

## CASE REPORT

# Simultaneous pleomorphic adenoma of the parapharyngeal space and contralateral submandibular gland. Case report

*Adenoma pleomorfo ad insorgenza simultanea nello spazio parafaringeo e nella ghiandola sottomandibolare controlaterale. Descrizione di un caso clinico*

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## SUMMARY

Herein the case is reported of a synchronous parapharyngeal space pleomorphic adenoma arising from the pharyngeal prolongation of the parotid gland and the contralateral submandibular gland, diagnosed in a young Caucasian female. Case reports and recent literature are presented. Upon physical examination, asymmetry of the lower part of the right side of the face with overlying intact skin, and a submucosal firm swelling, filling the ipsilateral side of the oropharyngeal wall, involving the right tonsillar bed which was medially displaced was immediately recognizable. The patient did not complain of dysphagia. Palpating the left submandibular region, a painless, mobile, rounded mass, 10 mm in diameter, apparently located in the submandibular gland, was detected. Magnetic resonance imaging showed that both lesions were well-defined and encapsulated. The surgical approach is discussed. Cytological diagnosis was that of a typical pleomorphic adenoma. To the best of our knowledge this is the second case report in the English literature of a concomitant pleomorphic adenoma located both in the parapharyngeal space and the submandibular gland.

KEY WORDS: Salivary glands • Parapharyngeal space • Pleomorphic adenoma

## RIASSUNTO

*Viene descritto il caso di un adenoma pleomorfo insorto in una giovane donna di razza caucasica in modo sincro nel prolungamento faringeo della ghiandola parotide e nella ghiandola sottomandibolare controlaterale. Vengono illustrati nel nostro lavoro casi simili. All'esame obiettivo della paziente era evidente un'asimmetria della parte inferiore del volto con cute sovrastante integra ed una tumefazione sottomucosa omolaterale della parete orofaringea con tonsilla che appariva mediatizzata. La paziente non riferiva disfagia. Alla palpazione era evidente una tumefazione di circa 10 mm della loggia sottomandibolare controlaterale, mobile, ben capsulata, non dolente. Una risonanza magnetica nucleare della regione dimostrava entrambe le lesioni ben evidenti e capsulate. Nell'articolo viene discusso l'approccio chirurgico utilizzato. L'esame istologico di entrambe le lesioni è stato di adenoma pleomorfo. A nostra conoscenza è il secondo caso descritto nella letteratura inglese di adenoma pleomorfo ad insorgenza sincro nello spazio parafaringeo e nella ghiandola sottomandibolare.*

PAROLE CHIAVE: Ghiandole salivari • Spazio parafaringeo • Adenoma pleomorfo

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## Introduction

The parapharyngeal space (PPS) is a well-defined anatomic region located as an inverted pyramid medially to the mandible and laterally to the superior pharyngeal constrictor muscle. It is filled with loose connective tissue in which a multitude of vital structures are found.

PPS primary tumours are rare. They represent up to 0.5% of head and neck neoplasms<sup>1</sup> and are benign in nature in approximately 80% of cases<sup>2</sup>. Of the heterogeneous group of neoplasms which can originate within this space, pleomorphic adenoma (PA) is the most common, accounting for about 50% of all cases diagnosed<sup>3</sup>. Approximately 5% of pleomorphic adenomas are located in the submandibular glands.

The case is reported of a synchronous PPS pleomorphic adenoma, arising from the pharyngeal prolongation of the parotid gland and the contralateral submandibular gland, diagnosed in a young Caucasian female.

## Case report

An otherwise healthy 28-year-old white female was referred to our Outpatient Clinic with a 9-month history of a painless mass in the right side of her neck and some swelling which she said had been present for 4 weeks.

Upon physical examination, asymmetry of the lower part of the right side of the face, with overlying intact skin, and a sub-mucosal firm swelling filling the ipsilateral side of the oropharyngeal wall, involving the right tonsillar bed which



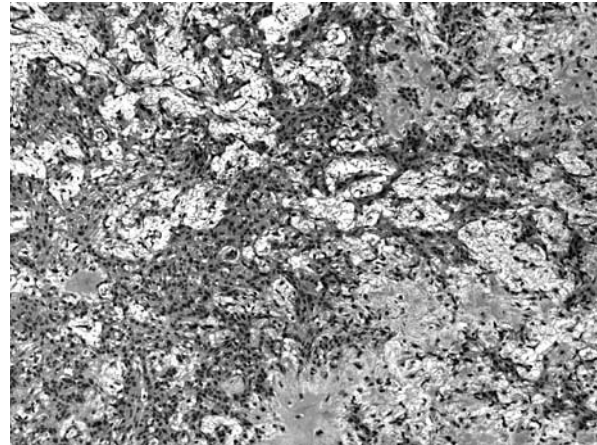
**Fig. 1.** MRI imaging showing expansive formations.

was medially displaced, was immediately recognizable. The patient did not complain of dysphagia. Upon palpating the left submandibular region a painless, mobile, rounded mass, 10 mm in diameter, apparently located in the submandibular gland, was detected. No other abnormalities were found in the head and neck region. Clinical history did not include previous exposure to radiotherapy.

At Magnetic Resonance Imaging (MRI) both lesions were found to be well-defined and encapsulated. In particular, the right PPS was fully occupied by a round shaped expansive formation measuring approximately 5 x 3 cm, which was hyperintense on T2-weighted images, had a low intensity signal on T1-weighted images and imprinted the lateral wall of the oropharynx, laterally displacing the ipsilateral internal pterygoid muscle; at the same time, a 1 x 1 cm rounded focal lesion was clearly present in the context of the contralateral submandibular gland. Neither of these lesions showed signs of infiltration of their capsule.



**Fig. 2.** Surgical view of parotidectomy.



**Fig. 3.** Histology: mixture of epithelial, myoepithelial and stromal components. Epithelial cells: nests, sheets, ducts. Stroma: myxoid, chondroid, fibroid

Trans-oral and trans-cervical fine-needle aspiration was then performed and samples of the lesions obtained revealed that they were of the same benign nature as pleomorphic adenoma.

The patient had not undergone any surgery or radiation therapy for this condition.

The need to proceed with surgical removal of both the parotid and the submandibular masses was fully discussed with the patient and informed consent was signed.

### Surgical procedure

The submandibular mass was excised through a 4 cm linear cervical incision along a natural crease of the skin in the left submandibular region.

As far as concerns the PPS lesion, this was surgically approached via a classic trans-parotid nerve-sparing technique through a modified Blair incision, the final result being total parotidectomy with no injury to any of the branches of the facial nerve. In dissecting and dividing the parotid tissue from the facial plane, mosquito artery forceps were used whereas to control bleeding both bipolar diathermy and vascular clips were used, depending upon the size of the vessel. Since the bony vertical ramus of the mandible did not represent an obstacle during dissection, mandibulotomy was not necessary. The procedure was completed uneventfully.

Gross appearance of the submandibular mass was that of a well demarcated lesion measuring 1 x 1 cm; the PPS lesion, measuring 5 x 3 cm, was encapsulated and presented as a single mass with a yellowish, irregular, bosselated surface. Subsequent microscopic examination confirmed the cytological diagnosis as its appearance was that of a typical pleomorphic adenoma with a very admixture of ductal epithelium, myoepithelium and stroma.

### Discussion

Concomitant pleomorphic adenoma of the major salivary glands is a rare but well documented occurrence.

The neoplasm most commonly involved in bilateral or multicentric simultaneous salivary gland location is the Warthin tumour, while pleomorphic adenoma (benign mixed tumour) is the second most frequent.

To the best of our knowledge this is the second case report in the English literature of a concomitant pleomorphic adenoma located both in the PPS and the submandibular gland.

The first concerned a simultaneous ipsilateral pleomorphic adenoma of the PPS and the submandibular gland in a Nigerian male who had already undergone a double distinct neck operation for the same problem, thus it could also be hypothesized that the PPS mass arose due to seeding of the submandibular tumour<sup>4</sup>.

PPS, otherwise referred to as peripharyngeal, lateral pharyngeal, pterygopharyngeal, pterygomaxillary, pharyngomaxillary, pharyngomasticator space, is a closed inverted truncated pyramid-shaped space located on 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 the 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, 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, the sheaths of the styloid muscles, the fascia over the posterior belly of the digastric muscle and all the visceral fascia and, therefore, the styloglossus muscle, which runs along the caudal aspect of this region, would be a better caudal demarcation line. Classically, it is divided into an antero-lateral or pre-styloid compartment containing some fat and the deep elongation of the parotid gland, and a postero-medial or post-styloid compartment in which the internal carotid artery, internal jugular vein, IX to XII cranial nerves and the sympathetic chain are found. The separation between these two subspaces 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>5-8</sup>. It is a direct consequence of such a variegated anatomical content that, in the presence of a PPS mass, the ENT surgeon is faced with a broad differential diagnosis of possible masses, including salivary gland neoplasms, schwannoma, paraganglioma, lymphadenopathy, lipoma.

There is a possibility of a multiple choice concerning the surgical approach to the PPS, which depends mainly on the size of the tumour, 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 post-operative problems, both func-

tional and cosmetic<sup>9</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<sup>10</sup>. The main advantage of a mandibular-splitting approach is that it allows greater exposure of the PPS; nonetheless, it usually causes more severe scarring, involves risk to the inferior alveolar nerve, and, because of the swelling of the soft tissues, may, sometimes, require tracheotomy.

The PPS tumour is very rare, accounting for < 0.5% of all head and neck neoplasms<sup>11</sup>. Benign diagnoses account for 71.4-82% of the various series of PPS neoplasms<sup>12</sup>. Of the large group of possible benign PPS tumours, the pleomorphic adenoma, originating either from the parotid gland or from some aberrant minor salivary gland, in the context of the PPS, is the most represented entity, accounting for approximately 40% of all benign PPS lesions<sup>11</sup> and 63% of all parotid gland tumours<sup>14</sup>.

Computed tomography (CT) and MRI are both of the utmost importance in the pre-operative evaluation of a PPS mass, with MRI being the more appropriate on account of its superior soft-tissue resolution and the possibility of offering multiple plane imaging of this region (MRI provides the most useful information regarding precise tumour margins and their relationship with the surrounding structures). 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>14</sup>.

At the moment, fine-needle aspiration biopsy (FNAB) is the easiest and safest procedure to obtain biopsy samples of PPS lesions. No reports of tumour seeding, after head and neck aspiration, have been described recently<sup>15</sup>. This is why some authors argue that FNAB may be superior to combined physical examination and radiological evaluation<sup>16</sup>. Pre-operative definitive diagnosis, with tumour typing, is less important and incisional biopsy of any PPS mass should definitely be avoided, in order not to run the risk of a significantly higher rate of recurrence (due to rupture of the capsule and consequent tumour spillage) and, more important, to avoid the risk of fatal complications such as carotid body tumour rupture.

Since the medical history showed that no previous radiotherapy or salivary gland surgery had been performed in this patient, the concomitant benign mixed tumour, in this case, could, in our opinion, be the result of an independent development of two distinct PAs or an example of a rare metastasizing mixed benign tumour, the latter being, by far, the least likely hypothesis since almost all of these tumours have a predilection for metastasizing to lung and bone<sup>17</sup> and are preceded by recurrence and further surgical treatment<sup>18</sup>.

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