

Life-Threatening Bleeding in Atypical Pleomorphic Adenoma of the Soft Palate

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Abstract

Introduction:

Atypical pleomorphic adenoma (PA) is an uncommon tumor, more frequent in submandibular and parotid glands. PA is classified as atypical when it presents hypercellularity, necrosis or hyalinization, dysplasia, capsular violation or distant metastases.

Case Report:

We described a case of a 39-year-old female presented with a slowly growing mass involving the soft palate. A life-threatening bleeding from PA with hemorrhagic shock occurred and required ligation of the external carotid artery with tracheotomy. A transoral en-bloc excision of the mass (70 x 50 x 40 mm) was performed. Pathological exam demonstrated an atypical PA, with hypercellular fields and myoepithelial and squamous differentiation.

Conclusion:

An appropriate diagnostic evaluation and a prompt intervention are essential to avoid dangerous complications, even for benign neoplasms.

Keywords: Atypical; Pleomorphic adenoma; Salivary gland tumors; Soft palate; Salivary glands.

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Introduction

Pleomorphic adenoma (PA) is the most frequent tumor of the salivary glands, usually located in the parotid (1). Concerning minor salivary glands, the most common site of origin of PA is the palate (2). This tumor involves women more frequently, growing slowly and being mainly asymptomatic. PA is a benign neoplasm characterized by epithelial, myoepithelial and stromal elements with architectural pleomorphism (3). A computed tomography (CT), or more preferably a magnetic resonance imaging (MRI), allows to detect PA with greater accuracy than fine-needle aspiration cytology (FNAC), since PA is characterized by a remarkable degree of morphologic diversity even within the single tumor (4-6).

We present a case of large, atypical PA of the soft palate with pre-operative severe bleeding. The patient has given a written informed consent to publish the case. An exemption was granted by the institutional ethics committee and the study was conducted in accordance with international ethical standards, such as 1964 Declaration of Helsinki and its later amendments.

Case Report

A 39-year-old female presented at our department with a slowly growing mass that involved the soft palate and protruded in the oropharynx and nasopharynx. Except for dust mite allergy, her medical history was non-contributory.

The lesion was first observed in 2013 in another hospital, where she underwent an MRI and a biopsy, that was consistent with PA. The imaging had shown a solid, hypervascular, exophytic lesion developing from the soft palate and measuring 2 x 2 cm. She was advised to undergo transoral surgical removal of the mass. However, she refused the treatment and never attended follow-up medical examinations during the subsequent 8 years.

In 2021, after a melena-induced syncope with hemorrhagic shock, she underwent several blood transfusions and an emergency tracheotomy in local anesthesia was performed due to the impossibility of intubating the patient.

The clinical examination revealed a large, dome-shaped, non-ulcerated, swelling of the soft palate with a lot of capillaries on its surface (Figure 1). The CT revealed an inhomogeneous

poorly defined 6 x 5 cm solid mass (Figure 2), whose vascularization was mainly provided by right external carotid artery.



Fig 1: Large, non-ulcerated, dome-shaped, swelling of the soft palate with several capillaries on his surface.

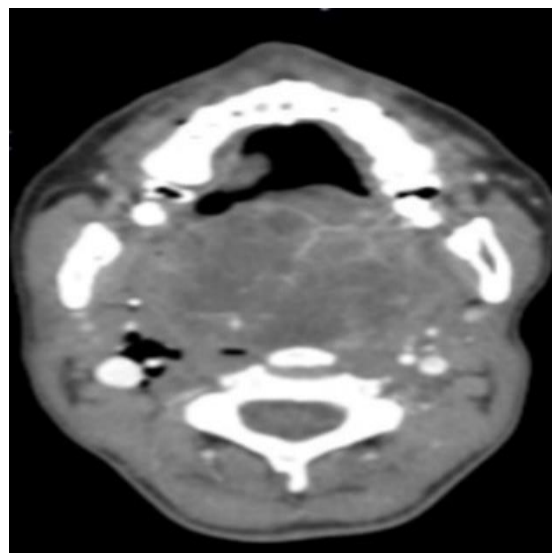


Fig 2: Axial CT scan of the atypical pleomorphic adenoma of the soft palate: an inhomogeneous poorly defined 6 cm mass involving both nasopharynx and oropharynx is visible.

Since another severe bleeding occurred four days after tracheotomy, the ligation of the right external carotid artery was performed under local anesthesia. A new biopsy of the lesion confirmed a PA. The patient underwent a transoral en-bloc excision under general anesthesia by means of electrocautery. The previous right external carotid artery ligation allowed a very low bleeding rate. The pathological exam showed a 70 x 50 x 40 mm mass (Figure 3). The remaining palate was sutured.



Fig 3: The excised atypical pleomorphic adenoma measuring 70x50x40 mm.

Post-operative course was uneventful, and the patient was decannulated after 5 days. The patient was discharged on the 9th post-operative day. A speech therapy was started after surgery and a palatal obturator was planned. Pathological exam demonstrated an atypical PA, with hypercellular fields and myoepithelial and squamous differentiation. Therefore, the patient underwent medical examination every two weeks in the first month, then once a month and performed a CT three months after surgery. No recurrence was observed at clinical examination and CT three months after surgery (Figure 4).



Figure 4. Clinical examination three months after surgery.

Discussion

Atypical PA is an uncommon tumor, more frequent in submandibular and parotid glands. PA is classified as atypical when it presents hypercellularity, necrosis or hyalinization, dysplasia, capsular violation or distant metastases (6).

Due to its rarity, little is known about the natural history and prognosis of this histopathologic variant.

Malignant transformation was observed in 13.8% of atypical PAs (7). In the absence of unambiguous guidelines, surgical excision and strict follow-up regimen are recommended.

We presented a large PA of the soft palate, with an extension of 7 x 5 x 4 cm. Such dimensions determined difficulties concerning the surgical procedure. Different surgical approaches for soft palate tumors have been proposed, including transoral, transcervical, and transmandibular (8).

The transoral approach granted us to be as unobtrusive as possible, since aesthetic result must be taken into consideration, especially when dealing with a young woman. It is not negligible that this mini-invasive approach allowed the patient to quickly recover the ability to swallow foods of any consistency and to speak in a fluent and understandable way, although with rhinolalia. Our case highlighted the risk of life-threatening bleeding also from a benign non-vascular neoplasia, which is typically a non-bleeding lesion.

Therefore, this report suggests the importance of an early management of PAs. Our patient, although affected by a benign neoplasm, faced life-threatening events that required a tracheotomy to manage airway and blood transfusions with ligation of the external carotid artery to manage bleeding. The latter allowed a safe transoral surgical approach with a minimal intraoperative bleeding.

Conclusion

In conclusion, an appropriate diagnostic evaluation and a prompt intervention are essential to avoid dangerous emergency situations, even for benign neoplasms. Further studies are necessary to establish the best post-operative follow up scheme for atypical PAs.

Declarations of conflicting interest

The Authors do not have any conflict of interest to declare.

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