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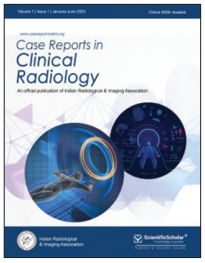

Case Report

Chondromesenchymal hamartoma of sinonasal region in a young child: Computed tomography, MR imaging, and pathological findings with brief review

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ABSTRACT

This report documents computed tomography and magnetic resonance imaging (MRI) findings for a case of sinonasal chondromesenchymal hamartoma and its recurrence occurring in an 11-year-old boy. We have described MRI dynamic contrast enhancement patterns in the recurrence of pathology, which, to the best of our knowledge, is the first documentation of such features. Sinonasal chondromesenchymal hamartoma is a rare pediatric benign hamartoma that can mimic aggressive inflammatory lesions or malignancies in the sinonasal region. A correct diagnosis is imperative to avoid unnecessary adjuvant therapy.

Keywords: Pediatric paranasal tumor, Nasal chondromesenchymal hamartoma, Computerized tomography, Magnetic resonance imaging, DICER1 tumors

INTRODUCTION

Nasal chondromesenchymal hamartoma (NCMH) is a very rare and benign tumor of the sinonasal tract mostly in infants and young children. NCMH patients present with locoregional symptoms due to mass effect. Symptoms can range from nasal obstruction to visual impairment and facial and dental pain. It has characteristic histopathological features similar to chest wall mesenchymal hamartoma in infancy.[1-5] We described the combined computed tomography (CT) and magnetic resonance imaging (MRI) features of NCMH and its recurrence in a young boy with a pathological review.[3,5-7]

CASE REPORT

An 11-year-old boy presented with a 6-month history of nasal obstruction and difficulty in breathing. Clinical examination showed obstruction of the posterior nasal cavity by a large polypoid fleshy mass. CT demonstrated a well-defined, expansile lesion in the ethmoid and sphenoid sinuses which was isodense to the cerebral cortex with hyperdense foci of the calcified matrix at the center. It caused scalloping and pressure remodeling of the sphenoid and ethmoid sinus walls without any frank bone destruction without orbital or intracranial invasion [Figure 1]. The lesion was devoid of fat or cystic component.

MRI revealed a brilliantly enhancing lesion that was homogeneously isointense to the gray matter on T1-weighted images (WIs) with few T1 hyperintense foci and homogeneously hyperintense

on T2WI. Non-enhancing ill-defined central T2WI and T2 gradient-echo-WI hypointense signals corresponded to the chondroid matrix elements [Figure 2].

The patient underwent debulking surgery on the lesion. On histopathology, microscopically, fragments of soft-tissue lesion were composed of lobules of the cartilaginous component with a spectrum of differentiation [Figure 3a-c] ranging from discrete nodules of mature hyaline cartilage and relatively cellular "immature" chondromesenchymal vague nodules; myxoid matrix showing mildly increased cellularity with few binucleate chondrocytes without appreciable cytologic atypia or pleomorphism [Figure 3d]. Amidst these cartilaginous components, bland spindled stroma were seen with increased vascularity, and scattered few multinucleated gaint cells and no mitotic figure.

Immunohistochemical stains were positive for vimentin in both spindle and cartilaginous components in a diffuse and strong pattern [Figure 4a], patchy expression of epithelial membrane antigen in both cartilaginous and spindle cell components [Figure 4b], positive for smooth muscle actin [Figure 4c in the spindle cell component, positive for S100 in the cartilaginous component [Figure 4d], and focal CD68 expression in spindle cell component [Figure 4e]. However, cytokeratin (AE1/AE3), desmin, and caldesmon were negative in both components. All features favored NCMH.

After 2 years, the patient presented with similar complaints and underwent an MRI which showed recurrence with comparatively very few discrete areas of cartilage. Dynamic contrast study showed progressive delayed enhancement without significant washout [Figure 5].

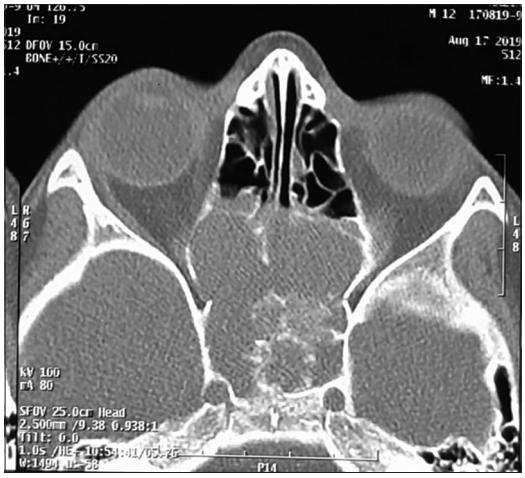

Figure 1: Computed tomography study with axial bone window showing expansile mass lesion in sphenoid and ethmoid sinus with irregular speckled areas of calcification, scalloping, and remodeling of bones.

DISCUSSION

NCMH is benign tumor consisting mainly of chondroid or cartilaginous tissues and mesenchymal elements that occurred in the nasal cavity. Hamartomatous lesions in the sinonasal tract may be epithelial, mesenchymal, or mixed.

In the sinonasal region, the spectrum of hamartomatous lesions is respiratory epithelial adenomatoid hamartoma, chondro-osseous respiratory epithelial adenomatoid hamartoma, and NCMH. NCMH is most commonly seen in the nasal cavity of children <3 months old with male predilection, but may be seen up-to-the second decade of life, and occasionally in adults.[1-8] Studies have shown genetic as well as non-genetic components to NCMH pathogenesis.[9]

The NCMH usually manifests as a polypoid expansile mass lesion occasionally showing paranasal, orbital, and intracranial extension without frank bone destruction.[8] Although benign, an NCMH may reveal bony remodeling and can raise suspicion for malignancy. Intracranial extension through the cribriform plate is not infrequent.[6,7] However, frank bony destruction as seen in malignant tumors is not a feature of NCMH.[10]

NCMH on CT is typically seen as a non-encapsulated and poorly defined mass often with or without cystic components; internal calcifications/chondroid matrix, and

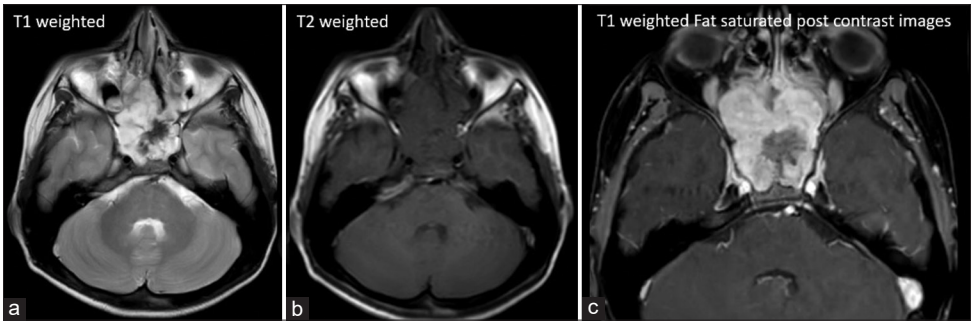

Figure 2: MRI study showed that mass lesion was homogeneously isointense to gray matter on T1-WIs, showed homogeneously hyperintense signals on T2-weighted axial images with central ill-defined hypointense signals. Lesion showed a diffuse heterogeneous enhancement with central as a non-enhancing region on post-contrast administration.

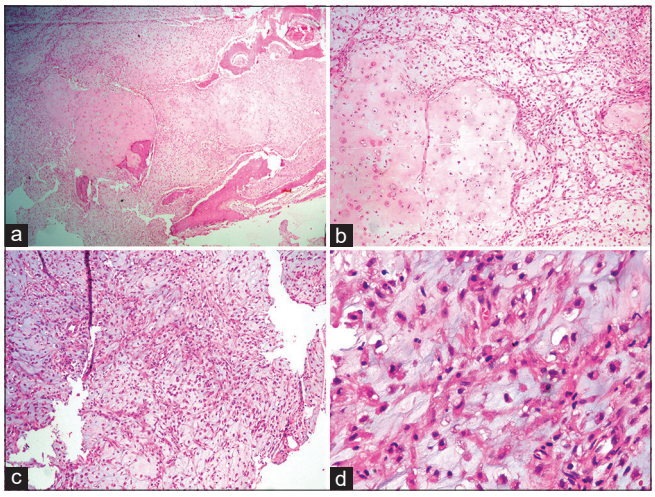

Figure 3: Hematoxylin-eosin (H and E). (a) Low-power view of spectrum of cartilaginous differentiation showing discrete lobules of hyaline cartilage interspersed with vague nodules of myxochondroid nodules and mesenchymal stromal cells and surrounding bony islands (hematoxylin-eosin, original magnification ×40). Medium-power view (b) showing lobules of hyaline cartilage interspersed with vague chondroid nodules with myxoid matrix blending imperceptibly with surrounding stromal cells (original magnification ×200). Medium-power view (c) showing predominantly chondromesenchymal elements composed of relatively cellular chondroid nodules with myxoid matrix interspersed with bland spindle cell stroma and blood vessels (original magnification ×200). High-power view (d) showing myxochondroid nodules with mildly increased chondrocytes and occasional binucleate chondrocytes without appreciable cytologic atypia or pleomorphism (original magnification ×400).

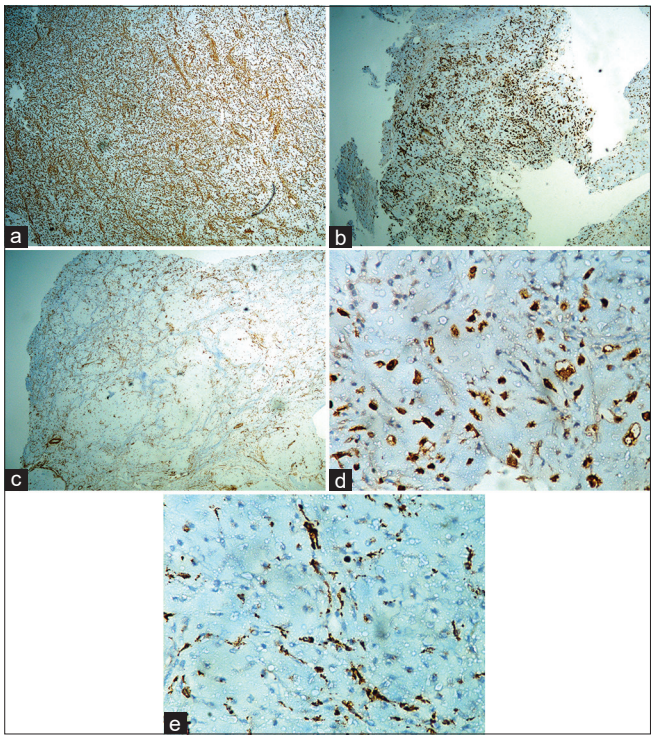

Figure 4: Immunohistochemistry: Vimentin (a) showing diffuse and strong expression in both cartilaginous and mesenchymal stromal components (VIMENTIN, original magnification ×40). Patchy epithelial membrane antigen (EMA) (b) expression in both cartilaginous and spindled stromal components (EMA, original magnification ×100). Smooth muscle actin (SMA) stain (c) showing cytoplasmic staining pattern of the stromal component (SMA, original magnification ×40) S100 stains (d) both cytoplasm and nuclei of the cartilaginous components (S100, original magnification ×400). CD68 (KP1) staining (e) shows rare expression in the spindled stromal areas (original magnification ×400).

very occasionally as non-enhancing mass.[6] On MRI, marked hyper-intensity on T2WI with no diffusion restriction is thought to be due to abundant stromal myxoid tissues with relatively low cellularity. Areas of calcification and chondroid matrix show corresponding low signal intensity on T2 and GEWI. In certain cases, the cystic change is so extensive that the tumor can mimic a meningoencephalocele and the latter can be distinguished based on a defect in the bone, without destruction of the anterior cranial fossa.[3] In some cases, intracranial extension through the cribriform plate in young adolescents needs careful evaluation to distinguish it from esthesioneuroblastoma.[5] Chondroma is rare in the pediatric age group and when an exception can closely mimic NCHM. Congenital salivary gland anlage tumor is another close mimic which is a benign midline lesion typically attached to the posterior septum or the posterior nasopharyngeal wall by a thin pedicle. Differential diagnosis of NCMH is listed in Table 1.

Table 1: The differential diagnosis of NCMH.

Nasal glioma
Rhabdomyosarcoma
Lymphoma
Chondroma, chondrosarcoma
Nasoethmoidal encephalocele and
Salivary gland anlage tumor

Microscopically, the NCMHs are a mixed morphological structure composed of varying proportions of proliferating mesenchymal and cartilaginous elements.[10]

Most areas with cartilage differentiation showed a vague nodular configuration and rarely appeared discretely nodular, although most nodules blended imperceptibly with the surrounding mesenchymal elements. The chondroid elements are arranged in lobules with intervening mesenchymal elements of collagen fibers, myxoid to spindle cell stroma, focal osteoclast-like giant cells, and erythrocyte-filled spaces resembling those of the aneurysmal bone cyst. The cases in which McDermott *et al.* initially described aneurysmal bone cyst-like areas as one of the characteristic patterns of NCMH were mainly newborn babies. However, the recently reported cases of adolescents and older age

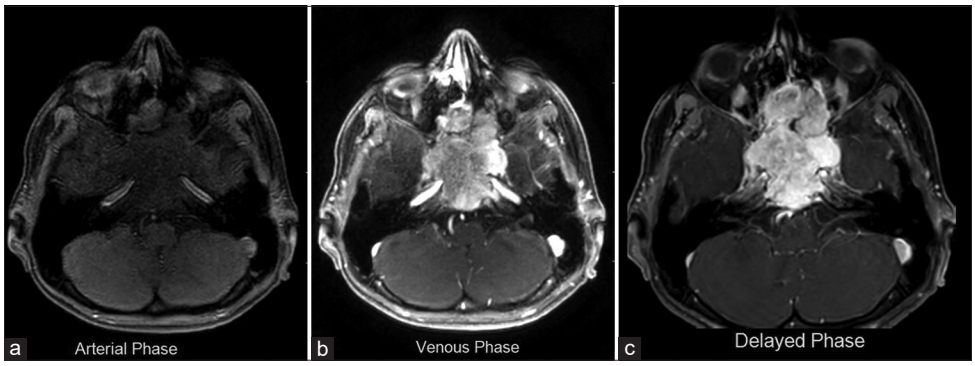

Figure 5: (a-c) In a known case of recurrent NCMH, lesion in posterior ethmoid and sphenoid sinus showed progressive enhancement on venous and more brilliant enhancement on delayed phase on dynamic contrast study.

patients contained no aneurysmal bone cyst-like areas as in our case.[1] The main histopathological differential diagnosis of NCMH is aggressive mesenchymal chondrosarcoma in which the mesenchymal cells are more cellular and small with hyperchromatic nuclei and peripheral condensed chromatin.[1]

CONCLUSION

We documented a recurrence of lesion of equal size within 2 years which could reflect a propensity for rapid growth from a residual microscopic tumor. Complete excision is usually curative. However, an incomplete resection might be followed by the persistent growth of a residual tumor. Multiple case reports have observed recurrence in association with DICER1 mutation.[8] We do not have confirmation of the DICER1 mutation in our case at the time of presentation.

TEACHING POINTS

Knowledge NCMH is essential to avoid potentially harmful therapies since it often simulates malignancy on imaging. Awareness of the imaging characteristics of NCMH can help in distinguishing it from other chondroid lesions of the sinonasal region. NCMH's association with DIECR1 mutation has very recently been established, and therefore in light of this, any patient with a DICER1-related tumor spectrum and new sinonasal mass should raise suspicion of NCMH. Furthermore, surgeons need to be vigilant for its recurrence and associated DICER1-related tumors, as NCMHs may be the earliest presentation for this disease spectrum.

MCQs

1. Which of the following are DICER1-related tumors?
    a. Pleuropulmonary blastoma.
    b. Juvenile granulosa cell tumors.
    c. Cystic nephroma, renal sarcoma, and Wilms tumor.
    d. Sinonasal chondromesenchymal hamartoma.
    e. All of the above.

Answer Key: e

2. Which of the following are hamartomatous lesions of sinonasal region?
    a. Mesenchymal chondrosarcoma.
    b. Respiratory epithelial adenomatoid hamartoma.
    c. Sinonasal chondromesenchymal hamartoma.
    d. Chondro-osseous respiratory epithelial adenomatoid hamartoma.
    e. All of the above.

Answer Key: e

3. Imaging features of NCMH are
    a. Polypoidal mass lesion with bony remodeling/ pressure erosion.
    b. Often show chondroid matrix with variable enhancement.
    c. Tends to cause locoregional mass effect in cases of large masses.
    d. All of the above.

Answer Key: d

Declaration of patient consent

Patient's consent not required as patient's identity is not disclosed or compromised.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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