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Congenital Nasal Chondromesenchymal Hamartoma

ABSTRACT

Objectives: To report the case of a congenital nasal chondromesenchymal hamartoma in a one-year-old female and review the literature, identifying problems encountered in confirming the diagnosis and in treatment of this patient.

Methods:

Design: Case Report

Setting: Tertiary Public General Hospital

Patient: One

Results: A one-year-old female with an intranasal mass noted at birth and with subsequent unilateral maxillary enlargement is described. Computed tomography showed calcifications and erosion of adjacent bony structures. Histopathology and immunohistochemistry of an intranasal biopsy were interpreted as chordoma, a malignant tumor. Following surgical excision, the final histopathologic diagnosis was chondroid hamartoma.

Conclusion: Only 20 cases of nasal chondromesenchymal hamartoma have been reported in the literature worldwide. These tumors may present clinically, histopathologically and radiologically as malignant tumors and may mislead even the experts. The whole clinical picture should be taken together to avoid misdiagnosis as a malignancy and to facilitate appropriate management.

Keywords: *nasal chondromesenchymal hamartoma, nasal masses in infancy, nasal chondroid lesions*

Nasal masses in infancy are infrequently encountered. Most are developmental anomalies such as encephalocoeles, gliomas and nasolacrimal duct cysts. The rest are neoplasms, primarily composed of teratomas and dermoid cysts. Occasionally, a variety of benign and malignant soft tissue tumors occur in children.¹

The majority of head and neck neoplasms in children are benign. Primary malignant neoplasms of the head and neck are not common and account for about 5 % of neoplasms

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occurring in childhood.² In children, the neoplasms are more frequently reticuloendothelial, neural or mesenchymal, as opposed to the predilection for epithelial neoplasms in adults.¹ Chondrogenic tumors of the maxillofacial region may be difficult to differentiate histologically and radiologically as to whether they are benign or malignant.

We present the case of an infant with a nasal chondromesenchymal hamartoma which was confused with a malignancy. This clinicopathologic entity is remarkable not only because of its rarity but also because it may present a diagnostic dilemma with major implications in management.

CASE REPORT

A term female was born at home with the assistance of a traditional birth attendant to a then 23-year-old G2P1 (1001) mother. Because of cyanosis and dyspnea at birth, the baby was rushed to a local hospital where physicians were unable to pass a suction catheter through the right nasal cavity. Glatzel's mirror test showed no misting from the nostrils, and a whitish mass was noted in the right nasal cavity. Subsequently, chronic mucoid discharge from the right nostril and a progressively bulging right maxilla were noted.

A computed tomography (CT) scan of the paranasal sinuses at three months of age revealed a mixed-enhancing, soft tissue mass at the right nasal antrum with irregular foci of calcifications, measuring 2.2 x 3.22 x 3.03 cm. The mass pushed the nasal septum medially and the medial wall of the right maxillary sinus laterally. No bony erosions, intraorbital or intracranial extensions were seen. Considerations were an ossifying fibroma, calcifying polyp or dentigerous cyst. Surgery was advised but deferred by the mother.

At eight months of age, repeat CT scans revealed a 3.0 x 3.95 x 3.33 cm mass, now occupying the right maxillary and ethmoid sinuses with extension in the anteromedial portion of the right orbit (*Figure 1*). There had been an increase in size of 83.7% over the past five months.

They finally agreed to admission, and repeat CT scans revealed a 3.20 x 4.03 x 3.4 cm ethmoidal mass (a 103 % volume increase in 9 months), with lysis of the right lamina papyracea, extension into the right intraorbital extraconal compartment with involvement of the medial rectus and inferior oblique muscle, and thinning of the medial wall of the right maxillary sinus (*Figure 2*). No intracranial extensions were noted. The radiological impression was an ethmoidal mass with malignant features.

An intranasal punch biopsy was interpreted as a mesenchymal neoplasm compatible with chordoma, a diagnosis later supported by slide review. Immunohistochemical stains were strongly positive to S100 and vimentin with non-immunoreactivity to cytokeratin. These findings were still interpreted as consistent with a chordoma.

Considering a highly malignant tumor, a right medial maxillectomy

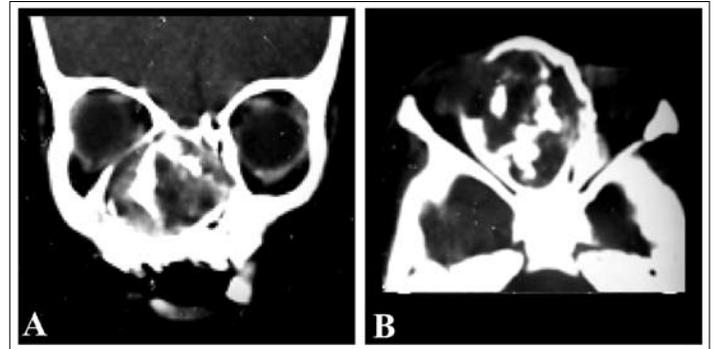


Figure 1. Paranasal sinus CT scan (A, coronal and B, axial cut) at 8 months of age showing an ethmoidal mass with extension into the right maxillary and ethmoid sinuses and the anteromedial portion of the right orbit

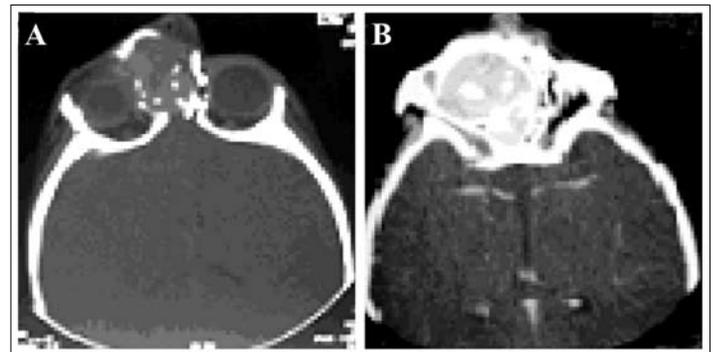


Figure 2. PNS CT scan (A, axial cut, bone window; B, axial cut with contrast) at one year of age showing an ethmoidal mass with lysis of the right lamina papyracea and extension into the right intraorbital extraconal compartment and thinning out of the medial wall of the right maxillary sinus



Figure 3. A right lateral rhinotomy with a Lynch extension revealed a well-circumscribed mass in the ethmoid sinus extending to the maxillary sinus and medial wall of the orbit

via right lateral rhinotomy combined with a Lynch incision was performed (*Figure 3*). Intra-operatively, an eggshell-thin anterior maxillary wall, a whitish mass filling the right ethmoid and maxillary sinuses, and septum deviated to the contralateral side were noted. The orbital wall was intact but the nasal roof showed small thinned-out areas. The patient tolerated the procedure without any problems or post-operative complications.

Grossly, the mass was cream to tan-colored, firm and well-

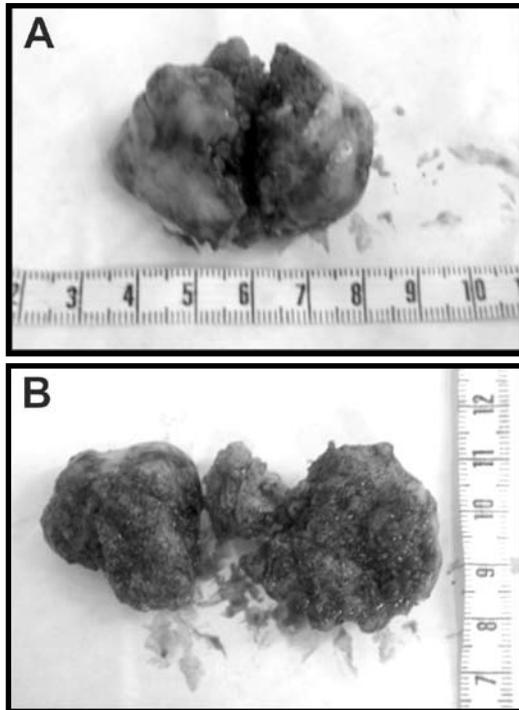


Figure 4. Gross appearance of the ethmoidal mass measuring 4 x 3.5 x 3 cm. (a) whole specimen; (b) cut surface

circumscribed, measuring 4 x 3.5 x 3 cm. The surface was smooth, but gritty and irregular when cut (Figure 4). No tumor hemorrhages were noted. Histologically, nodules of hyaline cartilage ranging from fairly immature looking chondroid stroma to islands of mature hyaline cartilage were found in a variably cellular stroma that was fibrocytic in some areas and loose and myxoid in others (Figure 5). Bundles of dense collagen as well as occasional spicules of bone were also noted.

The histopathologic diagnosis at this institution was a mesenchymal neoplasm with chondroid differentiation. Further analysis at the Memorial Sloan-Kettering Cancer Center and IMPATH Laboratories New York yielded a final pathologic diagnosis of chondroid hamartoma with osseous metaplasia.

DISCUSSION

Nasal chondromesenchymal hamartoma (NCMH) is a rare, destructive yet benign, tumefactive lesion involving the nasal cavity and paranasal sinuses. The term was first coined by McDermott *et al.* in 1998 to describe a distinct clinicopathological entity composed of a proliferation of mesenchymal and cartilaginous elements.⁴ Nasal chondroid lesions, nasal hamartoma, chondroid hamartoma and mesenchymoma are other terms used to refer to this disease entity. It usually affects infants less than 3 months of age although there have been reports of occurrence in adolescents and young adults.⁵ One case series reports occurrence in a 69-year-old female.⁶ Presently, there have

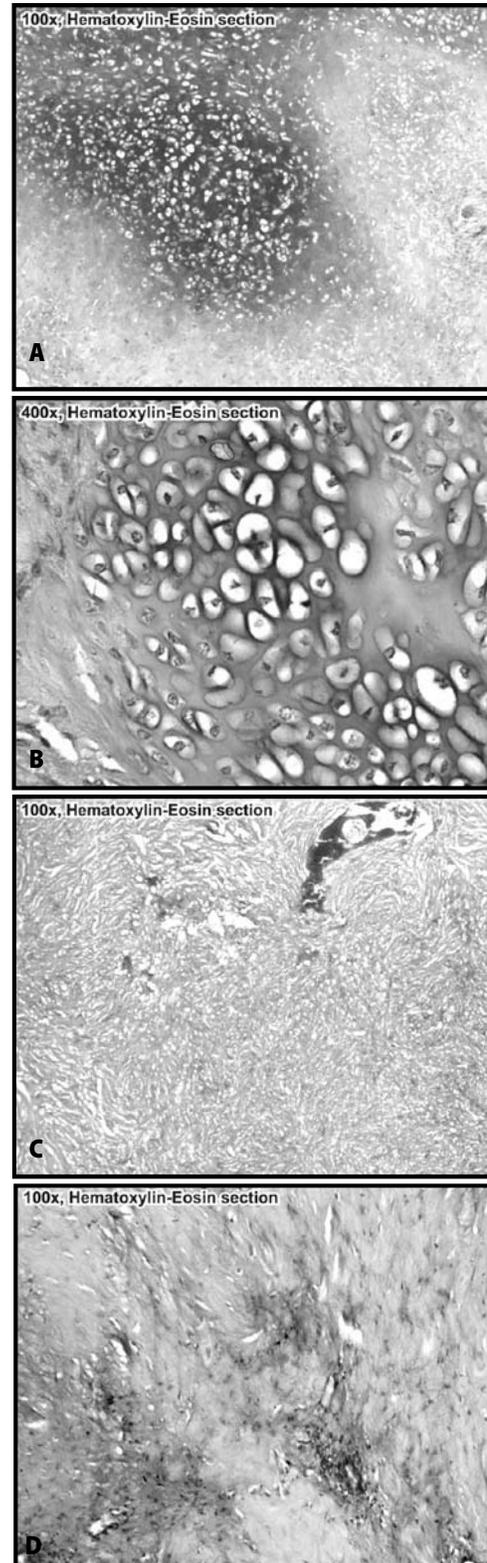
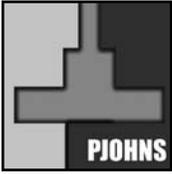


Figure 5. NCMH in this patient showing nodules of hyaline cartilage ranging from (A) fairly immature looking chondroid stroma to (B) islands of mature hyaline cartilage. Stroma is (C) fibrocytic in some areas and (D) loose and myxoid in others



only been 20 cases of NCMH reported in English medical literature.^{4,5,7-14}

Due to the paucity of cases, the pathogenesis of NCMH is not well understood. Hamartomas are characterized by an abnormal mixture of tissues indigenous to that area of the body, but with an excess of one or more of the tissue types. The development of hamartomas may involve errors during fetal growth or disturbances of immature tissues in the post-natal period.⁴ It is believed that fibroblasts and myofibroblasts are the major component cells in NCMH.⁹ NCMH was initially thought to be present at birth, indicating a developmental or congenital origin. However, with reports of occurrence in adolescents and adults, alternative explanations such as the role of inflammation, cytokines, and growth factors or the association with hormonal stimulation are being investigated.^{11,13}

Differentiation between NCMH and a malignant lesion like a chordoma may be difficult. NCMH have a seemingly infiltrative nature. As such, it may mimic malignant tumors in its clinical, radiologic or even histopathologic presentation. In the literature, presenting symptoms included a nasal mass, nasal obstruction, respiratory distress and a maxillary bulge which were all present in this patient. Orbital involvement can result in proptosis, enophthalmos or impairment of eye movement. Intracranial extension of the tumor can result in neurologic manifestations such as hydrocephalus and oculomotor disturbance.¹⁰ Our patient had no ocular or neurologic symptoms or actual orbital invasion despite a preoperative CT scan suggesting intraorbital involvement.

Radiologically, NCMHs may be nonencapsulated and ill defined, containing both solid and cystic portions. Calcifications may be present.⁴ The adjacent paranasal sinuses are frequently involved and erosion of the surrounding bone and extension to the skull base and orbital region are not uncommon. In a review of 19 cases by Johnson *et al.*,¹⁴ 67% demonstrated bony remodeling, thinning or erosion and 53% demonstrated intracranial extension through the cribriform plate to the anterior cranial fossa. 50% revealed internal calcifications while 40% revealed cystic components. In this patient, the rapid growth of the mass, calcifications and destruction of adjacent bony structures were interpreted as consistent with a malignancy.

Biopsy results may also be misleading. Histologically, NCMH is comprised of a variety of mesenchymal components with a focally lobular architecture of irregular islands of hyaline cartilage with occasional binucleated chondrocytes and cartilaginous islands.⁴ Thus, NCMH may morphologically overlap with chondroid lesions including malignancies such as chondrosarcoma and chordoma (Figure 6).

Immunohistochemistry may be helpful if the initial histological picture is unclear. Chordomas, which are tumors arising from embryonic notochordal remnants, and NCMH both stain positive for S100 and

vimentin which are markers for neuroepithelial cells and mesenchymal cells respectively. However, chordoma stains positive for cytokeratin while NCMH is negative.⁹ Negativity to cytokeratin in this case should have been a strong reason to discount a chordoma, a diagnosis which was strongly maintained by the pathologist.

Taking all these into account may elucidate how a nasochondromesenchymal hamartoma, a rare but benign lesion, was mistaken for a chordoma, an aggressive and malignant tumor. Accurate differentiation between NCMH and malignant tumors is very important, as a hamartoma's capacity for growth is usually limited and its biologic behavior is typically benign,⁸ making complete surgical excision sufficient therapy. When as in our case, a misdiagnosis of malignancy is made, excessively radical surgery and possibly, potentially harmful neo-adjuvant radiotherapy may result.

Despite its relatively low prevalence, NCMH should be considered as a differential diagnosis in patients with chondroid lesions in the maxillofacial region, particularly in infants. Caution should be exercised in diagnosing radiologically and histologically aggressive-looking maxillofacial tumors as malignant especially in infants. Ancillary studies may be necessary to further confirm a malignancy. As in all cases, individualized treatment of patients with head and neck tumors is very important. Factors such as age, co-morbid illnesses and location of the tumor should be considered in choosing treatment approaches.

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