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Case Report

Nasal chondromesenchymal hamartoma causing sleep-disordered breathing in an infant

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Abstract: Nasal chondromesenchymal hamartoma is an extremely rare neoplasm of the nasal and paranasal sinuses. We present the case of a 10-month-old boy with a huge nasal chondromesenchymal hamartoma that was complicated by sleep-disordered breathing. The mass was completely resected by image-guided endoscopic surgery and confirmed histopathologically as a chondromesenchymal hamartoma. In this report, we discuss the characteristics and treatment of this unusual tumor.

Keywords: Chondromesenchymal hamartoma, nasal mass, endoscopic excision, sleep-disordered breathing, infant

Introduction

Infants are vulnerable to sleep-disordered breathing (SDB) due to both anatomical and physiological predispositions. Adeno-tonsillar hypertrophy, laryngomalacia, stenosis of glottis or subglottic lesion, craniofacial anomalies and excessive airway secretion can cause SDB in infants [1]. In addition, infants with huge nasal mass are at risk of developing airway obstruction [1, 2]. Nasal chondromesenchymal hamartoma is a rare benign tumor that typically presents as a unilateral nasal mass in infant and children [3]. Complete resection is the treatment of choice, and treatment modalities depend on the size and location of the tumor as well as involvement of surrounding structures [4, 5]. In this report, we present a rare case of huge nasal chondromesenchymal hamartoma causing SDB that was successfully treated with image-guided endoscopic surgery.

Case report

A 10-month-old boy presented with a 6-month history of nasal congestion and mouth breathing. Recently, he showed loud snoring, episodes of breathing cessation witnessed by parents, irritability, and sternal recession during sleep. On physical examination, his nasal dorsum was widened and the area of the right nasolacrimal groove was swollen. Nasal endoscopic examination revealed a non-pulsatile, huge mass completely obstructed the right nasal cavity (**Figure 1**).

Computed tomography scan revealed a large, minimally enhancing homogeneous mass in the right nasal cavity. The mass filled the right nasal cavity completely, displaced the nasal septum, and extended to the right cribriform plate superiorly and palate inferiorly. Posterior choana was nearly obstructed by the mass. Remodeling and expansion of the adjacent bony structures were seen. Magnetic resonance images showed a well-demarcated expansile mass in the right nasal cavity. The mass had low signal intensity on T1-weighted images and high signal intensity on T2-weighted images. After administration of contrast medium, multifocal mottled enhancement within the lesion and peripheral outer enhancement were demonstrated. Intracranial extension was not identified (**Figure 2**).

Surgical excision via endoscopic approach was done. High definition endoscopic system (Conmed, Linvatec, Utica, NY, USA), navigation system (Fusion, Medtronic, USA), and pediatric

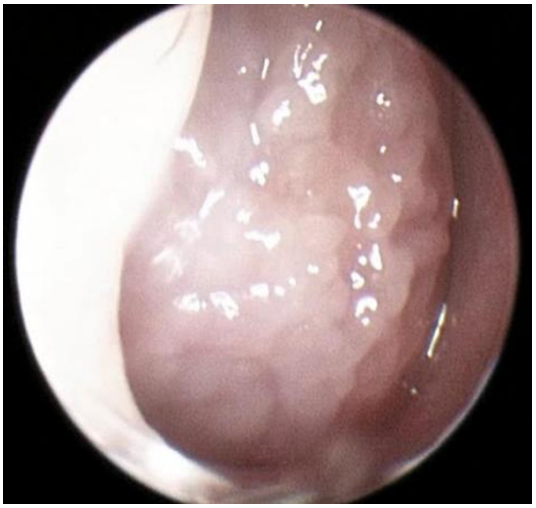

Figure 1. Nasal endoscopic examination shows a non-pulsatile, huge polypoid mass in the right nasal cavity.

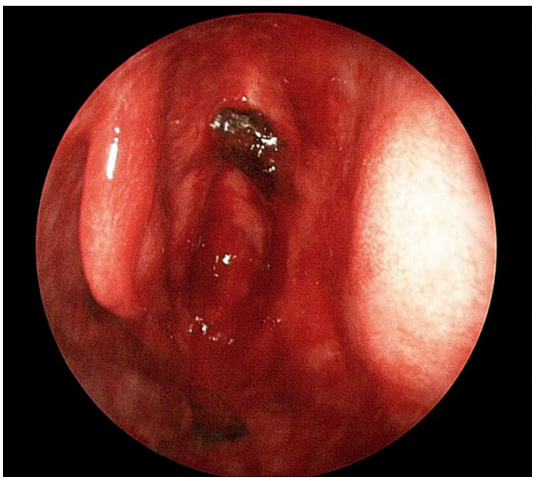

Figure 3. Intraoperative finding. The mass was originated from the roof of the right nasal cavity. Complete resection of the mass was achieved and electrocauterization was done at the site of origination.

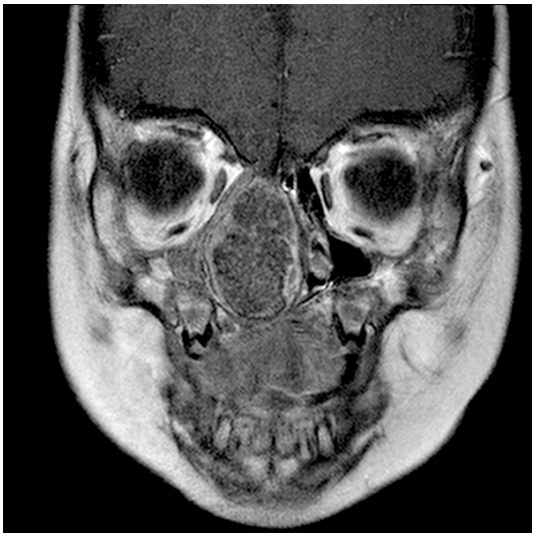

Figure 2. Coronal contrast-enhanced T1-wieghted MRI demonstrates heterogeneously enhancing intranasal mass filling the right nasal cavity. The mass is displacing the nasal septum. A huge mass filled total right nasal cavity, displaced the nasal septum, and extended to the right cribriform plate superiorly and palate inferiorly. Posterior choana was nearly obstructed. There is no intracranial extension.

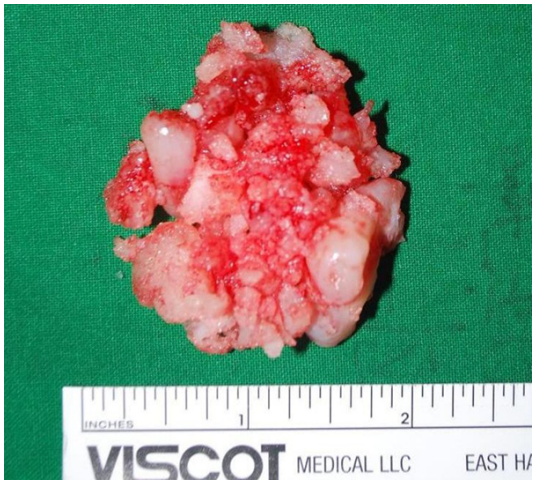

Figure 4. Gross appearance of the specimen. The mass was excised in a piecemeal manner. The specimen was consisted of multiple pieces of pink-tan colored soft tissue fragments.

endoscopic equipment were used. Intraoperatively, the mass was solid and appeared to originate from the roof of the right nasal cavity. But it was easily freed from the septum, the nasal floor and the middle turbinate. Resection of the mass was carried out in a piecemeal manner with microdebrider and cutting forceps. No significant bleeding occurred during the operation, and complete resection of the tumor mass was achieved (**Figures 3**, **4**). The recovery of the patient was uneventful, and he was discharged a day later. The patient's sleep disturbance was completely resolved.

Histopathologic examination showed multiple, irregular islands of chondroid tissue within spindle cell pattern with occasional binucleated mesenchymal cell proliferation (**Figure 5**). The spindle cells possessed reactivity for vimentin, S-100 protein, smooth muscle actin, epithelial membrane antigen, and myelin basic

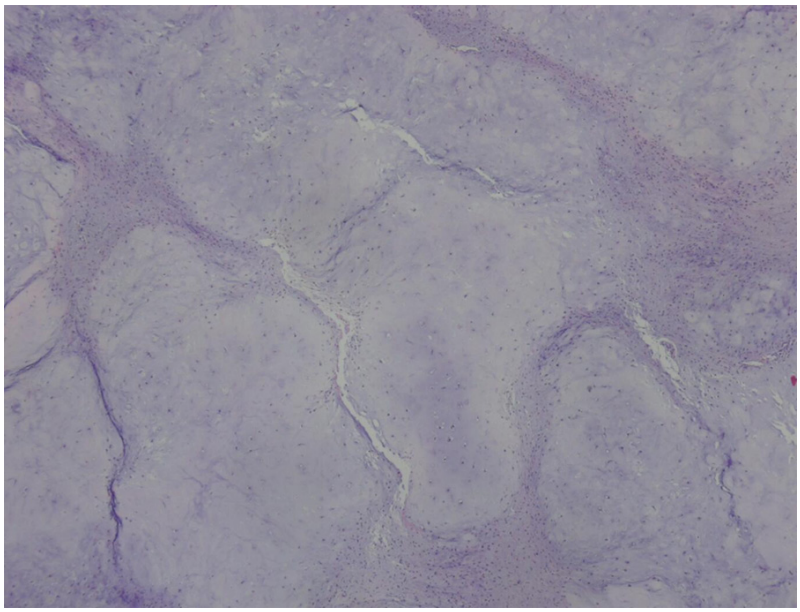

Figure 5. Histopathologic examination shows multiple, irregular islands of chondroid tissue within spindle cell stroma. Sheets of chondroid material with foci of mature and immature cartilage are seen (H&E stain, ×100).

protein were negative in immunohistochemical studies. At 18-month follow-up, there was no evidence of recurrence.

Discussion

A hamartoma is a malformation that results from excessive growth of tissues indigenous to the site of origin. Hamartomas are common in the skin, lung, liver, chest wall, kidney, and gastrointestinal tract [6]. In the head and neck, hamartomas are unusual. In rare instances, they have been reported in the oral cavity, nasal cavity, nasopharynx, hypopharynx, tongue, eustachian tube, and larynx [3]. Nasal chondromesenchymal hamartoma was first reported by McDermott *et al.* in 1988, so far about 20 cases have been reported in the literature. It presents as an intranasal mass in infants and young children with a male preponderance [5, 7, 8].

The clinical manifestations depend on the location and size of the lesion. Nasal obstruction, feeding difficulties, rhinorrhea, epistaxis, visual disturbance, otitis media and facial deformity are the symptoms of nasal chondromesenchymal hamartomas in infants [4]. Orbital involvement of the tumor can result in proptosis, ptosis, enophthalmos, or impairment of eye movement. An intracranial extension can result in neurologic signs such as hydrocephalus and oculomotor disturbance [5]. In this case, sleep disturbance and snoring were the main problems. The huge size, septal displacement, posterior growth and accompanied by adenoid hypertrophy could be the causes of airway obstruction during sleep.

CT scanning and MRI are helpful in both characterization and description of anatomic extent or invasion. Also, they can help determine the tumor's site of origin and aid in planning resection surgery. CT scan shows a nonencapsulated heterogenous soft-tissue mass, which are predominantly solid and cystic, with or without calcification. Calcification may be an important diagnostic clue which may help differentiate chondromesenchymal hamartoma from other nasal mass lesions, but is not always specific. On MRI, the signal intensity and enhancement of chondromesenchymal hamartoma are variable from case to case depending on the composition of the tumor. The signal is either high or low, homogeneous or heterogeneous. And the enhancement of the contrast medium is proportional to the degree of the tumor vascularity. Wang et al. reported that the MRI findings of chondromesenchymal hamartoma are inhomogenous with low signal intensity on T1-weighted images and with high signal intensity on T2-weighted images with strongly inhomogenous contrast enhancement, which were consistent with the present case [9].

Although benign, the radiological appearance of the chondromesenchymal hamartoma can raise suspicion for malignancy as a result of the surrounding bone, cortical thinning, displacement, and intracranial extension [6, 9]. Upon imaging, the differential diagnosis for a pediatric sinonasal mass is broad, with all entities being rare. It includes hemangioma, angiofibroma, nasoethmoidal encephalocele, nasal glioma, inverted papilloma, giant cell reparative granuloma, ossifying fibroma, chon-

dro-osseous respiratory adenomatoid hamartoma, and aneurismal bone cyst as benign pediatric tumors. Malignant pediatric tumors such as rhabdomyosarcoma, esthesionneuroblastoma, and chondrosarcoma should be excluded [9].

Histopathological examination of this tumor shows irregular islands of mature and immature hyaline cartilage with occasional binucleated chondrocytes. The islands of cartilage are well demarcated from the surrounding stromal tissues, which have a myxoid background. And spindle cells with occasional mitotic figures are seen [6]. The histopathological differential diagnosis includes chondrosarcoma and osteosarcoma [5].

Currently, the treatment of choice for a chondromesenchymal hamartoma is complete surgical excision [4, 5, 8, 9]. Local resection via endoscopic approach is recommended if the lesion is confined to the nasal cavity. But depend on tumor size and extension, external approach (midfacial degloving, sublabial, frontal craniotomy, etc.) can be necessary [7].

In the present case, the huge tumor was completely removed via image-guide endoscopic surgery. Due to the patient's age, narrow nasal cavity, and relatively huge tumor size, it could be difficult for endoscopic surgery. Jeyakumar *et al.* described the use of image-guided system had limited benefit in endoscopic surgery for 4-day-old infant [8]. However in the present case, high resolution endoscope system and image-guided navigation system were very helpful for safe surgery and to decrease surgical time.

Local recurrence of nasal hamartoma is rare but possible. Recurrence of chondromesenchymal hamartoma has been documented in patients in whom the tumor had not been completely resected [5]. Adjuvant therapy is unnecessary if there is complete surgical resection [4, 5, 8].

Conclusion

Huge nasal chondromesenchymal hamartoma is an extremely rare condition that can account for obstructive sleep apnea in infants. Treatment of this tumor is complete excision of the mass. Regardless of the size, when the mass is localized to the nasal cavity, endoscopic resection should be considered as the first treatment option. The image-guided navigation system can be helpful during surgery.

Disclosure of conflict of interest

None.

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