CASE REPORT

Journal of Otorhinolaryngology and Facial Plastic Surgery. 2023;9(1):1-5.

https://doi.org/10.22037/orlfps.v9i1.44710

Nasal Chondromesenchymal Hamartoma, a rare Tumor in Young Children: A Case Report

Nasim Raad[1,2], Jahangir Ghorbani[1,2*], Mahsa Rekabi[3], Maryam Yousefi Koosha[2], Mahboobeh Karimi-Galougahi[1,2]

1. Chronic Respiratory Disease Research Center, National Research Institute of Tuberculosis and Lung Diseases (NRITLD), Shahid Beheshti University of Medical Sciences, Tehran, Iran.
2. Department of Otolaryngology, Masih Daneshvari Hospital, Shahid Beheshti University of Medical Sciences Tehran, Iran.
3. Pediatric Respiratory Diseases Research Center, National Research Institute of Tuberculosis and Lung Diseases (NRITLD), Shahid Beheshti University of Medical Sciences, Tehran, Iran.

Article Info

Article Note:
Received: November 2023
Accepted: December 2023
Publish Online: December 2023

***Corresponding Author**:
Dr Jahangir Ghorbani

Email:
jjghorbani@gmail.com

Keywords:
Nasal neoplasm;
Chondromesenchymal hamartoma;
Paranasal sinuses

Abstract

Background: Nasal chondromesenchymal hamartoma (NCMH) is a very rare benign tumor of sinonasal tract with mixed components of predominantly mesenchymal and cartilaginous tissues which is mostly seen in infants and young children.

Case presentation: We present the case of an infant with a nasal chondromesenchymal hamartoma which was confused with a malignancy. This clinicopathologic entity is notable because of its rarity and also it may present a diagnostic dilemma with major implications in management.

Discussion: Nasal chondromesenchymal hamartomas (NCMHs) are rare, mostly benign lesions found in the nasal cavity and paranasal sinuses. They can be locally destructive and may be mistaken for malignant tumors due to their aggressive appearance. NCMHs consist of various lobulated mesenchymal components, with areas of fibro-osseous proliferation. They can cause nasal mass, obstruction, respiratory distress, maxillary bulge, or proptosis, and may extend to the adjacent paranasal sinuses. Radiological imaging is essential for differential diagnosis and evaluating surrounding tissues. Complete surgical excision is sufficient therapy for NCMHs.

Conclusion: We present a rare case of NCMH, which can be difficult to distinguish from malignant tumors. Although NCMH are benign lesions, there is a chance of recurrence following incomplete resection. Recurrence may mimic malignancy, and their differentiation is important for choosing appropriate treatment.

Conflict of Interest: The authors declare no conflict of interest.
Please cite this article as: Raad N, Ghorbani J, Rekabi M, Yousefi Koosha M, Karimi-Galougahi M. Nasal Chondromesenchymal Hamartoma, a Rare Tumor in Young Children: A Case Report. J Otorhinolaryngol Facial Plast Surg. 2023;9(1):1-5. https://doi.org/10.22037/orlfps.v9i1.44710

Background

Nasal masses in infancy are infrequently encountered. Most are developmental anomalies such as encephaloceles, gliomas and nasolacrimal duct cysts. The rest are neoplasms which are more frequently reticuloendothelial, neural or mesenchymal in origin as opposed to the predilection for epithelial neoplasms in adults (1). Nasal chondromesenchymal hamartoma (NCMH) is a very rare, benign tumour originates from nasal cavity and/or paranasal sinuses and despite of the most common sinonasal tumours that are epithelial in nature, these tumours have a mixed morphological structure comprised of predominantly mesenchymal and cartilaginous components (2).

Case Presentation

Journal of Otorhinolaryngology and Facial Plastic Surgery. 2023;9(1):1-5.
https://doi.org/10.22037/orlfps.v9i1.44710

A one-year-old girl presented to our outpatient clinic with painless facial swelling and proptosis over a period of two months. Swelling was on the left lateral wall of her nose and left maxillary sinus area and had been growing insidiously over time. The child also was a mouth breather. Her prior medical history was unremarkable. Examination revealed a non-tender, skin intact facial swelling located as described above and nasomaxillary groove seemed to become shallow. Anterior rhinoscopy showed a large, non- vascular submucosal mass originating from lateral wall of left nasal cavity, totally filling the cavity and pushing septum to contralateral side. Her left eye was proptosed and deviated laterally without limitation in movement. Left ear otoscopy showed effusion behind an intact ear drum. No palpable cervical lymphadenopathy was identified.

The child underwent further evaluation with imaging. Computed tomography (CT) scan and magnetic resonance imaging (MRI) demonstrated an expansile heterogenous mass involving left maxillary and ethmoid sinuses with extension to nasal cavity and deformity left orbital floor. Thinning of adjacent bones due to pressure remodelling without evidence of destruction noted on CT (Figure 1).

The mass was hypointense on T1-weighted images and hyperintense and heterogeneous on T2 weighted images are presented in Figure 2.

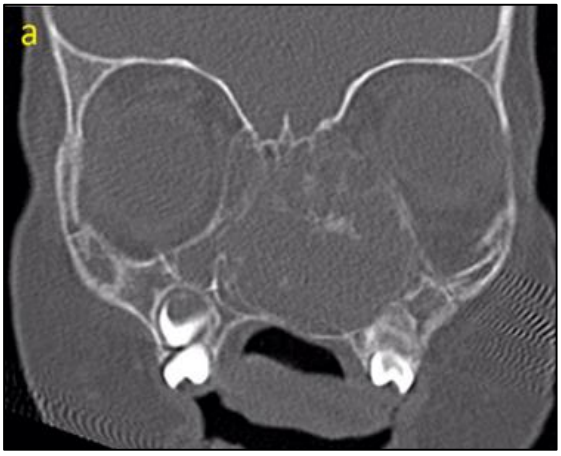

Figure 1. Coronal CT scan shows a huge, lobulated and well defined soft tissue mass filling both nasal cavities and anterior ethmoidal sinus with displacement of left eyeball. Thinning of adjacent bones caused by pressure remodelling is observed without evidence of frank destruction. Direct extension of lesion to intracranial and oral cavities is not evident.

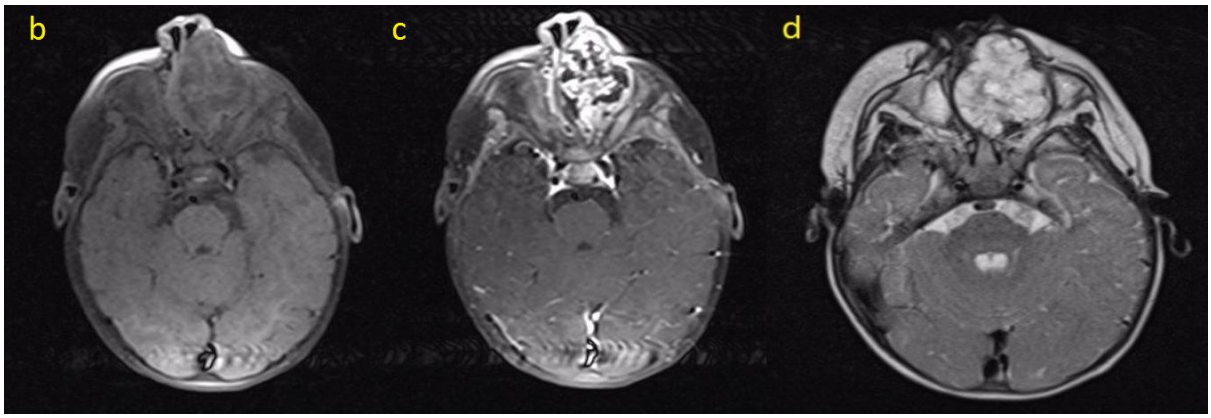

Figure 2. (b) Magnetic resonance imaging in axial cut showing in T1 weighted sequence a large, lobulated and mildly hypointense mass filling left maxillary sinus and both nasal cavities (c) Axial contrast-enhanced T1-wieghted MRI demonstrates heterogeneously enhancing intranasal mass filling total nasal cavity and left maxillary sinus (d) An axial T2-weighted MRI shows well-defined, huge soft tissue hyperintense mass with strong signal filling total nasal cavity and left maxillary sinus.

Biopsy of mass under general anesthesia was performed. On histopathologic analysis, the mass was consisted of nodules of cartilage varying in size and shape with different degree of maturation, surrounded by a loose spindle cell stroma. With immunohistochemical examination, diagnosis of the mass was reported as nasal chondromesenchymal

Journal of Otorhinolaryngology and Facial Plastic Surgery. 2023;9(1):1-5.
https://doi.org/10.22037/orlfps.v9i1.44710

hamartoma. We planned for endoscopic surgical resection of mass. A gross total resection of mass from nasal cavity and involved sinuses associated with resection of inferior turbinate was performed and involved Lamina papyracea and floor of orbit was removed. No further treatment was done and patient was discharged from hospital on the second postoperative day. However, she came back with nasal blockage, facial deformity and severe proptosis after one year. At that time there was two schools of thought, whether encountering a sinonasal malignancy in a two years old child or recurrence of previously reported chondromesenchymal hamartoma. This time it was planned for combined endoscopic and open surgical approaches to remove tumour from maxillary, ethmoid sinuses, floor and medial wall of orbit. Tumour resected completely by performing medial maxillectomy via lateral rhinotomy incision. The rest of lamina papyracea till orbital apex and floor of orbit was removed. Extension of tumour to upper part of pterygomaxillary fissure was noted which also resected. Titanium mesh was used for reconstruction of orbital rim and floor. The second histopathological report also confirmed the chondromesenchymal nature of mass.

During follow up, there was no clinical or radiological evidence of recurrence after 6 years (Figure 3).

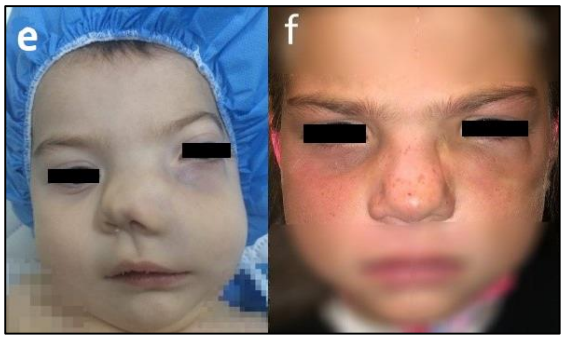

Figure 3. shows Clinical photography of the patient: (e) Pre operation (f) 6 years after operation**.**

Discussion

NCMHs are rare predominantly benign but locally destructive lesions involving the nasal cavity and paranasal sinuses and because of their aggressive appearance can be mistaken for a malignant tumour (3). Although nasal chondromesenchymal hamartoma is usually seen in infants and children, cases of adolescents and even older people have been reported(4).

The etiology of NCMH is thought to be due to an underlying genetic predisposition therefore accounting for the early presentation in the majority of cases (5). On histopathological examination, nasal chondromesenchymal hamartoma consists of focal lobulated various mesenchymal components. The major components are irregular, mature and immature hyaline cartilage islands(6).

Similar to other case reports in the literature, our histopathological findings revealed nodules of cartilage varying in size and shape with different degree of maturation, surrounded by a loose spindle cell stroma. There were areas of fibro-osseous proliferation including fibro-collagenization and chondral ossification. There was no significant mitotic activity or necrosis. Fibro-osseous dysplasia, chondosarcoma and osteosarcoma should be considered in histopathological differential diagnosis. However, mature and immature cartilage islands are not seen in fibro-osseous dysplasia and dissimilar to chondosarcoma and osteosarcoma there were no signs indicating malignancy.

NCMHs can be slow growing and therefore have a delayed presentation. Clinical features vary in relation to the size and location of lesion and include nasal mass, nasal obstruction, respiratory distress, a maxillary bulge or proptosis due to orbital involvement which all has been noted in our patient. Also intracranial extension of the tumour can result in neurologic deficits. NCMH can extend to the adjacent paranasal sinuses. The most frequently involved paranasal sinus is the ethmoid sinus (7).

Radiological imaging is essential for differential diagnosis and for evaluating surrounding tissues. NCMHs may be non-encapsulated and ill defined. The adjacent paranasal sinuses are frequently involved and

Journal of Otorhinolaryngology and Facial Plastic Surgery. 2023;9(1):1-5.
https://doi.org/10.22037/orlfps.v9i1.44710

remodelling, thinning or erosion of the surrounding bone are not uncommon(8, 9), as has been seen in our case.

NCMH may mimic malignant tumours in its clinical, radiologic or even histopathologic presentation. Differentiation between NCMH and malignant tumours is very important in view of making the best decision for their treatment. Complete surgical excision is sufficient therapy for these tumours unlike excessively radical surgery and possibly neo-adjuvant radiotherapy for malignancies. Although, nasal chondromesenchymal hamartoma has been considered as a benign lesion, the possibility of malignant transformation should be kept in mind (9, 10).

Recurrences have been reported following incomplete resections. In these cases radiotherapy and combined chemotherapy is effective in regression of residual tumour when complete resection is not possible. Adjuvant therapy is not recommended after complete resection(11).

Conclusion

We present a rare case of NCMH and highlight the differentiation between these naturally benign tumours and a malignant one may be difficult clinically, radiologically and histopathologically, however the recurrence cases should not be interpreted as a malignancy since there is low chance of recurrence following incomplete resection and presence of residual tumoural tissue. As in all head and neck tumours, treatment of benign and malignant sinonasal tumours is two totally different entities with different outcomes.

Acknowledgements

Not declared.

Patient Consent

Written informed consent was obtained from the patient for publication and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Conflict of Interest

The authors declared no conflicts of interest.

Financial Support

The authors declared that there was no financial support.

Authors' ORCHIDs

Nasim Raad
https://orcid.org/0000-0003-1372-3710
Jahangir Ghorbani
https://orcid.org/0000-0001-6209-1081
Mahsa Rekabi
https://orcid.org/0000-0003-1187-3555
Maryam Yousefi Koosha
https://orcid.org/0000-0002-5954-5166
Mahboobeh Karimi-Galougahi
https://orcid.org/0000-0001-9880-3802

References

1. Estimar RS, Zafra MAM, Lopa RAB. Congenital Nasal Chondromesenchymal Hamartoma. Philippine Journal of Otolaryngology Head and Neck Surgery. 2009;24(2):23-6.

2. Thirunavukkarasu B, Chatterjee D, Mohindra S, Dass Radotra B, Prashant SJ. Nasal Chondromesenchymal Hamartoma. Head and neck pathology. 2020;14(4):1041-5.

3. Cho YC, Sung IY, Son JH, Ord R. Nasal chondromesenchymal hamartoma: report of a case presenting with intraoral signs. Journal of oral and maxillofacial surgery : official journal of the American Association of Oral and Maxillofacial Surgeons. 2013;71(1):72-6.

4. Nakagawa T, Sakamoto T, Ito J. Nasal chondromesenchymal hamartoma in an adolescent. International Journal of Pediatric Otorhinolaryngology Extra. 2009;4(3):111-3.

5. Bahubeshi A, Bal N, Rio Frio T, Hamel N, Pouchet C, Yilmaz A, et al. Germline DICER1 mutations and familial cystic nephroma. Journal of medical genetics. 2010;47(12):863-6.

6. Johnson C, Nagaraj U, Esguerra J, Wasdahl D, Wurzbach D. Nasal chondromesenchymal hamartoma: radiographic and histopathologic analysis of a rare pediatric tumor. Pediatric radiology. 2007;37(1):101-4.

7. Sarin V, Singh B, Prasher P. A silent nasal mass with ophthalmic presentation. Orbit (Amsterdam, Netherlands). 2010;29(6):367-9.

8. Norman ES, Bergman S, Trupiano JK. Nasal chondromesenchymal hamartoma: report of a case and review of the literature. Pediatric and developmental pathology : the official journal of the Society for Pediatric Pathology and the Paediatric Pathology Society. 2004;7(5):517-20.

Journal of Otorhinolaryngology and Facial Plastic Surgery. 2023;9(1):1-5.

https://doi.org/10.22037/orlfps.v9i1.44710

9. Mason KA, Navaratnam A, Theodorakopoulou E, Chokkalingam PG. Nasal Chondromesenchymal Hamartoma (NCMH): a systematic review of the literature with a new case report. Journal of otolaryngology - head & neck surgery = Le Journal d'oto-rhino-laryngologie et de chirurgie cervico-faciale. 2015;44(1):28.

10. Li Y, Yang QX, Tian XT, Li B, Li Z. Malignant transformation of nasal chondromesenchymal hamartoma in adult: a case report and review of the literature. Histol Histopathol. 2013;28(3):337-44.

11. Mattos JL, Early SV. Nasal chondromesenchymal hamartoma: A case report and literature review. International Journal of Pediatric Otorhinolaryngology Extra. 2011;6(4):215-9.