

# A Rare Case Report of Pleomorphic Adenoma of the Submandibular Gland Extending to Involve the Parapharyngeal Space

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## ABSTRACT

*Pleomorphic adenomas are benign tumors of the salivary glands, most commonly involving the parotid gland. Only 5% of pleomorphic adenomas involve the submandibular gland, of which tumors extending into the parapharyngeal space (PPS) are very rare, and locate in complex anatomical regions. The purpose of this article is to report a rare case of pleomorphic adenoma of a submandibular gland extending to involve the PPS. The incidence, differential diagnosis and surgical treatment of a pleomorphic adenoma involving the PPS are discussed.*

**Key words:** Pleomorphic adenoma, Submandibular gland, Parapharyngeal space (PPS).

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## ÖZET

### Nadir Görülen Parafaringeal Mesafeye Uzanım Gösteren Submandibüler Bezin Pleomorfik Adenomunun Olduğu Bir Olgu Sunumu

*Pleomorfik adenomlar tükürük bezlerinin selim tümörlerindedir ve en sık parotis bezinde gözlenir. Pleomorfik adenomların %5'i submandibüler bezi tutar ve çok nadir durumlarda tümör parafaringeal mesafeye (PPS) uzanım göstererek karmaşık anatomik bölgelerde yerleşebilir. Bu çalışmanın amacı nadir görülen ve PPS'ye uzanım göstermiş submandibüler bez pleomorfik adenoma olgusunu sunmaktır. PPS'ye uzanım gösteren pleomorfik adenoma olgusunun görülme sıklığı, ayırıcı tanısı ve tedavi yöntemleri de tartışılacaktır.*

**Anahtar kelimeler:** Pleomorfik adenom, Submandibüler bez, Parafaringeal mesafe (PPS)

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## INTRODUCTION

Salivary gland tumors represents about 1-4% of all neoplasias, affecting the parotid gland in more than 70% of cases, submandibular gland in 5-10%, sublingual glands in 1%, and minor salivary glands in 5-15%. Pleomorphic adenoma is the most frequent benign tumor of the salivary glands. Approximately 5% of pleomorphic adenomas are located in the submandibular glands<sup>[1]</sup>.

Parapharyngeal space (PPS) tumors are rare, representing only about 0.5% of head and neck neoplasms. The distribution of PPS tumors is as follows: pleomorphic adenoma (40%), paraganglioma (20%), neurogenic tumors (14%), malignant salivary tumors (13%), miscellaneous malignant tumors (7%), and miscellaneous benign tumors (6%). This wide variety of tumors, coupled with the difficulty of surgical access to the PPS, poses a challenge in selecting the optimum approach to maximize visibility, ensure complete removal of the tumor, and minimize morbidity<sup>[2]</sup>.

Here we report a case of pleomorphic adenoma of the submandibular gland extending into the PPS.

## CASE REPORT

We present herein the case of a 42-year-old patient with the chief complaint of a painless swelling in the left submandibular region for the past six years, with difficulty in swallowing.

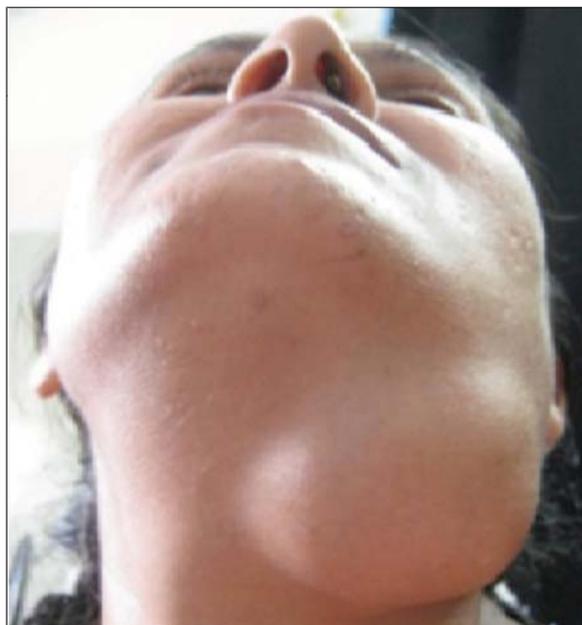
Extraorally, the swelling was seen 1 cm beneath the inferior border of the mandible. It was solitary, mobile, oval in shape, approximately 12 x 8 cm, and rubbery in consistency; the overlying skin was intact and not crossing the midline (Figure 1).

Intraorally, the swelling was well encapsulated and seen in the ipsilateral side of the oropharyngeal wall, approximately 5 x 3 x 4 cm, and firm in consistency; the left portion of the tongue was seen to be pushed down. The tonsillar bed was medially displaced and was immediately recognizable in the oral cavity. The patient had difficulty in swallowing and speaking (Figure 2).

Contrast magnetic resonance imaging (MRI) revealed the details of the tumor of the submandibular gland and its extension into the PPS (Figure 3).

True cut biopsy was done, and the histological examination confirmed the diagnosis of pleomorphic adenoma (Figure 4).

Surgical resection of the tumor was planned under general anesthesia. Transcervical incision 2.5 cm beneath the inferior border of the mandible over the swelling was done, and the incision was continued



**Figure 1. Solitary tumor seen in the submandibular region.**



**Figure 2. Intraoral view showing extension to involve the parapharyngeal space.**

sub-platysmally, with the marginal branch of the mandibular nerve preserved. Through blunt dissection, the tumor was exposed completely, and the facial vessels were identified and ligated (Figure 5).

Lateral swing mandibulotomy was performed through a lip-splitting incision to expose the tumor extending into the PPS. With careful blunt dissection and preservation of vital structures, the tumor was removed in toto (Figure 6).

There had been no recurrence at the one-year follow-up.



Figure 3. MRI contrast showing tumor extension and involvement.

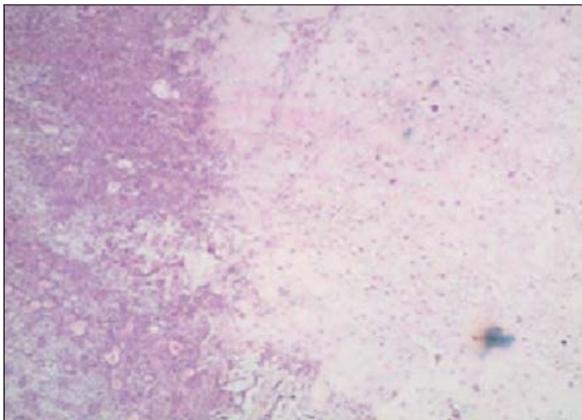


Figure 4. Histopathology.

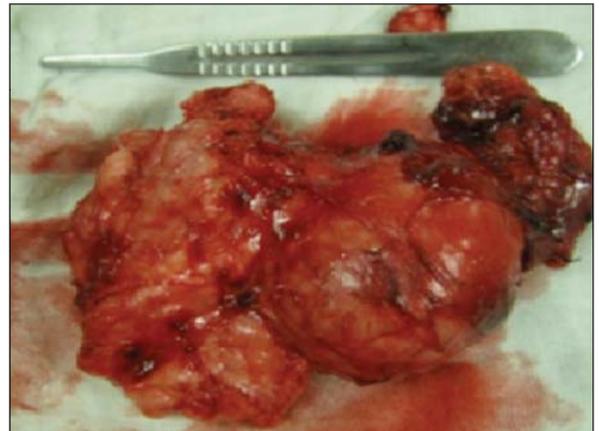


Figure 6. Surgical specimen.



Figure 5. Exposure of the tumor.

#### DISCUSSION

The name pleomorphic adenoma was suggested by Willis, and pleomorphic adenoma represents 45-74% of all salivary gland tumors<sup>[3]</sup>.

The prevalence is more common in females, affecting adults more frequently than the young. In 1989, Schultz-Coulon<sup>3</sup> reviewed 31 cases of giant pleomorphic adenoma and found female predominance of around 64.5%, with an age range of 20-40 years.

Pleomorphic adenomas in the PPS can arise either de novo or in the deep lobe of the parotid gland and extend through the stylomandibular tunnel into the PPS. The origin of de novo pleomorphic adenoma is probably from displaced or aberrant salivary gland

tissue within a lymph node or due to direct seeding of the submandibular gland during surgery<sup>[4,5]</sup>.

Parapharyngeal space (PPS) tumors are infrequent, mostly benign and present few symptoms, making an early diagnosis very difficult. They represent up to 0.5% of head and neck neoplasms and are benign in nature in 80% of cases. Of the large group of possible benign PPS tumors, pleomorphic adenoma, originating from either the parotid gland or some aberrant minor salivary gland, is the most represented entity, accounting for approximately 50% of all benign PPS lesions<sup>[4]</sup>.

Computed tomography (CT) and MRI are both of the utmost importance in the preoperative evaluation of a PPS mass, with MRI being the more appropriate due to its superior soft-tissue resolution and the possibility of offering multiple plane imaging of this region. In a study on 51 patients with a PPS neoplasm, both benign and malignant, CT and MRI were able to localize the mass in 95% and 84% of patients, respectively<sup>[5]</sup>.

The PPS, otherwise referred to as peripharyngeal, lateral pharyngeal, pterygopharyngeal, pterygomaxillary, and pharyngomasticator space, is a closed, inverted, truncated pyramid-shaped space located on the both sides of the neck, lateral to the fascia overlying the superior pharyngeal constrictor and tensor and the levator palatini muscles and into the pterygoid lamina and ramus of the mandible. It is located superiorly alongside the middle fossa base of the skull and beside the fascia over the spinal column and paraspinal muscles. Inferiorly, it has been reported to reach the level of the hyoid bone. However, this is one of the most controversial boundaries, and some authors argue against it, pointing out that caudally, near the angle of the mandible, the PPS is functionally obliterated by the fusion of the fascia around the submandibular gland, sheaths of the styloid muscles, fascia over the posterior belly of the digastric muscle, and all the visceral fascia and, therefore, the styloglossus muscle, which runs along the caudal demarcation line. Classically, it is divided into an anterolateral or pre-styloid compartment containing some fat and the deep elongation of the parotid gland and a posteromedial or post-styloid compartment in which the internal carotid artery, internal jugular vein, IX<sup>th</sup> to XII<sup>th</sup> cranial nerves, and the sympathetic chain are found. The separation between these two sub-spaces is represented by the tensor-vascular-styloid fascia, which is a fairly thick envelope filling the gap between the tensor veli palatini muscle and the medial pterygoid plate, skull base, styloid process, and the relative musculature<sup>[5]</sup>.

The intricate vascular, neural, and lymphatic structures of the PPS call for the differential diagnosis to include not only tumors originating in the PPS, but also extension of tumors originating in adjacent structures and metastases at the PPS level (Table 1)<sup>[6]</sup>.

There is a possibility of multiple options concerning the surgical approach to the PPS, which depends mainly on the size of the tumor, its location with regard to surrounding vital structures and the surgeon's experience. The treatment of choice to approach the PPS should meet two basic criteria: wide surgical exposure to guarantee a safe and radical dissection and minimal postoperative problems, both functional and cosmetic<sup>[7]</sup>.

Traditional PPS surgery mainly uses the transcervical and transparotid approaches. Malone et al. and Hamza et al. described the resection of PPS tumors using the transcervical approach alone in 90-100% of cases. Hughes et al.<sup>[10]</sup> published a series of 172 cases using the transcervical and transparotid approaches in 94% and mandibular osteotomy in only 2% of resections<sup>[8]</sup>.

A choice can be made between mandibular-splitting and mandibular-preserving procedures. The use of mandibulotomy to gain access to the PPS was first described by Butlin in 1885. The main advantage of a mandibular-splitting approach is that it allows greater exposure of the PPS; nonetheless, it usually causes more severe scarring and involves risk to the inferior alveolar nerve<sup>[7]</sup>.

**Table 1. Differential diagnoses of parapharyngeal tumors**

**Benign tumors**

- Pleomorphic adenoma
- Monomorphic adenoma
- Warthin's tumor
- Branchial cyst
- Sjögren's paraganglioma
- Schwannoma
- Neurofibroma

**Malignant tumors**

- Lymphoma
- Adenoid cystic carcinoma
- Adenocarcinoma
- Squamous cell carcinoma
- Neurofibrosarcoma
- Sarcoma

The primary concerns with the mandibulotomy access approaches are:

- difficulty with mandibular healing,
- preservation of the occlusion, and
- loss of mental nerve sensation.

To minimize the risk of sensory disturbances, osteotomies distal to the mental foramen have been described. More recently, osteotomies at the mandibular angle as well as step or oblique osteotomies of the body proximal to the mental nerve have been described<sup>[7]</sup>.

Pleomorphic adenomas have a high recurrence rate. Many investigators have emphasized adequate surgical management of the initial tumor to avoid recurrence. Recurrence depends on the type of the surgery and lesions of the pseudocapsule. Due to intraoperative maneuvers and insufficient preoperative diagnostics, there are chances of underestimation of tumor extensions, particularly in cases with multifocal origin. The incidence of recurrence is around 0.5-10%<sup>[9]</sup>.

The incidence of malignant transformation in pleomorphic adenomas ranges 1.9% to 23.3%. Some authors have postulated that the risk of malignant transformation increases from 1.6% in tumors with less than five years of evolution to 9.5% for those presenting for more than 15 years<sup>[10]</sup>.

In conclusion, pleomorphic adenomas of the submandibular gland extending to involve the PPS are very rare. As such parapharyngeal tumors are very rare and locate in complex anatomical regions, careful clinical and radiological examination must be done for better prognosis, and surgical resection is the mainstay of treatment.

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