOMA- A DIAGNOSTIC DILEMMA

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How to cite: Syeda Iqra Usman et al., Nasal Condromesenchymal Hamartoma- A Diagnostic Dilemma Int J Histopathol Interpret 2023; 12(2), 1-5 DOI: https://doi.org/10.56501/intjhistopatholinterpret.v12i2.747

Received : 23/02/2023 *Accepted:04/01/2023* *Web Published: 10/07/2023*

Abstract

Nasal chondromesenchymal hamartoma (NCH) is a very rare and benign tumor of sinonasal tract and usually presents in infants and young children with varied symptoms depending upon the site of tumor in nasal cavity or paranasal sinuses. Here we present a case of 12 years old female who presented with on and off nasal obstruction for 1 year. On the basis of clinical and radiological findings she was diagnosed as a case of ethmochoanal polyp but radiological findings also showed bony thinning because of which malignancy could not be ruled out. But surprisingly her histopathological findings were in favour of chondromesenchymal hamartoma which was confirmed by vimentin and S100 positivity.

Keywords: Hamartoma, Nasal endoscopy, S-100, Paranasal sinus

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INTRODUCTION

Nasal chondromesenchymal hamartoma (NCH) is a very rare benign tumor of sinonasal tract and mostly presents in infants and young children.[1] It usually arises as growth in nasal septum or vestibule. Involvement of ethmoidal and maxillary sinus is rather rare.[2] Patient presentation and common symptoms are dependent on location and size of tumor that includes nasal obstruction, difficulty in breast feeding, epistaxis, rhinorrhea, and middle ear effusion. Additionally, involvement of the orbit or cranial cavity may cause exophthalmos or enophthalmos, oculomotor disorders, and neurological dysfunction.[3]

CASE REPORT

A 12 years old female presented to out-patient department with complaint of on and off nasal obstruction for 1 year. She also complains of on and off headache and difficulty in breathing. There was no history of rhinorrhea, discharge from ear, epistaxis or any visual impairment.

An otolaryngologic evaluation revealed a large intranasal mass. On examination her vitals were stable and on nasal endoscopy large pale polypoidal mass was noticed involving the right nasal cavity. Non contrast computed tomography paranasal sinuses showed a large soft tissue density completely involving right ethmoidal sinus, right frontal sinus and right nasal cavity with evidence of extension into nasopharynx and severe airway luminal compromise.

Bone thinning and bone destruction was noted in the walls of all paranasal sinuses on right side, right middle and inferior turbinate, bilateral nasal bone, nasal septum and right lamina papyracea.

It was also causing mass effect in the form of deviation of nasal septum towards left side and bulging of right lateral wall of ethmoid causing compression of right maxillary sinus with hypodense collection in right maxillary and sphenoid sinus. On the basis of clinical and radiological findings a provisional diagnosis of ethmochoanal polyp was made and malignancy was also kept as one of the differential due to mass effect, bony thinning and destruction as noted on non-contrast computed tomography. Endonasal endoscopic surgery was performed.

Multiple tissue pieces were received for histopathological examination which aggregately measured 3.5x3x0.8cm with largest tissue piece measuring 3.5x1.8x0.4cm. Microscopically, fragmented tissue pieces showed pseudostratified ciliated columnar lining with underlying lamina propria showing dense non-specific inflammation, fragments of immature cartilage, fibromyxoid stroma, dilated vascular spaces and metaplastic bone formation.

Immunohistochemistry was applied for confirmation which showed diffuse strong cytoplasmic positivity for vimentin and diffuse strong nuclear and cytoplasmic positivity for S-100. Ki67 proliferation index was 2-3%.

On the basis of history, clinical, radiological and histopathological findings she was diagnosed as a case of nasal chondromesenchymal hamartoma (NCH).

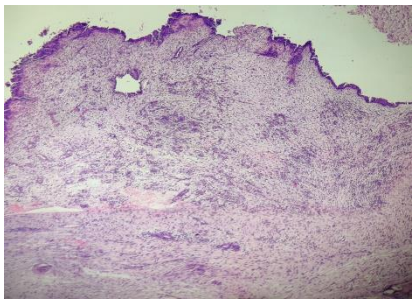

Figure 1: Section showing columnar lining with underlying Fibromyxoid stoma having inflammation (H & E, 40X)

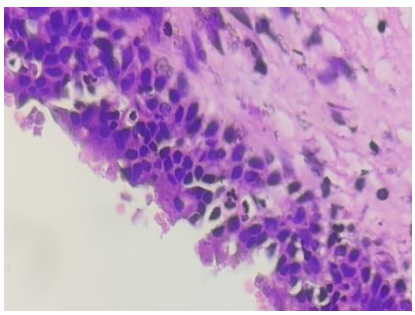

Figure 2: Section shows pseudostratified ciliated columnar lining with underlying lamina propria showing dense non-specific inflammation (H & E, 400X)

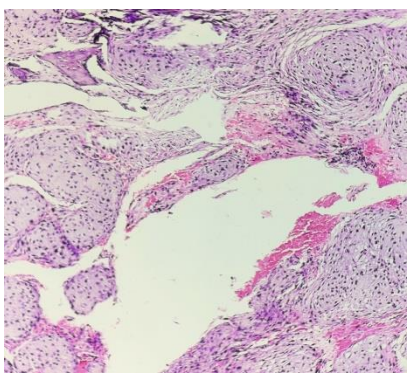

Figure 3: Sections shows area of cartilaginous differentiation (H & E, 400X)

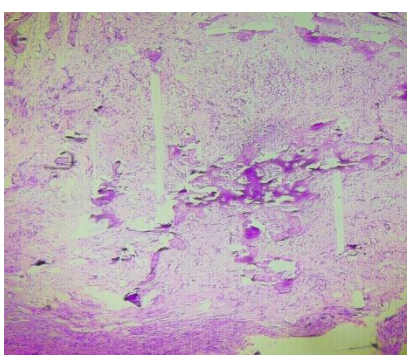

Figure 4: Section shows metaplastic bone formation (H & E, 100X)

DISCUSSION

The term 'Hamartoma' was first introduced by Albrecht (1904) which means some kind of error in the growth of the three germinal layers. Clinically it may mimic as malignancy but histologically it is composed exclusively of components derived from the local tissues for the given anatomical site which are abnormal in amount, ratio, or distribution.[4] Hamartomas of the nasal cavity and nasopharynx can be classified as epithelial, mesenchymal and mixed epithelial and mesenchymal types. They include chondromesenchymal

hamartoma, respiratory epithelial adenomatoid hamartoma (REAH) and seromucinous hamartoma. Each of these has characteristic features, but occasional lesions will show overlapping features of multiple types.[5]

McDermott et al. were the first to recognise NCMH as a distinct clinic-pathological entity in 1998.[6] Exact pathogenesis is yet not known but early age of presentation points towards genetic predisposition. DICER 1 mutation has been studied in this context. On histopathological examination they show similarities to other mesenchymal hamartomas consisting of islands of chondroid tissue such as hyaline cartilage, areas of calcification, and mesenchymal cellular elements such as spindle cells and myxoid stroma.[7] It has varied clinical presentations ranging from nasal obstruction to neurological manifestations due to mass effect and compression of surrounding structures. In our case patient presented with nasal obstruction, headache and difficulty in breathing.

Although it is a benign entity but on radiology it can mimic malignancy as it causes local destruction. This was true in our case also because bone thinning and destruction was noted on non-contrast computed tomography. There is a long list of differential diagnosis on imaging which includes hemangioma, angiofibroma, nasoethmoidal encephalocele, nasal glioma, inverted papilloma, giant cell reparative granuloma, ossifying fibroma, chondro-osseous respiratory adenomatoid hamartoma, and aneurismal bone cyst as benign pediatric tumors.[8] In our case on the basis of clinical and imaging modalities provisional diagnosis of ethmochoanal polyp was made.

NMCH can have varied histological findings but there are certain components, initially described by McDermott et al. in 1998 that are found in all or the majority of cases that includes lobular cartilaginous areas that can be hypercellular or normocellular. These may have sharp borders, or may blend into the surrounding stroma. The stroma typically consists of spindle cells or myxoid tissue or both. Osseous tissue along with cystic blood-filled spaces has also been reported in majority of cases.

These findings were true in our case also.[9] Histopathological findings were in contrast to provisional diagnosis of ethmochoanal polyp which shows edematous, fibrotic or loosely myxoid stroma covered by respiratory epithelium with infiltration of mixed inflammatory infiltrate including lymphocytes, plasma cells, eosinophils, neutrophils and mast cells.10 Immunohistochemical staining demonstrates vimentin positivity in the stromal-mesenchymal elements and S-100 positivity in both the mature cartilage and spindled areas.6 Both the immunohistochemical stains showed diffuse positivity in our case.

Main modality of treatment is complete excision and recurrence occur only if the excision has not been done completely. As it is a benign neoplasm no radiotherapy of chemotherapy is needed. In our case also tumor was excised and patient dis not had any fresh complaints at 3month and 6 months of follow up.

On the basis of histopathological findings and immunohistochemistry most of the differential diagnosis were ruled out in our case and ki-67 proliferation index was low which pointed towards a benign entity.

CONCLUSION

Nasal chondromesenchymal hamartomas are very rare benign neoplasms with very few reported cases. They can be misdiagnosed on clinically and radiologically as malignant neoplasms because they can cause mass effect, bone thinning and bone destruction, hence histopathological examination must always be performed along with immunohistochemistry to avoid unnecessary chemoradiations. This case provides insight that nasal hamartoma can be confused with sinonasal polyps and malignancy hence thorough work up and examination is required for proper treatment.

Financial support and sponsorship:

Nil

Conflicts of interest

There are no conflicts of interest

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