Nasal Chondromesenchymal Hamartoma
- A Case Report -

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Hamartoma refers to a mass of disorganized overgrowth of
mature, specialized cells and tissues indigenous to the organ in
which it occurs. Although the cellular elements are mature and
identical to those found in the remainder of the organ, they do
not reproduce the normal architecture of the surrounding tis-
sue. McDermott et al. first described nasal chondromesenchy-
mal hamartoma (NCMH) as a tumefactive process of the nasal
passages and contiguous paranasal sinuses in seven children in
1998.1 NCMH commonly presents as respiratory difficulty, an
intranasal mass or facial swelling in young infants and children.
NCMH is usually benign and is cured by a complete excision.
However, invasion of the surrounding soft tissue and bone, es-
specially intracranial invasion has also been identified.2 NCMH
is histologically composed of well-demarcated nodules of carti-
lage with some variation in the cellular density and maturation
of the chondrocytes, a myxoid to spindle cell stroma, focal osteo-
clast-like giant cells in the stroma, and erythrocyte-filled spaces
resembling those of an aneurysmal bone cyst.1

Twenty cases of NCMH have been reported in the English
literature. We report a case of NCMH in a 14-year-old boy along
with the histological and immunohistochemical analyses.

CASE REPORT

A 14-year-old boy presented with a 5 cm sized mass in the
left maxillary sinus accompanied by facial swelling and a loose
tooth of a 20 day duration. The physical examination revealed
swelling of the left maxillary area with tenderness and tooth
loosening. His prior medical history was unremarkable. The
laboratory data were normal. A CT scan revealed a 5 cm sized
lobulating mass in the left maxillary sinus with destruction and
remodeling of the adjacent bone (Fig. 1). The mass showed ho-

eral heterogeneous enhancement on contrast enhancement. The mass had destroyed the anterior maxillary sinus wall and was bulging into the face. The tumor also involved the alveolar process of the left maxilla and the soft tissue of the retroantral space. A preoperative wedge biopsy was performed. The histological diagnosis was a myxoid mesenchymal tumor and a definite diagnosis was deferred. A subtotal left maxillectomy with a bone graft was performed. The excised mass was composed of partly encapsulated, solid and cystic fragments of soft tissues, measuring 11.0 × 9.0 cm in size. The cut surface was pale brown and firm with ossified and myxoid areas (Fig. 2).

A histological examination of the hematoxylin and eosin stained

Fig. 1. A CT scan reveals a 5 cm sized lobulating mass (arrows) in the left maxillary sinus with adjacent bone destruction and remodeling. The mass destroys the anterior maxillary sinus wall and makes a bulging contour to the face. The mass shows peripheral heterogeneous enhancement.

Fig. 2. The tumor is well defined and partly encapsulated. The cut surface is pale brown, solid and cystic with cartilage.

Fig. 3. Histologic features of NMCH. (A) Hyaline cartilage component is most prominent. (B) The spindle cell component has fibrous matrix with variable myxoid change and occasional sclerotic change.
sections, and immunohistochemical analysis for CD34 (1:30, Neomarker, Fremont, USA), S-100 protein (1:100, Zymed, San Francisco, CA, USA), desmin (1:100, Zymed, San Francisco, CA, USA), smooth muscle actin (1:100, Zymed, San Francisco, CA, USA), pan-cytokeratin (1:100, Zymed, San Francisco, CA, USA), and p63 (1:50, DAKO, Glostrup, Denmark) were performed.

Microscopically, the tumor had a relatively well-delineated boundary with mucosal ulceration. The mass consisted of mesenchymal tissues containing hyaline cartilage, osteoid and spindle cell components. The hyaline cartilage component was the most prominent (Fig. 3A). The chondrocytes were distributed evenly within lacunar spaces. The lacunar spaces were occasionally markedly dilated. The chondrocytes contained small round nuclei with condensed chromatin and finely granular eosinophilic cytoplasm that was often vacuolated. The periphery of the hyaline cartilage component showed a transition to a spindle cell component. The spindle cell component contained a fibrous matrix with variable myxoid or sclerotic changes (Fig. 3B). Individual cells of the spindle cell component had oval to spindle nuclei and indistinct eosinophilic cytoplasm. Thick hyalinized eosinophilic osteoid-like trabeculae were focally present. No mitosis was found in the hyaline cartilage component but was occasionally found in the spindle cell component. Atypical
mitosis was not present. There were no osteoclast-like giant cells or aneurysmal bone cyst-like areas.

Immunohistochemically, all the mesenchymal cells tested positive for vimentin, and the chondrocytes tested positive for S-100 protein. The spindle cell component showed focal immunoreactivity for desmin, smooth muscle actin and CD34. (Fig. 4). There was no immunoreactivity for pan-cytokeratin and p63.

No further treatment was done and there has been no evidence of recurrence at 10 months after the excision.

### DISCUSSION

NCMH is presumed to be an upper respiratory analogue of chest wall mesenchymal hamartoma, which also occurs in young infants and children. The present case was older than most reported cases, and cases of adolescents and even older people have been reported.

Histologically, our case had the typical features of NCMH. Prominent hyaline cartilage, myxoid spindle cells and osteoid were noted. However, there were no osteoclast-like giant cells or aneurysmal bone cyst-like areas. The aneurysmal bone cyst-like area was reported to be variably present in NCMH. The cases in which McDermott et al., initially described aneurysmal bone cyst-like areas as one of the characteristic patterns of NCMH were mainly new born babies. The recently reported cases of adolescents and older age patients contained no aneurysmal bone cyst-like areas. Twenty one reported cases including the present case were analyzed for a correlation between age and aneurysmal bone cyst-like areas (Table 1). In 18 cases with in formation, the male and female ratio was 13/5. When the age was categorized into two groups, NCMHs in patients younger than 12 months were significantly associated with the presence of aneurysmal bone cyst-like areas (p<0.004) (Table 2).

The positive immunoreactivity for vimentin and S-100 protein and the negativity for pan-cytokeratin are in concordance with previous reports. In our case, smooth muscle actin, desmin and CD34 were positive in the spindle cell component. The reported expression pattern of smooth muscle actin was variable.

Nasal chondromesenchymal hamartomas usually test negative for desmin and one case report described a negative immunoreactivity for CD34. Our case showed unusual immunopositivity for desmin and CD34, which was also reported in a previous case. The expression of smooth muscle actin, desmin and CD34 is considered to be non-diagnostic for nasal chondromesenchymal hamartomas.

In our case, the mass was locally aggressive and was accompanied by bony erosion and extension to the soft tissue. NCMH can extend to the adjacent paranasal sinuses, most frequently the ethmoid sinus. Intracranial extension is not uncommon, and a case mimicking a meningoencephalocele was reported.

The locally aggressive radiological findings of NCMH and the rarity of mesenchymal tumors in the sinonasal tract can lead to a misdiagnosis, particularly when limited specimens have been obtained. In our case, the preoperative biopsy specimen was composed entirely of myxoid spindle cell lesion without other components. Therefore, it was suspected of being a benign or malignant myofibroblastic neoplasm, such as nodular fasciitis, myxofibrosarcoma or infantile fibrosarcoma.

However, the main histopathological differential diagnosis of NCMH is mesenchymal chondrosarcoma arising in the nasal cavity. Mesenchymal chondrosarcoma has been described as a particularly aggressive neoplasm in skeletal locations with a high tendency for late recurrence and delayed distant metastas-
A mesenchymal chondrosarcoma comprises small monotonous mesenchymal cells with a hemangiopericytomtous pattern and islands of relatively well-differentiated and comparatively benign appearing cartilage. In contrast to NMCH, the mesenchymal cells of a mesenchymal chondrosarcoma are more cellular and small with hyperchromatic nuclei and peripheral condensed chromatin.

NCMH shows benign clinical features. A complete excision is usually curative. However, an incomplete resection might be followed by the persistent growth of a residual tumor. This case underwent a complete excision, and there was no evidence of a recurrence ten months after surgery.

REFERENCES