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Case Report

A RARE CASE OF A HUGE PLEOMORPHIC ADENOMA OF MINOR SALIVARY GLANDS IN THE PARAPHARYNGEAL SPACE

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ABSTRACT

Parapharyngeal space lesions account for only 0.5% of head and neck tumours. A 63-year-old male presented primary pleomorphic adenomas arising de novo from minor salivary glands in the parapharyngeal space. We describe the clinical features, pathology, radiological findings and treatment of this lesion. The goal when presented with such a tumour is a correct diagnosis and the in toto excision of the tumor to avoid any recurrence.

1. Introduction

Parapharyngeal space tumours are very rare, accounting for some 0.5% of neoplasms of the head and neck. The majority (70%-80%) are benign and 40-50% of these originate in the salivary glands (1). The most common tumour in the parapharyngeal space (PPS) is pleomorphic adenoma which can arise *de novo* or from the deep parotid lobe and extend into the PPS. The origin of *de novo* pleomorphic adenoma is probably from displaced or aberrant salivary gland tissue within a lymph node (2). However, pleomorphic adenoma arising *de novo* in the parapharyngeal space is extremely rare. The case of a 63-year-old male, his management and treatment is presented.

2. Case Presentation

A 63-year-old male presented dysphagia and a painless swelling of the left submandibular region. On intraoral examination there was a smooth firm bulge of the soft palate and left lateral pharyngeal wall (Figure 1). The submandibular swelling was palpable and ballotable (Figure 2).

Posterior nasal examination showed the extension of the swelling into the nasopharynx.

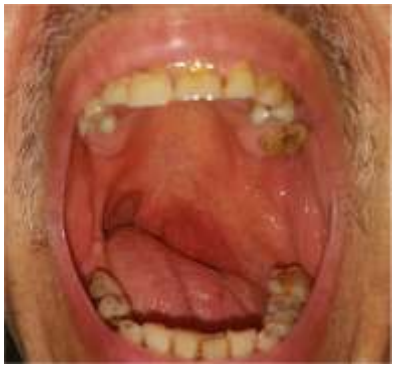

Figure 1 - Intraoral view showing the displacement of soft palate on left side.

There was no significant lymph node enlargement in the neck. Clinical examination did not reveal involvement of any of the cranial nerves. CT scan showed heterogeneously enhancing tumour with areas of necrosis measuring 6 x 4 x 8 cm in the left parapharyngeal space, extending from the skull base to the floor of the mouth.

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Caliber reduction of the left jugular vein at the level of the petrous bone was evident (Figure 3). Fine needle aspiration cytology (FNAC) revealed a diagnosis of pleomorphic adenoma.

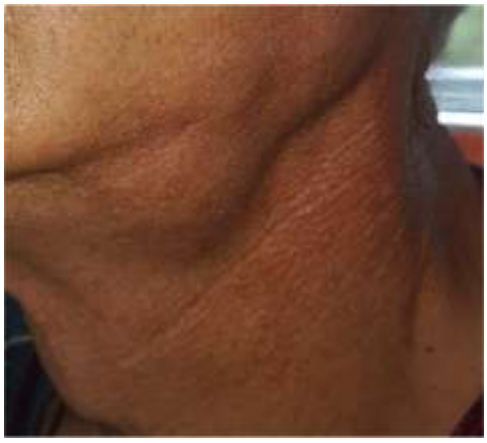

Figure 2 - Neck view showing a swelling on left submandibular region.

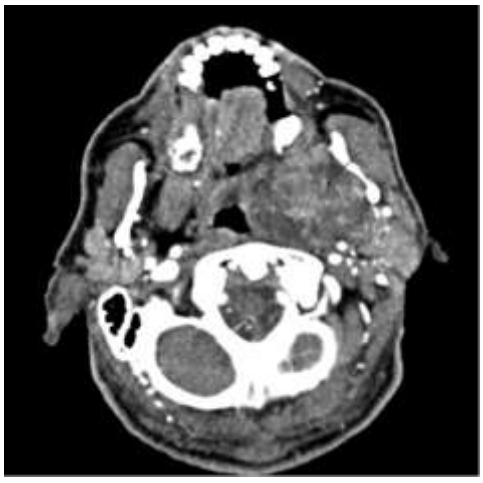

Figure 3 - CT Scan showing a well-defined mass in the left parapharyngeal space suggesting a tumour of benign origin and caliber reduction of the left jugular vein

Therefore, balloon catheter was positioned in the left jugular by right transfemoral venous access two hours before the intervention. Trans-cervical approach was used to access to the left parapharyngeal space (Figure 4), the tumour was separate from the deep parotid lobe and was completely excised. On gross examination the lesion was 8 × 6 cm with a whitish, lobulated and glistening surface (Figure 5). Histopathological examination showed a neoplasm having an admixture of epithelial and stromal components. Ducts lined by inner epithelial and outer myoepithelial cells were seen surrounded by a chondro-myxoid stroma typical of pleomorphic adenoma. Postoperative period was uneventful and patient was discharged after ten days. Repeat CT scan done after 6 months and at 1 year of follow up did not show any evidence of residual or recurrent disease. Patient is free of disease to date.

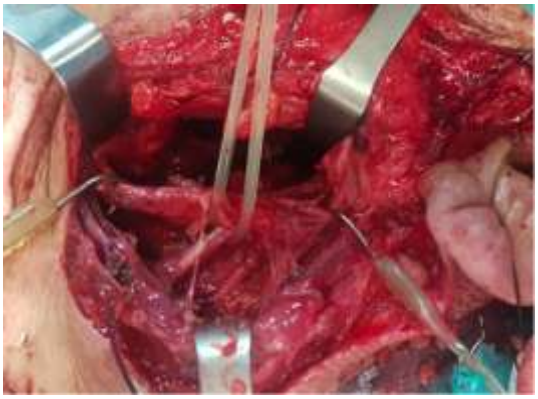

Figure 4 - Surgically exposed lesion through transcervical approach. Exhibition by string of the internal jugular vein

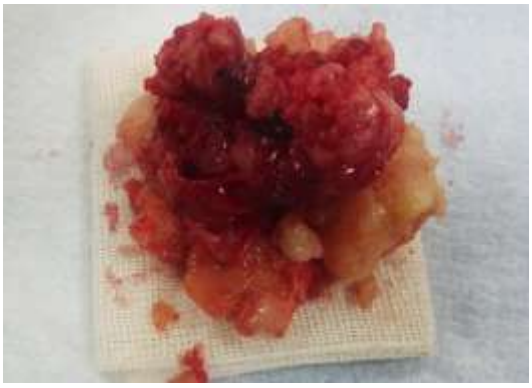

Figure 5 - Excised tumour

3. Discussion

Tumours arising in the minor salivary glands account for 22% of all salivary gland neoplasms (3). 18% of them are benign, the remaining are malignant. Of all the benign tumours, pleomorphic adenoma is the most common (3). The typical site of this adenoma is on the minor salivary glands of the palate followed by the lip, buccal mucosa, floor of the mouth, tongue, tonsil, pharynx, retro molar area and nasal cavity (3-6).

A literature search was performed on PubMed by searching using the keywords “pleomorphic adenoma”, “minor salivary gland” and “parapharyngeal space”. 97 articles were found. Articles that pointed to the presence of pleomorphic adenoma of minor salivary gland in the parapharyngeal space were selected. 9 manuscripts that enrolled a total of 11 patients were reviewed. It appears that the average age at which the disease is diagnosed is 38.5 years. Only one case of pediatric involvement is reported. The size of the diagnosis tumor is quite large with an average of 6.8 x 5.6 cm. The surgical approach used is variable: in 2 cases the mandibular swing approach (18.5%), in 6 cases the trans-cervical approach (54%), in 2 cases the combined trans-oral and trans-cervical approach (18.5%) and in 1 case only the trans-oral approach (9%). In no case has there been disease recurrence (7-15). (Table 1)

	Age (years)	Size (cm)	Treatment	Recurrence
Varghese et al. (2003) [7]	40	5x4	Mandibular swing approach	No
Brigger et al. (2006) [8]	16	4.5x3.5	Trans-oral approach	No
Hakeem et al. (2009) [9]	20	7x6	Trans-cervical approach	No
	53	6x5	Trans-cervical approach	No
Rawat et al. (2012) [10]	42	7x6.5	Mandibular swing approach	No
Hwang S et al. (2013) [11]	34	8x6	Trans-cervical + trans-oral approach	No
Akın et al (2014) [12]	50	5x4	Trans-cervical approach	No
	56	4x3	Trans-cervical approach	No
Laturiya et al. (2016) [13]	27	6.5x5.5	Trans-cervical + trans-oral approach	Not specified
Bist et al. (2017) [14]	60	10x8	Trans-cervical approach	No
Malhotra et al. (2017) [15]	26	12x10	Trans-cervical approach	No

Table 1 - Cases of pleomorphic adenoma of minor salivary glands in the parapharyngeal space and its management

According Varghese *et al*, Pleomorphic adenoma of the parapharyngeal space is rare and occurs *de novo* from displaced or aberrant salivary gland tissue within this space (7). Another source of such a tumour is the deep lobe of the parotid gland (16). In our case, the only symptoms of the tumor were dysphagia and a painless swelling. However, these lesions of the parapharyngeal space may show additional symptoms, such as otalgia, neuralgia, palsies of cranial nerves (IX, X, XI) or trismus. Classical findings of benign parapharyngeal swelling are a submucosal swelling in the lateral pharyngeal wall with a possible extension to retromandibular fossa or the submandibular region, and is characteristically ballotable on palpation (16-18). CT scan and magnetic resonance (MR) are gold standard diagnostic tools in these cases to determinate the extent of disease, local spread and also help to determine the type of tumour. Contrast enhancement is important for differential diagnosis with vascular and neurogenic tumours. Also an intact fat plane helps to distinguish benign or malignant tumours. The presence of a layer of fibroadipose tissue between the tumour and deep lobe of parotid can help to distinguish tumours of the deep lobe of a parotid gland from tumour arising *de novo* in parapharyngeal space(19). MR has been shown to be superior to computed tomography but in our case CT was enough to define the characteristics and limit of the lesion (20-22).

Fine needle aspiration cytology (FNAC) is the modality of choice for obtaining a biopsy sample for diagnosis because this technique avoids (2) tumour seeeding and the subsequent multinodular recurrence possible using incision biopsy (23). Histopathologically, pleomorphic adenoma is an epithelial tumour of complex morphology, possessing epithelial and myoepithelial elements arranged in a variety of patterns and embedded in a mucopolysaccharide stroma. The treatment of pleomorphic adenoma is essentially surgical (2,3,16,24). These tumours have a false capsule with possible dehiscences, and therefore a resection of an adequate margin of normal surrounding tissue is necessary to prevent recurrence(19). The parapharyngeal space is however, a complex anatomic region. Within this potential space are cranial nerves IX, X, XI, and XII, the sympathetic chain, carotid artery, the jugular vein and lymph nodes. In fact, our case evidenced a caliber reduction of the left jugular vein at the petrous bone and therefore balloon catheter was positioned in the left jugular by right transfemoral venous surgical procedure. Traditionally, the approaches of choice for this tumour in PPS are the transcervical and parotid approaches (25-26). The only exception is Hughes *et al*. (16) who recorded the use of mandibular osteotomy though only in 2% of resections. Overall, this technique is useful in the case of malignant tumours. Another possible approach is the transoral approach described for the first time by Ehrlich (27) but we prefer to use trans-oral approach in small, non vascular tumours (28-33) due to there being no adequate control of an eventual haemorrhage. Therefore McElroth *et al*. (34) use this approach along with ligature of the external carotid artery to remove PPS tumours. For this reason, we consider the trans-cervical approach the gold standard for large benign PPS tumours.

In conclusion, pleomorphic adenoma arising *de novo* in the parapharyngeal space is rare. The goal in such cases is a correct diagnosis and *in toto* excision to avoid any possible recurrences.

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