Case report

Giant pleomorphic adenoma of the parotid gland: an unusual case presentation and literature review

Adenoma pleomorfo gigante della ghiandola parotide: caso clinico e revisione della letteratura

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SUMMARY

Pleomorphic adenoma is the most common type of all salivary gland tumours. Although uncommon, cases of giant pleomorphic adenomas have been described in the medical literature, the majority involving the parotid gland. This paper describes an unusual case of a giant adenoma arising in the parotid gland. The patient underwent surgical resection of the giant tumour, which was one of the largest pleomorphic adenoma reported in recent literature. This case has prompted us to evaluate the behaviour of those benign tumours, which suggested that aesthetic and social morbidity is sufficient to justify, when possible, early tumour excision, despite the relatively low risk of malignant transformation. Management of this unusual tumour is discussed, and the literature on giant parotid tumours is reviewed.

KEY WORDS: Giant pleomorphic adenoma • Parotid tumours • Neck mass

RIASSUNTO

L'adenoma pleomorfo rappresenta il più comune istotipo tumorale interessante le ghiandole salivari. Quantunque non siano comuni, la letteratura medica internazionale ha descritto alcuni rari casi di adenoma pleomorfo gigante. La maggior parte di essi era di origine parotidea. Questo articolo vuole descrivere un inusuale caso di adenoma gigante di origine parotidea. La crescita abnorme che la massa tumorale ha avuto in questo caso, ne ha determinato la discesa per gravità verso il torace del paziente. Il paziente è stato sottoposto ad exeresi chirurgica del tumore, che è apparso essere uno dei più voluminosi adenomi pleomorfi riportati nella letteratura recente. Questo atipico caso ci ha portato a riflettere in merito al comportamento clinico di siffatte forme tumorali benigne. È evidente come le morbidità estetiche e sociali siano sufficienti a giustificare, quando possibile, una precoce asportazione del tumore, malgrado il relativamente basso rischio di trasformazione maligna. Di seguito discuteremo la gestione clinica e chirurgica di questo raro caso. Forniremo inoltre una revisione della letteratura in merito.

PAROLE CHIAVE: *Adenoma pleomorfo gigante • Tumori parotidei • Tumefazioni cervicali*

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Introduction

About 70% of all salivary gland tumours arise in the parotid gland, and approximately 85% are benign; of these, 80% are pleomorphic adenoma [1]. These tumours are almost uniformly characterized by a slow-growing, painless mass, usually varying from 2 to 6 cm in diameter when resected [2]. In 4% of cases, tumours may be malignant.

Cases of giant pleomorphic adenomas have been reported in the parotid gland, presenting as an irregular multinodular mass that can weigh more than 8 kg [3]. Most cases of giant adenomas were seen before the 1980s, but some have been published recently [4]. Given the relative ease of diagnosis of pleomorphic adenomas based on clinical and cytological findings, and the low risk of malignant degeneration, some authors have an expectant management policy for those patients who do not desire surgery [1]. This is in contrast to traditional management, which emphasizes that "aggressive treatment of primary and recurrent mixed tumours is necessary" [5] due to their malignant potential.

This paper describes an unusual case of a giant pleomorphic adenoma arising in the parotid gland, along with the reasons for diagnostic delay. This case prompted us to evaluate the behaviour of benign tumors that do not undergo malignant transformation. This case and others like it demonstrate that aesthetic and social morbidity is sufficient to justify, when possible, early tumour excision, despite the relatively low risk of malignant transformation.

Case report

An 83-year-old Caucasian man complained of a slow-growing mass on the left side of his face for more than 30 years. The patient's social history was significant for the events surrounding dismissal from work. The patient lived alone, isolated from his family, and was homeless. He denied knowledge of any past illness, but admitted to not having seen a physician for more than 30 years. He affirmed that the mass had begun to develop about 30 years ago, and had slowly enlarged without symptoms. Since that time he refused any interaction with his family and healthcare providers.

The patient came to our attention when the mass has become too large to allow normal walking. After a week of frequent visits, we finally gained the patient's confidence and took over his management.

Clinical examination showed a giant, firm, multinodular, irregular and painless mass measuring approximately 35 cm x 28 cm, involving the left parotid and cervical region (Fig. 1). The arrangement of the mass along the chest had the typical appearance of a beard.

Despite of the large dimension of the mass, there were no signs of facial nerve palsy, and the skin that covered the lesion did not present ulcerated areas. CT scan and magnetic resonance angiography were performed to evaluate the extent of the mass (Fig. 2). The lesion had an arterial blood supply from the facial artery and venous drainage in the internal jugular vein (Fig. 3). The main hypothesis for diagnosis was a benign tumour of the parotid gland, most likely pleomorphic adenoma.

Incisional biopsy was performed and confirmed the suspicion of adenoma.

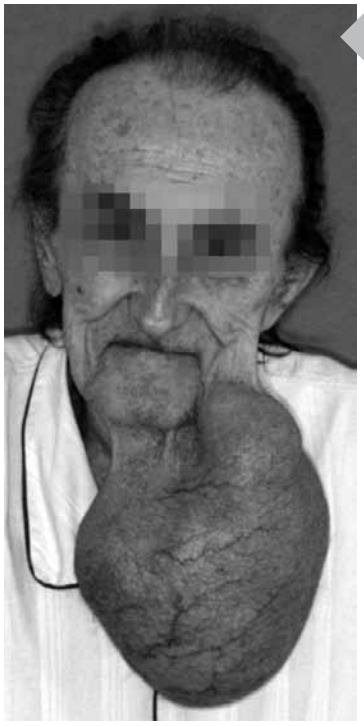

Fig. 1. The patient at initial examination, showing a large, multinodular left neck mass extending from the parotid region onto the chest.

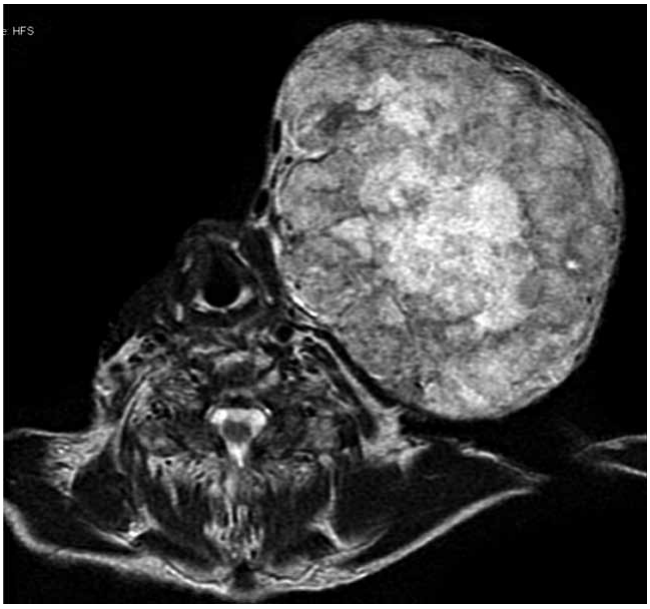

Fig. 2. Pre-operative imaging evaluation. MRI showing the giant mass. The outline of the heterogeneous lesion is clearly demarcated, and tissue planes preserved.

The tumour was excised under general anaesthesia. Despite the size of the mass, a clear plane of dissection was found. The sternocleidomastoid muscle was greatly hypertrophied, and the tumour displaced it posteriorly. Due to many years of traction, the mass had descended well below the angle of the mandible and the facial nerve was avoided without difficulty. Skin flaps were raised off the sides of the mass to provide sufficient tissue for neck closure.

The post-operative course was uneventful.

Macroscopically, the excised mass measured 33 cm x 27

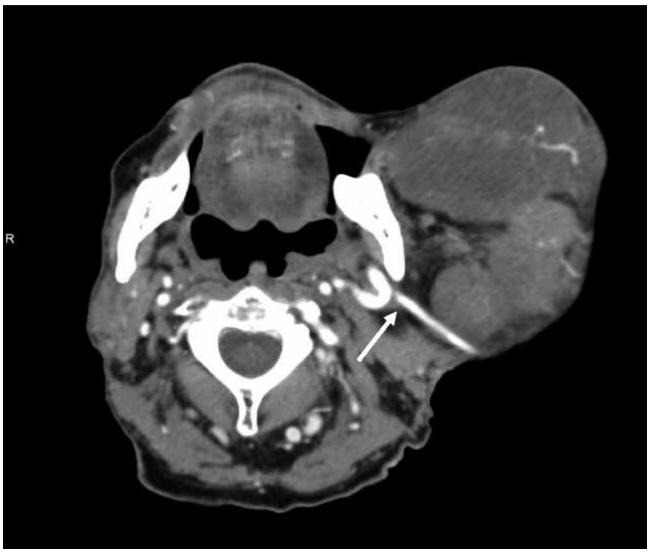

Fig. 3. Pre-operative CT scan showing the tumour pedicle (arrow) arising from facial artery. It measured about 15 cm in length, and demonstrated numerous feeding vessels.

Fig. 4. Intra-operative image showing the multinodular tumour and the main vascular pedicle (arrow).

cm x 16 cm and weighed 7.3 kg (Fig. 4).
Microscopically, the tumour was composed of islands and strands of epithelial cells in a hyaline stroma. Spindle and plasmocytoid myoepithelial cells in a myxoid stroma were also abundant. All areas of the surgical specimen were microscopically analyzed, and none showed evidence of malignant transformation. On histological analysis and immunohistochemistry, the lesion was identified as a pleomorphic adenoma with negative surgical margins.
The patient presented good aesthetic and functional results (Fig. 5), without signs of facial nerve dysfunction (House Brackmann grade I). At five-year follow-up the patient was doing well, without clinical or radiographic evidence of recurrence.

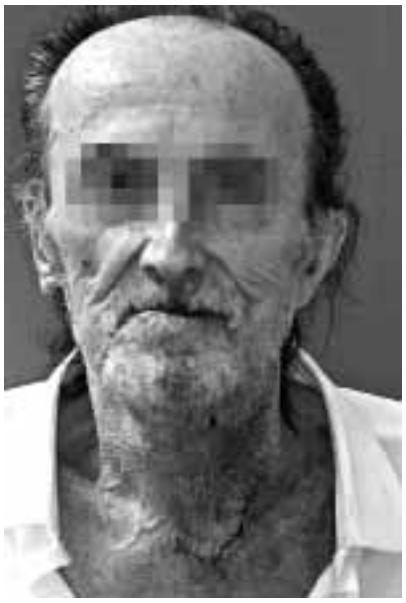

Fig. 5. Post-operative image showing clinical aspect after giant tumour removal with facial nerve preservation.

Discussion

Pleomorphic adenoma is the most common salivary gland tumour. The main site of occurrence is the parotid gland, affecting patients of any age, most frequently between the fifth and sixth decades of life [3]. Although uncommon, cases of giant pleomorphic adenomas have been described the majority of which involved the parotid gland. The first case of giant pleomorphic adenoma published in medical literature was reported by Spence in 1863 [6], who described the treatment of a mixed tumour > 1 kg.
In 1956, Short and Pullar [7] published an English language review of massive pleomorphic adenomas and a case-report of a 2.3 kg adenoma. In 1989, Schultz-Coulon [8] reviewed 31 cases of giant pleomorphic adenomas of the parotid gland. The author found a female predominance (64.5%), with an age range from 20 to 40 years old, and a tumour weight between 1 to 27 kg.
Buenting [9] reported the 10 largest pleomorphic adenomas published in the English language literature, and found a mean tumour weight of 7.8 kg; nine of 10 occurred in females, with a mean age of 56 years. His case was the 5th largest pleomorphic adenoma reported (6.85 kg).
In our case, the patient was a man who presented a mass with a 30-year history of evolution that weighed 7.3 kg, more than the Buenting case report.
In most of the cases described in the literature, the lack of information and patient's negligence are considered as the most relevant aspects influencing the treatment delay. In our case, it must be considered that the patient was homeless and away from his family.
The incidence of malignant transformation in adenomas ranges from 1.9% to 23.3% [10]. The risk increases in tumours with long-standing evolution, recurrence, advanced age of the patient and location in a major salivary gland [11]. Some authors postulated that the risk of malignant transformation increases from 1.6% in tumours with less than 5 years of evolution, to 9.5% for those presenting for more than 15 years [12].
The classic clinical history of carcinoma ex-pleomorphic adenoma is a slow-growing mass for many years, with a recent fast growth phase. A case of a giant PA with malignant transformation with this typical history was reported in 2005 by Honda [4] in a 72-year-old woman with a slow growing parotid lesion for 20 years, with a rapid increase in the last 3 months.
In the Schultz-Coulon [8] review, 3 of 31 cases of giant adenomas showed areas of malignant transformation. In our case, although the patient presented all the characteristics for an increased risk of malignancy, clinically and histologically there was no such evidence.
Neglecting even a benign parotid tumour carries an increasing risk of facial nerve injury when treatment is performed. The bony and muscular deformity associated with such tumours is uniformly disfiguring and incapaci-

tating. Although it is generally accepted that the majority of all giant adenomas remain non-malignant, this case and others like it should serve to remind us that the clinical course of these masses can be far from benign. We believe that early excision of parotid pleomorphic adenomas is desirable if the patient will tolerate surgery.

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