## **CASE REPORT**

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# Squamous cell carcinoma of the pterygopalatine fossa -A case report

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#### ABSTRACT

The pterygopalatine fossa is an inverted pyramid-shaped space of the viscerocranium, protected by bony structures. Surgical access to this anatomical space is difficult, especially for tumor resection. There are numerous open surgical techniques for accessing this space, but nowadays, minimally-invasive endoscopic approaches are preferred in order to increase postoperative quality of life and reduce postoperative morbidities.

The tumors of the pterygopalatine fossa can be benign or malignant, and can occur primarily in the fossa or as secondary extensions from the surrounding regions through the multiple canals and foramina in its walls. Squamous cell carcinomas of this space have been described to appear as extensions from the nasopharynx, the paranasal sinuses or through perineural extension from the cutaneous branches of the maxillary nerve.

In this paper the authors present a rare case of squamous cell carcinoma of the pterygopalatine fossa, which was excised in an endoscopic transnasal approach after preoperative selective embolization.

KEYWORDS: pterygopalatine fossa, squamous cell carcinoma, endoscopic surgery

#### **INTRODUCTION**

The pterygopalatine fossa (PPF) is a challenging space for surgery due to its important vital relations, like the skull base and the orbit, but also due to the vascular and nervous elements that pass through it. Pathological processes in this anatomical space are uncommon. Both malignant and benign tumors can occur in this space. Most commonly, extensions from nasopharyngeal juvenile angiofibromas or nerve sheath tumors (schwannomas) are found in the PPF<sup>1</sup>. Other tumors involving this space have been described, like pleomorphic adenomas, lymphomas, renal carcinoma metastasis or craniopharyngiomas<sup>24</sup>.

Squamous cell carcinomas of the PPF are rare, and usually occur secondarily by direct extension from adjacent structures like the nasopharynx and the maxillary or the sphenoid sinuses<sup>5</sup>. Other types of dissemination of malignancies in the PPF were described, like perineural spread along the maxillary nerve, or the more unlikely hematogenous and lymphatic routes<sup>5</sup>. Primary squamous cell carcinoma in the PPF is considered to be highly improbable due to the lack of squamous epithelial tissue in the fossa<sup>5</sup>.

Surgery of the PPF is difficult because the space is well bounded and hard to explore. Traditional surgery for this fossa includes different open-approach techniques, like midfacial degloving, lateral rhinotomy with medial maxillectomy, which have significant postoperative morbidities and sequelae<sup>1</sup>. Modern techniques are described and used due to technological advances in endoscopic surgery, and it is considered that endoscopic-assisted surgery of the PPF can provide better visualization and potentially reduce surgical risks<sup>1,6</sup>.

This paper presents the case of a 33-year-old woman who underwent endoscopic surgery for a right PPF tumor. Histopathology revealed the diagnosis of squamous cell carcinoma.

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A 33-year-old female patient presented to the clinic complaining of gradual sensation of numbness and pain on the right infraorbital region, along with headache and facial pressure. The symptoms appeared a couple of months before presentation and increased in intensity.

Initial clinical examination and endoscopy of the nose and nasopharynx showed no specific lesions of the regions examined. No prior significant conditions were known or related by the patient, no malignancies or long-term treatment.

The patient underwent a contrast-enhanced computed-tomography (CT) of the nose, paranasal sinuses and skull base. This revealed an opaque mass in the right PPF, determining an anterior bulge and thinning of the posterior wall of the right maxillary sinus, associated with mucosal thickening of the right sphenoid sinus, posterior ethmoid and nasopharynx (Figure 1). No extensions of the mass were revealed into the infratemporal fossa or the skull base (Figure 2).

Also, the patient underwent a selective angiography of both common carotid arteries and their terminal branches, which revealed a contact between the tumor and the right maxillary artery, the tumor receiving blood supply directly from it. During the same procedure, a selective embolization of the arterial branches and of the maxillary artery was performed.

Approximately 24 hours after embolization, surgery for tumor removal was performed. The surgical technique consisted in an endoscopic transnasal-transmaxillary approach of the PPF. After the endoscopic medial maxillectomy was performed, the bulging of the posterior maxillary wall was visible. This wall was paper-like thin, and it was carefully removed, opening the PPF (Figure 3). The tumor was excised in toto, with a rather difficult dissection from the PPF contents and fatty and connective tissue. Some vascular sources were encountered, but bleeding was only minor and controlled through bipolar cauterization (Figure 4). In the same procedure, paraseptal and transethmoidal sphenoidotomy were performed, as well as posterior ethmoidectomy. Several fragments from the sphenoid sinus, posterior ethmoid and also from the nasopharynx were sent for extemporaneous histologic examination. Minimal nasal packing was necessary after tumor removal, and the patient had a favourable postoperative outcome.

After surgery, the tumor was sent to pathology for examination. The histological and immunohistochemical diagnosis were of squamous cell carcinoma for the tumor, and chronic inflammation from the other fragments that were sent.

Figure 1 Preoperative cranio-facial computed-tomography, axial section.

The mass in the right PPF can be observed, as well as the anterior bulge and thinning of the posterior maxillary wall; note also the opacity in the right sphenoid sinus and posterior ethmoid.



**Figure 2** Preoperative cranio-facial computed-tomography, coronal section. The tumor in the right PPF can be observed, and also the anterior displacement of the posterior wall of the maxillary sinus.



**Figure 3** Intraoperative aspect. Note the removal of the paper-thin posterior wall of the maxillary sinus (black arrows), revealing the tumor in the PPF.

The patient was advised to seek further oncological treatment and care in the oncology unit, but she refused medical treatment and started her own alternative treatment. Approximately six months after surgery the tumor reoccurred in the PPF and in the parapharyngeal space.

#### DISCUSSIONS

The pterygopalatine fossa is a narrow, inverted pyramid-shape space situated directly posterior to the maxillary sinus<sup>1,5</sup>. The boundaries of the PPF are complex and are represented by the posterior wall of the maxillary sinus anteriorly, the pterygoid process of the sphenoid bone posteriorly, the pterygomaxillary fissure laterally and the ascending process of the palatine bone medially. The superior wall is the greater wing of the sphenoid bone. In the inferior part, the fossa becomes narrow and continues with the palatine canal<sup>1,5,7,8</sup>. Aside from the complex bony structure of the PPF, its contents make it even more difficult to operate. Its contents are represented by important nervous and vascular structures, which pass from the PPF to complex networks of foramina and canals into the surrounding regions<sup>1</sup>. The vascular structures are branches of the maxillary artery and nervous structures are part afferent and efferent fibers of the pterygopalatine ganglion<sup>9-11</sup>.

Lesions of the PPF itself are rare, but some types of inflammation, infection and tumors can occur in the space primarily or by extension from related spaces through its canals and foramina<sup>5,11,12</sup>.

Tumors in the PPF were presented, both benign and malignant, and a variety of histopathological types are described, such as teratomas, neurofibromas, neurofibrosarcomas, choristomas, as well as adenocarcinomas, mucoepidermoid carcinomas or adenoid cystic



**Figure 4** Intraoperative aspect. Three-hand surgery for better visualization and dissection of the tumor.

carcinomas, the latter three usually occurring from the deep lobe of the parotid gland<sup>13,14</sup>.

Squamous cell carcinoma of the PPF is very unlikely to appear as a primary tumor, since this space does not typically contain epithelial tissue. However, this type of carcinoma can occur via direct spread or perineural spread from the maxillary or sphenoid sinuses, from the nasopharynx or from the cutaneous territory of the maxillary nerve<sup>5</sup>. In our case, the histological and immunohistochemical diagnosis was of squamous cell carcinoma, and there was no other primary tumor discovered using intraoperative biopsies that were sent for analysis.

Regarding the preoperative diagnosis of PPF tumors, it is difficult to reach due to the location of this anatomical space, which is well bounded by bony structures, and risky to biopsy. Even though most lesions of the head and neck are accessible for biopsy through simple procedures, this is extremely difficult if not impossible in the PPF<sup>13</sup>. A preoperative diagnosis obtained through biopsy or fine-needle aspiration would be very helpful for treatment planning<sup>13,15-18</sup>. Imaging techniques, such as CT and MRI, are helpful in determining the primary tumor and its extensions. Greater attention should be paid in determining the tumor extension laterally into the infratemporal fossa, superiorly into the skull base, or the posterior extension which can involve the cranial nerves<sup>1,19</sup>.

Surgical access in case of PPF pathology has greatly evolved with the development of endoscopic techniques and adoption of minimally invasive operative approaches. These approaches, like the transnasal or transmaxillary, give excellent visualization and greater ergonomics to the surgeon, also being safe and with far less morbidities than the open techniques<sup>1</sup>. In our case, we used a transnasal, transmaxillary approach, with an endoscopic medial maxillectomy and opening of the PPF through the posterior wall of the maxillary sinus. This approach offered good visualization of the tumor and its surroundings, whilst being minimally invasive for the patient. The tumor was excised macroscopically in toto. Bleeding was minimal probably due to preoperative selective embolization of the direct arterial branches of the tumor and branches of the maxillary artery. Angiography and selective embolization are usually necessary in the preoperative stage for tumors considered highly vascular, like juvenile nasopharyngeal angiofibromas, but are also useful in other tumors, in order to reduce operative blood loss and improve visualization. The procedure should occur 24-48h prior to surgery, in order to minimize tumor revascularization<sup>1,20</sup>.

Even though tumor resection was macroscopically complete, further oncological treatment and survey was necessary and recommended. However, the patient refused further treatment and started alternative treatments. This led to an unfavourable outcome, with tumor recurrence approximately six months after surgery.

In this case, as in most oncological cases, we consider that complex and combined treatment schemes, with surgery, radiotherapy and chemotherapy would be indicated and have more favourable results than surgery alone.

#### CONCLUSIONS

The PPF is a difficult anatomical space to access surgically, but modern techniques have led to an improvement in surgical approaches and quality of life for the patient. Using endoscopic techniques, there is reduced postoperative pain and oedema, and fewer neurovascular complications, improving the postoperative course of the patient.

Even though pathological conditions of the PPF are rare, several inflammations, infections and tumor can occur in this space, and the diagnosis as well as the treatment are challenging for the attending physician.

Primary squamous cell carcinoma of the PPF is considered highly unlikely; in our case, another primary tumor was not revealed even after extensive diagnostic procedures, before, during and after surgery. In this case, we consider that the tumor might have been an extension from a primary nest in the nasopharynx or the sphenoid sinus, even though the biopsies were negative.

Although surgical techniques have made extensive progresses in this area of surgery, and, nowadays, complete resection of some tumors is possible, combined schemas of oncologic treatment should be used, leading otherwise to unfavourable results. Conflicts of interests: None

**Contribution of authors:** All authors have equally contributed to this work.

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