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Case Report

Pediatric nasal septum pleomorphic adenoma: a case report

Jialian Feng, Yatian Liu, Jianguo He, Xiao Jiao

Department of Otolaryngology, The Fourth Affiliated Hospital, Zhejiang University School of Medicine, N1 Shangcheng Road, Yiwu 322200, Zhejiang, China

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Abstract: Pleomorphic adenoma (PA), the most common benign salivary gland tumor, is rarely found in the nasal cavity and paranasal sinuses, particularly in pediatric patients. This report presents a case of PA in the nasal septum of a 14-year-old girl who presented with unilateral epistaxis and progressive nasal obstruction. The tumor was excised from the left anterior nasal septum via endoscopic sinus surgery, and PA was confirmed through histopathological examination. This case emphasizes the importance of including PA in the differential diagnosis of pediatric sinonasal masses, despite its rarity, and underscores the necessity of meticulous surgical planning to prevent recurrence. Further studies are needed to better understand the long-term outcomes and optimal management strategies for this rare condition in children.

Keywords: Nasal cavity, diagnosis, endoscopic surgery, children

Introduction

Pleomorphic adenoma (PA) is the most common benign salivary gland neoplasm, primarily occurring in major salivary glands (80%), with less frequent occurrence in minor salivary glands (10-15%) [1]. Its presence in the sinonasal region is exceedingly rare, accounting for approximately 0.4% of PA cases [2]. Sinonasal PA (SNPA) presents significant diagnostic and therapeutic challenges due to its rare location and potential for misdiagnosis. Several studies have explored the pathogenesis of SNPA. Some suggest that these tumors may arise from ectopic salivary gland tissue or embryonic epithelial remnants in the nasal cavity and paranasal sinuses [3], while others propose a potential origin from the vestigial vomeronasal organ [4, 5]. However, the exact mechanisms remain unclear.

Clinically, SNPA typically manifests with nonspecific symptoms such as nasal obstruction, epistaxis, and, occasionally, facial swelling or pain [6]. These symptoms overlap with those of more common sinonasal conditions, which can delay diagnosis. Imaging techniques, such as computed tomography (CT) and magnetic resonance imaging (MRI), are essential for evaluating these tumors, defining their extent, and aiding in surgical planning [7].

Histopathologically, SNPA is characterized by a mixture of epithelial and myoepithelial cells embedded in a stromal background, which may include myxoid, chondroid, or fibrous components [1, 8]. Compared to PA in major salivary glands, SNPA typically exhibits higher cellular density and less stromal differentiation, which can complicate diagnosis, particularly in distinguishing SNPA from other benign and malignant sinonasal tumors.

The primary treatment for SNPA is complete surgical excision. Endoscopic methods are increasingly preferred due to their minimally invasive nature and lower associated morbidity [9]. Achieving complete resection is crucial to reduce the risk of recurrence, which is primarily linked to incomplete initial excision [10]. Long-term monitoring is necessary to check for recurrence, given the pleomorphic nature of the tumor and its potential for late recurrence.

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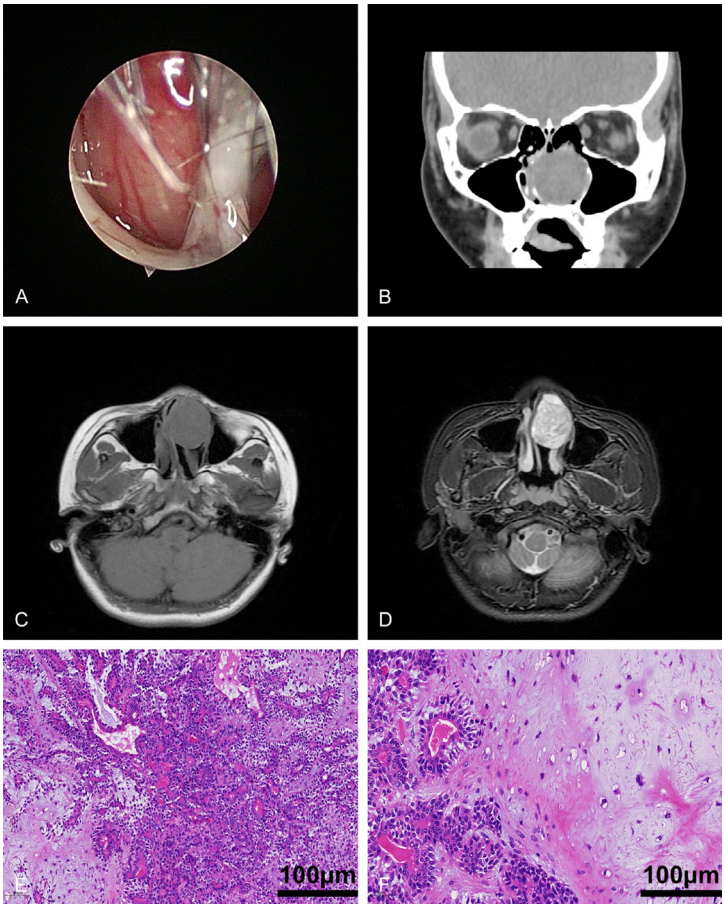

Figure 1. Nasal endoscopy showed that a smooth, pink mass with a rich vascular supply. (A) CT imaging showed a neoplasm in the left nasal cavity measuring 3.5 × 2.5 cm, with notable compression and rightward deviation of the nasal septum. (B) MRI demonstrated the soft tissue mass occupying the left nasal cavity. T1-weighted imaging (T1WI) showed a low signal intensity (C), while T2-weighted imaging (T2WI) displayed a high signal intensity (D). (E) HE stain, ×100 magnification, and chondroid stroma (F), HE stain, ×200 magnification.

Case report

A 14-year-old female patient presented with unilateral epistaxis persisting for 2 years and unilateral nasal obstruction for 1 year. She also reported a runny nose, teary eyes, occasional sneezing, and a mild decrease in her sense of smell, though she was otherwise healthy with no significant medical history. Clinical examination revealed swelling of the left external nose. Nasal endoscopy identified a smooth, pink mass with a rich vascular supply in the left nasal cavity, which bled easily upon contact (**Figure 1A**).

CT imaging of the paranasal sinuses showed an oval-shaped soft tissue mass in the left nasal cavity, measuring approximately 3.5 × 2.5 cm, with the nasal septum deviated to the right. The paranasal sinuses were well-developed with no signs of abnormal densities or bony destruction (**Figure 1B**).

MRI revealed a round mass in the left nasal cavity measuring 3.7 × 3.1 × 2.4 cm. The mass showed low signal intensity on T1-weighted images (**Figure 1C**), heterogeneous high signal intensity on T2-weighted images, and iso-high signal intensity on diffusion-weighted imaging. The nasal septum appeared compressed and deviated to the right (**Figure 1D**).

The patient underwent endoscopic resection of the nasal septal mass under general anesthesia. Intraoperatively, a pedunculated, round, pink mass with a smooth surface and rich vascularity was identified, originating from the anterior part of the left nasal septum.

Histopathological examination revealed a tumor composed of glandular epithelial and myoepithelial components, interspersed with abundant mucinous and chondroid stroma (**Figure 1E**, **1F**). The mucinous stroma predominated, with no tumor capsule observed, which is typical in cases with extensive mucinous components. Myoepithelial cells formed duct-like structures secreting eosinophilic material, lined by luminal cells and several layers of non-luminal cells merging into the mucinous stroma. These cells exhibited diverse morphology, with areas of squamous metaplasia noted. The tumor stroma was primarily composed of mucinous, chondroid, and hyaline-like components. Based on these findings, a diagnosis of PA was established.

The patient's postoperative course was uneventful, and follow-up one year after surgery showed no complications or signs of recurrence.

Discussion

PA, a neoplasm characterized by its varied histological profile - consisting of neoplastic epithelial tissue interspersed with mucous or chondroid components - is commonly referred to as a mixed tumor. This tumor predominantly occurs in the major salivary glands, such as the parotid, submandibular, and sublingual glands, while its occurrence in minor salivary glands is relatively rare [9, 11, 12]. PA frequently manifests in the palatal region, but documented cases also occur in the nasal cavity, external auditory canal, larynx, pharynx, trachea, and lacrimal glands [13]. Malignant transformation of PA in the nasal cavity is considered rare. In the nasal cavity, PA typically arises from the bony or cartilaginous sections of the nasal septum, with approximately 20% originating from the lateral nasal wall [14-16]. Instances in the nasal sinuses are generally attributed to secondary spread.

PA in the sinonasal region is uncommon, with the majority of cases reported in women between their third and sixth decades of life. The occurrence of SNPA in children is exceedingly rare, with a review of 39 cases showing a mean age at diagnosis of 50.5 years (range: 21-80 years) [9]. The slow progression and benign nature of PAs often lead to delays in diagnosis [17]. Studies indicate that the rate of malignant transformation in PAs of major salivary glands is approximately 6% [12]. If a PA exhibits a shift from slow to rapid growth or becomes painful, the risk of malignant transformation should be considered. Initial diagnosis can be supported by CT and MRI scans to detect signs of bone destruction or resorption, but a definitive diagnosis requires histopathological examination [7, 10].

In pediatric cases, nasal masses present a broad differential diagnosis, ranging from benign conditions like juvenile nasopharyngeal angiofibroma to malignant entities such as rhabdomyosarcoma. Given the variability in presentation, histopathological confirmation remains essential for a definitive diagnosis [18].

Histological examination revealed a tumor composed of epithelial, myoepithelial, and mesenchymal components within a fibrous stroma. The variability of the stroma, often featuring myxoid and chondroid areas, is characteristic of PA. Although the tumor appeared encapsulated during surgery, histopathology showed no distinct capsule, underscoring the need for thorough excision [9, 18, 19]. The histological diagnosis of PA can be confirmed through immunohistochemical staining, marked by positive expression of cytokeratin, vimentin, S100 protein, smooth muscle actin (SMA), and glial fibrillary acidic protein (GFAP) [20, 21]. Unfortunately, this patient did not undergo these tests.

Complete surgical resection with negative margins is the preferred treatment for PA. Surgical options include endonasal endoscopic resection, midfacial degloving, lateral rhinotomy, and partial maxillectomy. In this case, an endoscopic approach was chosen, reflecting current best practices for benign sinonasal lesions. Endoscopic surgery offers enhanced visualization, enabling precise excision while preserving adjacent structures, thereby reducing patient morbidity and facilitating faster recovery [15, 22]. In another case, a patient also underwent endoscopic surgery, with no significant adverse reactions post-treatment [23]. Endoscopic surgery is minimally invasive and provides a clear surgical field. For pediatric patients, when the tumor is localized within the nasal cavity and sinuses, complete resection under nasal endoscopy may be the optimal treatment option. However, due to the potential for recurrence and malignant transformation, postoperative follow-up is essential.

Recurrence rates for benign PA are generally low. Compagno et al. reported that intranasal mixed tumors have a recurrence rate of approximately 10% [1], highlighting the need for long-term follow-up, particularly in cases with positive margins or incomplete resection. Rha et al. found that one out of seven patients (14.3%) experienced recurrence within an average follow-up period of 34.4 months [24]. Regular follow-ups with imaging, such as MRI or CT scans, are recommended for early detection of potential recurrences [25].

In conclusion, we report a rare case of a 14-year-old female with a nasal PA, a condition seldom seen in children. This case underscores the diagnostic and therapeutic challenges associated with managing benign salivary gland tumors in uncommon locations.

Disclosure of conflict of interest

None.

Address correspondence to: Xiao Jiao, Department of Otolaryngology, The Fourth Affiliated Hospital, Zhejiang University School of Medicine, N1 Shangcheng Road, Yiwu 322200, Zhejiang, China. Tel: +86-18314952140; E-mail: 8017057@zju.edu.cn

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