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Case Report

Massive Recurrence of a Giant Pleomorphic Adenoma of the Hard Palate

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Abstract

Pleomorphic adenoma PA is the most common salivary gland tumor. Although benign, PA is known for a high propensity for recurrence which depends on the initial management. It occurs usually in major and accessory salivary glands specially the parotid gland. The localization of the disease in the hard palate is relatively rare. We report the case of a 63 years-old woman who presented a massive recurrence PA of the hard palate after inadequate initial management leading to a therapeutic impasse. We tried to focus on the clinical and histological features of PA and analyze the different circumstances leading to recurrence.

Keywords: Pleomorphic Adenoma; Hard Palate; Recurrence; Surgery; Radiation Therapy

Introduction

Pleomorphic adenoma (PA) or ''mixed tumor, salivary gland type", in reference to its dual origin of epithelial and mesenchymal elements, is the most common tumor of salivary glands accounting for about 40% to 70% [1]. Large salivary glands especially the parotid gland is the most concerned. These tumors may also arise from minor salivary glands in different parts of oral mucosa; the palate is the most common site followed by the upper lip and the buccal mucosa [2]. PA usually presents as a slow enlarging, firm and painless mass which doesn't alarm patients who often seek medical consultation lately. Although considered as benign tumor, there is a significant risk of malignant transformation to carcinoma ex pleomorphic adenoma in 3% to 7% of cases [3]. Surgery, with complete removal of the tumor is the basis of the treatment [4]. Incomplete resection expose to a high risk of recurrence [5,6]. Adjuvant radiation therapy (RT) could be of great help in cases of unclear margin or gross residual disease [7].

Observation

A 63 old-years woman presented in Salah Azaez institute, head and neck surgery department with a massive enlarging oral mass as chief complaint. Nasal obstruction, headache and difficulties in breathing and swallowing solid food were also reported. In medical history the patient was treated three years ago for a tumor of the hard palate by surgical excision and adjuvant RT due to insufficient surgery with positive margins. Clinical examination found a moderate limitation of mouth opening and a gross firm painless oral mass with large implantation in the hard and soft palate extended posteriorly to the pharyngeal wall and laterally to the right buccal mucosa measuring 6cm in greatest diameter. Nasal endoscopy showed a fungating mass filling the left nasal cavity extended to the choane and the rhinopharynx. CT of the facial massif showed voluminous-88x49x66 mm process with tissular and cystic components in the right side of the rhino and oropharynx, with extension to the right deep facial spaces, pterygoïd muscles and the left nasal

cavity. Biopsy concluded to a mixed tumor, salivary gland type. A palliative surgical treatment was performed including right hemi-palatectomy and reductive surgery of the tumor. A sufficient temporary liberty of airways was obtained.

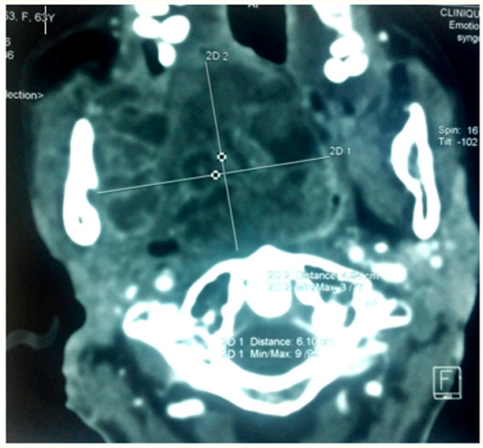

Figure 1: Facial massif CT scan, axial section showing a voluminous process of hard and soft palate extended to buccal mucosa, parapharyngeal and prevertebral spaces.

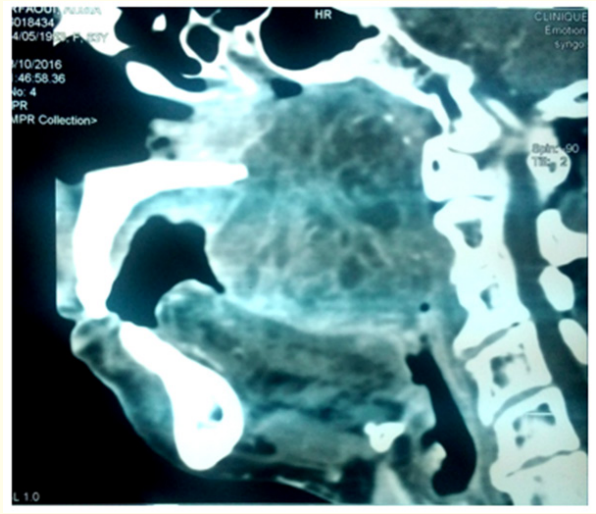

Figure 2: The same process as fig-1 in sagittal section.

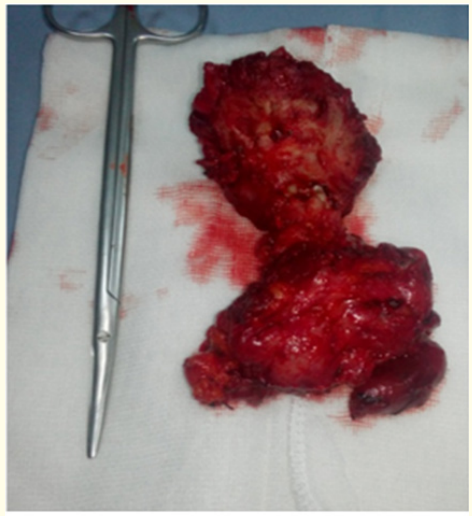

Figure 3: Surgical specimen.

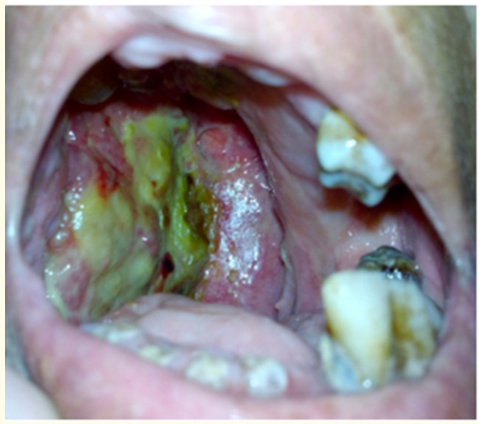

Figure 4: Oral cavity aspect one month after surgery.

Discussion

Salivary glands tumors are relatively uncommon counting for about 2 to 3% of all tumors and 5% of head and neck tumors [8]. Tumors occurring in minor salivary glands account for 20% to 40% of all salivary glands precisely 22% according to Spiro [9].73% of

salivary glands tumors are PA [10]. PA often affects large salivary glands with predilection for the parotid gland, less frequently accessory and minor salivary glands. Corresponding to minor glands, the palate is the most common site for PA followed by the upper lip and the buccal mucosa [2].

As presented in this case, PA usually occurs in the fourth to the sixth decade with slight predominance of the female gender [5,11]. Mixed tumor usually manifest with a slow enlarging, firm, and painless mass [4,12]. Few cases showing ulceration, bleeding and pain were reported by Pigaroo and al, were attributed to repeated trauma of the lump due to chewing [4].

Authors reported in the same series a large delay between first symptoms and diagnosis ranging from 3 months to 10 years [4]. The differential diagnosis includes mainly, palatal abscess, cysts, malignant tumors, and oral papilloma [10].

Diagnosis of PA is established on basis of history, physical examination and histology. An incisional biopsy is necessary to confirm the diagnosis [13]. Histological examination emphasizes the dual origins of mixed tumor showing the epithelial and myoepithelial elements arranged in a variety of patterns within a mucopolysaccharide stroma. The tumor does not have a true capsule and extends through normal glandular tissue in the form of finger-like pseudopodia [14,15].

A recurrent genetic event, consisting of chromosomal rearrangement most commonly t (3; 8) (p21: q12), was recently identified in all major cytogenetic groups of PA [16]. This event is thought to be responsible of promoter swapping between PLAG1 characterized as a proto-oncogene and CTNNB1. PLAG1 is also involved in the second most frequent translocation of PA t (5; 8) (p13; q12) which results in increased expression of PLAG1 [17]. This plays in the favor of the theory that PA cells originate from a single pluripotent cell type capable of differentiation into a variety of somatic phenotypes [18].

Malignant transformation and propensity for recurrence are the two big deals about PA. In fact, carcinoma ex pleomorphic carcinoma (Ca ex PA) is a carcinomatous transformation within a primary mixed rumor [19]. It counts for about 6.2% of all PA and about 11.6% of all malignant salivary glands tumors. Ca ex PA usually occurs in parotid and submandibular glands. Minor salivary glands are not spared especially for the soft and hard palate [20]. A rarer phenomenon concerning some malignant behavior of PA was described. In fact few metastatic cases of PA were reported although the primitive tumor remains histologically benign [21,22]. Therefore metastatic PA is recognized as a histological sub-type of PA by the world health organization [23].

Although benign tumor, PA management could be difficult due to its propensity for recurrence. Microscopic finger-like or pseudopodia of neoplastic cells may extend beyond visible tumor [24]. Leaving these extensions during an inadequate surgery may result in recurrence even after many years. Capsular violation, tumor rupture and simple enucleation must be avoided as well when dealing with PA [4,5]. Recurrent mixed tumors are typically multinodular with satellite islands within the remaining salivary gland [10]. Abandoning enucleation technique reduced dramatically recurrence rate of PA from 20 to 40% to less than 4% [25,26]. Mixed tumor of minor salivary gland are less likely to relapse. In a series of 20 cases of PA of hard palate operated, no cases of recurrences were reported in one years of follow up [4]. In English literature 2 cases of palatal PA recurrence in children out of 16 were reported [6,27]. Although reported to be risk factors for recurrence for some authors, patient age and sex are controversial [28]. In our case we attribute the recurrence probably to an insufficient initial surgery. We do not dispose of data concerning the initial operation. Adjuvant RT was indicated because of involved surgical margins. Residual disease in the parapharyngeal space is another hypothesis. According to Polat and colleagues, parapharyngeal space is a close-up space with no barrier to tumors extension and it and hold in many anatomic limitations for surgery [5].

Management of recurrent PA should involve a combination of surgical resection and adjuvant RT [5,7]. In fact salvage surgery is less likely to be curative than initial complete resection [5]. Although carrying the risk of malignant transformation and a higher long-term morbidity, adjuvant RT could be of great contribution in controlling multi-nodular and residual disease [7]. Renehan and colleagues in an analytic study of recurrent PA of the parotid gland reported that surgery alone has a recurrence rate of 43% as opposed to surgery and RT a 4% cumulative recurrence rate at 15 years [29]. In our case, adjuvant RT was not performed because of the high morbidity due to the previous irradiation. Besides the debulking surgery kept a voluminous residual tumor, we judge adjuvant RT was useless.

Many methods of reconstruction of soft tissues and bony defects were reported in the literature. Reconstructions technique using palatal, buccal mucosa and obturator flaps were used [4,6]. In our case, the defect was large as well as the residual tumor and as a result reconstruction was not possible.

Conclusion

PA is the most common tumor occurring in minor salivary glands. The hard palate is a relatively frequent localization. Local control of PA could be difficult due its high propensity for recurrence. Initial surgery may be decisive. A total resection with large free margins is recommended. Adjuvant RT could be of great help in case of residual disease inaccessible to surgery or when surgical margins are involved. Finally a long term follow-up is necessary due to the risk of late recurrence after many years.

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