PLEOMORPHIC ADENOMA OF THE NASAL SEPTUM: A RARE CASE REPORT OF A 14 YEAR-OLD PATIENT

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ABSTRACT

Pleomorphic adenoma is the most common benign tumor of the salivary glands. It arises mainly from the parotid, less frequently from the minor salivary glands of the oral cavity and, exceptionally, from the accessory glands widespread in the upper aero-digestive tract. Intranasal localization is rare and mainly affects the glands sited in the nasal septal mucosa. There are isolated case reports in literature regarding adult patients. We report a rare case of a young 14-year-old girl with pleomorphic adenoma arising from the nasal septum.

1. Introduction

Pleomorphic adenoma is the most common benign tumor of the salivary glands. It occurs mostly in the major salivary glands, particularly the parotid. It may also involve, less frequently, the minor salivary glands of the lips¹ and hard and soft palate². Cases have been reported in the upper respiratory tract, the most favorite site of origin is the nasal cavity, followed by the maxillary sinus and the nasopharynx³.

Other unusual sites, reported in literature, include the lacrimal gland⁴ and the external auditory canal⁵. Nasal pleomorphic adenoma may originate from the nasal septum mucosa, even though the vast majority of the minor salivary glands are located within the lateral nasal wall and especially in the turbinates².

We present a rare case of a 14-year-old girl with pleomorphic adenoma arising from the nasal septum.

2. Case Report

A 14-year-old girl presented a 6-month history of progressive unilateral left-sided nasal obstruction. A history of allergic rhinitis and local steroid treatment was reported. There was no history of nose bleeding, previous trauma to the nose, visual defect, pain or fever. The general health was satisfactory and her weight was stable.

Rhinoscopy revealed a large, elastic-hard mass filling the left nasal cavity, arising from the nasal septum with a broad base. It was normal mucosa covered without ulceration or bleeding on touch or spontaneously (Figure 1a and b). The right nasal cavity and the nasopharynx were clear. There were no palpable lymph nodes of the neck. No imaging was done preoperatively because the whole lesion was visible.

The tumor was removed by a trans-nasal sub-perichondral resection under general anesthesia, by emitrashfixion incision on caudal border of septal cartilage. It was totally excised; no piece of septal cartilage was removed because it didn't involve a tumor. The resection margins were free from tumor invasion.
Histological examination revealed the presence of two components: epithelial and mesenchymal, and thus the mixed aspect of the lesion led to the diagnosis of pleomorphic adenoma. Her post-operative course was uneventful. No clinical and/or radiological recurrence had occurred 5 years after operation.

Moreover a viral involvement has been discussed, Malinvaud et al. reported three cases with relationship between pleomorphic adenoma of the nasal septum and Epstein Barr virus.

The first case of intranasal pleomorphic adenoma reported in literature was in 1929.

Nasal pleomorphic adenoma is extremely rare, although three large series have been reported by Spiros et al. with 40 cases, by Compagno and Wong with 40 cases and by Suzuky et al. with 41 cases. Incidence is predominantly female and usually between the third and fifth decades of life.

A typical symptom is unilateral nasal obstruction which gradually worsens due to the slow growth of the tumor, as our patient reported in her anamnesis. Less common symptoms may appear and they are related to the size of the mass and to the position occupied inside the nasal cavity; they include epistaxis, external swelling of nasal pyramid, epiphora and mucopurulent rhinorrhea. Our patient may have had a delay in diagnosis due to a comitant history of allergic rhinitis that caused nasal obstruction.

Nasal pleomorphic adenoma appears as a unilateral mass, directly attached to the septum or to the lateral nasal wall, with polypoid aspect, pale or with pinkish-grey coloration, with a smooth surface and a firm or soft consistency. The sizes are variable in a range from <0.5 cm to >7 cm. The clinical features of the mass may denote a benign nature, in particular the smooth surface without signs of ulceration, no bleeding on touch, and no invasion of surrounding structures.

Histologically, nasal pleomorphic adenoma is a mixed tumor composed of two components: epithelial and myoepithelial elements and a quota of mesenchymatous stroma. It differs from pleomorphic adenoma of the major salivary glands for greater cellularity and predominance of epithelial component on stromal elements. This can lead to misdiagnosis as a carcinoma. It can present a capsule with variable thickness and it has a tendency to grow encapsulated.

Immunohistochemical staining shows the mixed nature of the lesion. It is performed using antibodies of various citokeratins, S100 protein, glial fibrillary acid protein (GFAP), α-smooth muscle actin (SMA), vimentin and the Ki67 proliferation marker.

Definite diagnosis of the pleomorphic adenoma is based on histological examination and immunohistochemical.

Radiological exams as CT or MRI are not specific for diagnosis of pleomorphic adenoma; however they are useful in case of very large lesions in the preoperative phase to identify the margins of the mass, the invasion of surrounding structures and possible bone destruction. The diagnosis is often accidental during endoscopic examination.

When a mass in the nasal cavity is found, the possibility of it being a pleomorphic adenoma must be considered and a differential diagnosis with benign lesions should be made, such as nasal polyps, papillomas, inverted papilloma, osteomas and angiofibromas and malignant lesions such as squamous cell carcinoma, adenocarcinoma, adenoid cystic carcinoma, mucoepidermoid carcinoma, melanoma and olfactory esthesioneuroblastoma.

Despite the benign nature of pleomorphic adenoma, the tumor may have a malignant behavior, giving local recurrence or, more rarely, cervical metastasis or distant ones.

Compagno et al. reported 3 cases of local recurrence of 40 patients examined. Intranasal pleomorphic adenoma have a low rate of recurrence (10%) compared with recurrence rates of the major salivary gland tumors.

Figure 1 - Endoscopic view of septal pleomorphic adenoma into left nasal fossa; a (up): view of the whole lesion; b (down): relationship with inferior turbinate and closure of nasal valve region.

3. Discussion

Coffee The pathogenesis of the tumor is still unknown. Three theories have been formulated to identify the precursors of the septal pleomorphic adenoma and herein explain its particular presence in this anatomical site: ectopic embryonic epithelialized cells on the nasal septum mucosa; remnant of the vomero-nasal origin, an epithelium-lined duct in the cartilaginous nasal septum degenerated in early fetus; or directly from completely matured salivary gland tissue.

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The possibility of recurrence is less than the same tumor of the major salivary glands\textsuperscript{12}. Risk factors for tumor recurrence are the prevalence of myxoid stroma, the invasion of the capsule or its irregularity and multinodularity\textsuperscript{19}. Compagno et al. also reported a malignant transformation rate of 2.5-10% with a female predominance\textsuperscript{12}. In our case there are no clinical or radiological signs of malignancy.

The treatment of intranasal pleomorphic adenoma is surgical and it concerns a wide, local resection with a histological clear margin in order to prevent recurrence. The choice of surgical technique is founded on localization and dimension of the mass. It can be performed using several surgical approaches: intranasal resection, transnasal endoscopic, mid facial degloving, lateral rhinotomy and external rhinoplasty\textsuperscript{3}. As with other nasal tumors, the study of olfactory function is not mandatory\textsuperscript{20}. The intranasal resection of the mass, with direct (as the surgery of the nasal septum) or endoscopic approach, is the best surgical technique in the case of small lesions, thus avoiding external scars, bleeding and pain, favoring a more rapid recovery post-operative and reducing the duration of the patient’s hospitalization.

Some authors have sustained adjuvant radiotherapy after evidence of residual disease\textsuperscript{22}. Long-term follow up and careful endoscopic examination are necessary to exclude the malignancy and reduce the loco-regional recurrence rate.

4. Conclusions

Intranasal pleomorphic adenoma is rare. The possibility of a pleomorphic adenoma of the nasal septum should be taken into consideration in the diagnosis of an intranasal mass in patients with unilateral nasal obstruction or epistaxis. It is important to highlight that early detection can lead to a less aggressive treatment with a less invasive approach. Histopathological and immunohistochemical studies can confirm the diagnosis of pleomorphic adenoma. Surgical excision with a wide margin resection is the treatment of choice for preventing the appearance of recurrence. Pleomorphic adenoma is a benign tumor but, in view of the potential for recurrence or malignant transformation, it follows that a careful endoscopic and radiologic long-term follow up is necessary.

References