Solitary benign follicular hyperplastic lymphoid tumor in parapharyngeal space

Solitärer gutartiger folliculärer hyperplastischer lymphoider Tumor im parapharyngealen Raum

Abstract

Tumors of the parapharyngeal region account for only 0.5% of all the head and neck tumors. This article presents a case of a 36-year-old female, who presented with an extensive mass in the right preauricular and parapharyngeal space. The tumor was excised successfully using a transcervical approach with temporary lateral mandibulotomy. The histopathology report showed a “solitary benign follicular hyperplastic lymphoid tumor in parapharyngeal space”.

Introduction

The parapharyngeal space (PPS) is an inverted pyramid-shaped deep space in the head and neck region. The bottom and apex of the inverted pyramid are the skull base and the hyoid bone pyramid, respectively. The PPS is medially bounded by the pharyngo-basilar and the bucco-pharyngeal fascia and laterally bounded by the superficial layer of the deep cervical fascia curving around the medial surface of the pterygoid muscles. The tensor-vascular-styloid (TVS) fascia, a layer that extends from the inferior border of the tensor velipalatini (TVP) muscle and posterior-laterally and inferiorly to the styloid process and muscles, separates the PPS into a pre-styloid compartment and a post-styloid compartment (Figure 1). Posterior to the styloid musculature lies the carotid artery with jugular vein and sympathetic chain with cranial nerves IX through XII [1]. PPS tumors are very rare and account for only 0.5% of all head and neck tumors [2].

Zusammenfassung

Nur 0,5% aller Kopf-Hals-Tumoren sind im Parapharyngealraum lokalisiert. Dieser Artikel berichtet über einen Fall einer 36-jährigen Frau, die sich mit einer großen Raumforderung im rechten Parapharyngealraum vorstellte. Der Tumor wurde erfolgreich über einen transzervikalen Zugang mit temporärer, lateraler Mandibulotomie entfernt. Der histopathologische Befund zeigte einen „solitären benignen folliculären hyperplastischen lymphoiden Tumor im Parapharyngealraum”.

Figure 1: Schematic axial view of pre-styloid (yellow) and post-styloid (pink) parapharyngeal spaces. IJV = internal jugular vein, ICA = internal carotid artery

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Both benign and malignant tumors may arise from any structure contained within the PPS. 70–80% appear to be benign and 20–30% appear to be malignant. Most of the tumors arising from the posterior compartment are of neurogenic origin, while salivary gland tumors are predisposing the anterior compartment [3]. Approximately 50% of the tumors have a salivary origin, 20% are neurogenic and the remaining 30% are represented by tumors such as benign and malignant lymphoreticular lesions, metastatic lesions and carotid body tumors. Schwannomas (neurilemmomas) are the most common neural tumors next to salivary gland tumors found in the PPS [4].

Case description

A 36-year-old female patient attended the outpatient clinic of the oral and cranio-maxillofacial surgery department with an increasing chief complaint of difficulty in swallowing. Other symptoms were otalgia, neuralgia and foreign body sensation in the throat, voice changes, and obstructive sleep apnea for the past 10 years. She went to different specialists, general and ENT surgeons. They recommended her not to undergo surgery due to high surgical risks.

The patient neither had any significant medical or dental history nor any deleterious habit. On clinical examination, a swelling was present in the right periauricular region that was oval, measuring approximately about 3 x 4 cm in size and was firm and painless. None of the cervical lymph nodes were palpable.

On intraoral examination, the uvula was pushed towards the left side with bulging of the soft palate with a smooth overlying mucosa. The right tonsil and the airway were pushed towards the midline of the oropharynx with an extension to the upper border of the laryngopharynx. Due to the massive mass the soft palate was not moving during the “Ah” sound.

An initial fine-needle aspiration cytology (FNAC) evaluation of the right soft palate bulged mass led to the initial diagnosis of a hemangioma. Based on this finding, a MR angiography study was conducted and a large, well defined tumor in the right periauricular and parapharyngeal region measuring about 7.5 x 5.5 x 3 cm in size was seen (Figure 2). It showed no contact with the internal carotid artery (ICA). However, vascular structures could be identified which were communicating with the right side external carotid artery (ECA).

During surgery, tracheostomy was performed first. It was planned to excise the tumor via a standard periauricular and trans-cervical approach in combination with a temporary mandibulotomy. The periauricular incision was not sufficient to address the intended tumor even though the superior part of the tumor bulged and protruded through the sigmoid notch of the mandible. Therefore, visor incision was employed additionally 2.5 cm below the inferior border of the right mandible along a skin crease extending from the right angle of the mandible to the hyoid bone. After reflecting the flap, the facial artery and vein were ligated. The right submandibular gland and lymph node were spared. The marginal mandibular nerve was identified and reflected superiorly. Right mandibulotomy procedure was performed. After retracting the right medial pterygoid muscle laterally the apex of the tumor could be exposed. The rest of the dissection could be managed bluntly with the finger the outer surface of tumor up to the skull base and finally the tumor could be removed in total (Figure 3). During the whole procedure, no association was revealed between the mass and ICA or ECA. During closure, the osteomized mandibular bone was fixed with 2.0 mini-plates.

The final diagnosis “solitary benign follicular hyperplastic lymphoid tumor” was confirmed on histopathological examination (Figure 4).
Figure 3: A) Visor incision was performed 2.5 cm below the inferior border of right mandible along a skin crease extending from the right angle of mandible to hyoid bone. B) Shows tumor exposed from PPS. C) Approaches to PPS: transoral + mandibulotomy (green); transcervical submandibular (yellow), transparotid (blue), and transcervical + mandibulotomy (red). D) Finally excised PPS tumor.

Figure 4: A) A hyperplastic follicle with cellular polarization: an eccentric mantle cap, a pale zone and dark zone from top to bottom. Some tangible body macrophage are evident and they are more prominent in dark zone. B) Although these neoplastic follicles have thin mantle, they lack cellular polarization and tangible macrophage.

Discussion

PPS tumors, although rare, are always a challenge for surgeons because of their complex anatomy, scarce symptomatology and difficulties in diagnosis. Most of the PPS tumors are benign (70–82%) [5], [6], [7], [8], [9]. The most common PPS tumors originate from the salivary gland tissue of the deep parotid lobe or other salivary tissue. In some cases, PPS tumors are originated from ectodermal residua associated with the cranial nerves and the cervical sympathetic chain or from the glomus body [10]. Tumors with a salivary gland etiology are with 30–48% the most frequent, followed by tumors of neurogenic origin (20–32%) [5], [6], [7], [8], [9], [11]. Just a small number of studies claim tumors with neurogenic origin as the most frequent [5], [12]. PPS tumors with neurogenic origin are often paragangliomas [6], [10], [12]. However, there are multiple other and sometimes very rare entities like mucoepidermoid carcinoma, adenoid cystic carcinoma, salivary malignancies, lymphoretic-
ular lesions, ganglio-neuroblastoma, myofibroblastic sarcoma or metastatic diseases described in other publications [4], [5], [6], [7], [9], [11], [12], [13]. The most frequent symptoms reported in literature are swelling of the neck (46%), pain (20%), dysphagia (13%), and pharyngeal wall swelling (9%) [9], [12]. Adequate 3D imaging is mandatory in these tumors to analyze the following parameters: pre- or post-styloid localization, size and extension, relation to the parotid gland and big cervical blood vessels as well as tissue characteristics of the tumor. Imaging methods contributed the most to diagnosis.

MRI and CT are recommended for imaging of the PPS [9]. However, some authors have reported the superiority of MRI over CT because of its better soft tissue differentiation and the possibility of combination with MRI angiography for better identification of the carotid artery [10], [12]. In contrast, CT imaging, although less precise in soft tissue evaluation, better depicts the styloid process and possible bony erosions [12]. Anterior displacement of the carotid artery and the internal jugular vein is suggestive for post-styloid tumors. This can be seen mostly in neuromas, schwannomas and paragangliomas, while salivary lesions rather displace the vessels posteriorly [9], [14]. In pre-styloid tumors, a preserved fat plane between the deep lobe of the parotid gland and the tumor is indicative of a separate tumor originating from the extra-parotid minor salivary glands [7]. CT with contrast and MR imaging with gadolinium are often used together to provide complementary information, which allows delineation of size, exact localization and probable cause of these tumors [6], [15].

The PPS is a relatively dangerous space to access due to the hiding behind the mandible and the close localization to the superficial parotid lobe, the facial nerve, the carotid artery and the jugular vein, lingual nerve, spinal accessory nerves and hypoglossal nerve. Surgery is the method of choice for PPS tumor management. The surgical access should be planned foresightful on the bases of the preoperative imaging results. They can be accessed using a transcervical-transparotid or transcervical approach with or without temporary mandibular osteotomy. The choice of approach should be dictated by tumor localization, size and its relation to the carotid artery and jugular vein. Also, the relation to the skull base and of course whether it is a benign or a malignant tumor should be considered. External transcervical approaches are preferred for better neurovascular control and surgical radicality [7], [11], [16]. In literature, a transcervical approach is preferred in the majority of the cases. It is indicated for the removal of tumors originating from the minor salivary glands as well as for Schwannomas and Paragangliomas in the post-styloid space [7], [8], [12], [17]. The transcervical approach allows direct access to the PPS. Removal of the tumor is often improved by removal of the submandibular gland, external and facial vessel ligation and dividing digastric, stylohyoid and styloglossal muscles and stylo-mandibular ligament and anterior dislocation of the mandible [1], [8], [10], [11]. The transcervical-transparotid approach is suitable for pre-styloid tumors arising in the deep lobe of the parotid gland [3], [5]. Parotidectomy incision is extended to the level of the greater cornu of the hyoid bone. This approach requires identification and preservation of the main trunk of the facial nerve. The transoral approach can be used in carefully selected cases [5], [6], [9], [16], [18]. Other authors do not recommend an intraoral approach because of the risk of injuring the lingual blood vessels and the risk for uncompleted tumor extirpation [7], [11], [12]. A combination of a transcervical and transoral approach may be used for the removal of benign sizeable tumors without mandibular osteotomy [5], [16].

In cases in which a transcervical approach in combination with a temporary mandible osteotomy is needed, it is recommended to secure the airway by tracheotomy first [9], [11], [12]. Osteotomy can be performed at the mandibular ramus or in the median or premolar region of the mandible without harming teeth and the mandibular nerve. Literature describes both single [19] and double osteotomies [3], [7], [16], [20]. Malignant tumors of the PPS often require adjuvant therapy (radio and/or chemotherapy). The main reasons for this are high-grade malignant tumors and unclear surgical margins [7], [12], [16]. In literature, data on PPS tumor control are often incomplete. The frequency of benign tumor recurrence seems to be low [5], [7], [9]. In contrast, malignant PPS tumors seem to have a poor prognosis [5], [7], [8], [11], [15]. Pathohistological diagnosis has to be performed before surgery. In addition to open incisional biopsy which may carry the risk of tumor dissemination, tumor rupture and/or hemorrhage [11], [12], FNAC may be helpful for preoperative diagnosis of PPS tumors [1], [12], [13], [14], [21], [22]. Unfortunately, as it was in the presented case, FNAC correlates with definite histopathological findings in 65% only. However, the sensitivity for detection of malignant diseases seems to be with around 90% more reliable [7]. The main benefit of FNAC in evaluating PPS tumors is the low risk of complications. Therefore, it should be used as method of first choice to diagnose those tumors. It is particularly recommended when lymphoma, metastatic lesions and other malignant neoplasms are suspected. In contrast, it seems to be not as meaningful in paragangliomas [7], [9]. The most common postoperative complications are facial nerve lesions or lesions of cranial nerves in patients with removed neurogenic tumors [11] or malignant tumors and paragangliomas [7], [12], [15], [16]. In rare cases, first bite syndrome is described in the literature as well [23]. Many squeal of surgical treatment are transitory and a permanent deficit is described in 11% [12].

Conclusions

Parapharyngeal solitary benign follicular hyperplastic lymphoid tumors are rare benign neoplasms. They are
usually detected late after considerable signs and symptoms appear. FNAC should be employed for preoperative diagnosis first, because open incisional biopsy may carry the risk of tumor dissemination, tumor rupture and/or hemorrhage. CT or MRI are the preferred imaging modalities and total surgical excision is the treatment of choice. Surgical access should be planned foresightful based on the preoperative imaging results to prevent unnecessary incisions. Submandibular visor incision in combination with a temporary mandibulotomy enables the best vision in the PPS during surgery and can be recommended for the resection of large PPS tumors. In non-malignant tumors blunt dissection by one finger is mostly adequate for tumor removal, even when the tumor extends up to the skull base. Recurrence of benign PPS tumors is rare after complete excision.

Notes

Competing interests

The authors declare that they have no competing interests.

References


Erratum

By mistake, two authors were missing in the original publication. They have been added.