Case report

Synovial sarcoma of the parotid gland: a case report and review of the literature

Sarcoma sinoviale della ghiandola parotide: caso report e revisione della letteratura

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Summary

Parotid gland tumours are very heterogeneous, being benign in 80% of cases, and generally arising from epithelial cells. Nevertheless, a small group of non-epithelial tumours representing just 5% of all salivary gland neoplasms has also been reported, the most common of these being haemangioma, especially in children. However, lymphomas, neuromas, neurofibromas, lipomas and sarcomas can also be found. Synovial cell sarcoma is a high grade histological variety of sarcoma and is generally located near large joints and bursae of the lower extremities, such as knee, tendon sheaths and bursal structures. It is rarely found in the head and neck region due to its lack of synovioblastic tissue. Herewith, the case of a young female, affected by a synovial sarcoma of the left parotid gland, is presented and a review is made of the literature on this rare specific localization focusing on management and outcome.

Key words: Head and neck sarcomas • Parotid gland tumours • Surgery of parotid gland • Synovial sarcoma

Introduction

Sarcomas are malignant soft tissue tumours representing only 1% of all malignant tumours and 1% of all head and neck malignancies. Synovial cell sarcoma (SS) is a high grade histological variety of sarcoma and it is the fourth most common entity after malignant fibrous histiocytomas, liposarcomas and rhabdomyosarcomas. It typically occurs in young adults with a male/female ratio of 2:1. It is located predominantly near large joints and bursae of the lower extremities such as knee, tendon sheaths and bursal structures, but is rarely found in the head and neck (representing only 3% to 10% of synovial cell sarcomas and quite often the correlation with synovial structures remains unclear), because this region is poor in synovioblastic tissue. However, the most common sites of localization, in the head and neck region, are hypopharynx and parapharyngeal spaces. In fact, it is possible to find the majority of SSs in the paravertebral connective tissue spaces, while they are less common in the larynx. Histological diagnosis is generally a late post-operative finding. Due to the rarity of this entity, few cases have been described in the literature and treatment is still not well codified.

Case report

A 31-year-old female had an 8-month history of a progressive swelling in the left parotid region. Physical examination revealed a firm mass 3 x 2 cm, associated with local facial pain and ipsilateral otalgia. Ultrasound examination revealed an enlarged left parotid gland, occupied, in the middle-upper region of its deep lobe, by a moderately expansive process measuring 31 x 21 mm with a solid anechoic structure, suggesting a Warthin tumour. Magnetic resonance imaging (MRI) confirmed a high volume left parotid mass, with a normal gland profile, without involvement of adjacent vascular structures, bones or adipose tissue. Regional lymph nodes were not enlarged.
Fig. 1. Pre-operative MRI FSE DP/T2 weighted with contrast in axial plane (A) and FLAIR TRS in coronal plane (B) showing a left parotid mass close to the mandibular ramus and the temporo-mandibular joint.

Fine-needle aspiration biopsy (FNAB) revealed normal acinar cells with oncocytic metaplasia. The patient denied any history of trauma or facial surgery. Total parotidectomy with facial nerve preservation was then carried out and was followed by a regular post-operative course. Histology revealed an unencapsulated, lobulated, whitish solid mass composed of cellular fascicles and sheets of uniform, small, ovoid cells with plump and spindle-shaped pale nuclei and inconspicuous cytoplasm. Loosely arranged fascicles in a myxoid stroma and collagenized areas were observed. Vasculature was prominent producing a haemangiopericytoma-like pattern in most areas. Immuno-histochemistry revealed tumour cell positivity for vimentine, CD99, Bcl-2 and focally for epithelial membrane antigen (EMA), whereas no reactivity for CD34, CAM5.2, KL1, AE1/AE3, CK14, S100, GFAP, p63, CD117, HHF35, ASMA was detected (Figs. 2, 3). Cytogenetic tests showed the characteristic \((x:18), \text{SyT/SSX1}\) translocation. Morphologic, immunophenotypic and molecular findings were compatible with primary monophasic synovial sarcoma. Therefore, the patient underwent ipsilateral selective neck dissection (negative for metastasis) and adjuvant radiotherapy (total dose 50 Gy). She is free of disease at 3-year follow-up.

Discussion

Salivary gland tumours are mainly located in the parotid (80%) and in the submandibular glands (10-15%) while sublingual and ancillary glands are less frequently involved (5-10%)\(^7\). About 80% of parotid gland neoplasms are benign and more than 95% occur in adults. Their histopathological aspects are heterogeneous. The most common parotid gland tumour is pleomorphic adenoma (60%) followed by Warthin adenoma (5-15%). As far as concerns malignant neoplasms the largest percentage is represented by mucoepidermoid carcinoma (8-15%) followed by adenoid cystic carcinoma (5%). Finally, there is a small group of non-epithelial tumours representing just 5% of all salivary gland neoplasms and generally they are benign. The most represented of these is haemangioma, especially in children. However we can also find lymphoma, neuromas, neurofibromas, lipomas and sarcomas. Synovial sarcoma (SS) is the fourth most common variety of sarcoma after malignant fibrous histiocytoma, liposarcoma and rhabdomyosarcoma\(^8,9\). Two subtypes of SS are described in the literature: monophasic and biphasic. The latter contains spindle and epithelioid cells, while the former contains only spindle cells. As far as concerns SS, 99% present an identifiable translocation between chromosome 18 and X that results in the fusion of the SYT genes located on chromosome 18 and the SSX-1 or SSX-2 gene on chromosome X. Finding this translocation is very important to confirm the histological diagnosis of SS, especially for the monophasic type allowing differential diagnosis with other spindle
cell tumours, such as haemangiopericytoma, neurogenic sarcoma, fibrosarcoma and leiomyosarcoma. Some Authors indicated biphasic tumours as more aggressive than the monophasic type, whereas another study was not in agreement with this difference and, indeed, found no differences between the mentioned subtypes. The American Joint Committee on Cancer has classified all synovial sarcomas as high grade malignancies, irrespective of their phenotypes. A systematic review of the literature, selective for SS arising from the parotid gland, has shown controversial opinions among the reported cases. Sex, age at onset, histopathological pattern, size, prognosis and treatment were then evaluated. Parotid gland SS occurs primarily in males with a high-prevalence rate in young adults (> 90% of cases presenting before 50 years of age). The biphasic type would appear to be more common than the monophasic type and generally more aggressive. Tumour size has been reported to be highly variable (from 0.6 mm to 7 cm) and appears to be correlated with prognosis as generally occurs in the other localizations of SS (tumours larger than 5 cm). Although several reports have suggested an aggressive course for head and neck synovial sarcoma, our case and the review of parotid localizations suggest a relatively good prognosis, probably due to the easy exploration and early recognition of every parotid swellings.

A review of the literature showed an overall disease-free survival at 10 and 15 years ranging from approximately 45% to 50%. This rate has improved since 1977 with a 40% 5-year survival reported in 1977, a 55% 5-year survival rate in 1982, a 56% 5-year survival rate in 1992 and a 60% 5-year survival rate in 1996. Long-term follow-up is necessary because of the high incidence of distant metastases especially to the lungs (main cause of death in approximately 50% of the patients).

An analysis of 36 cases of SS involving the head revealed a high prevalence of parotid localizations (14/36) suggesting that this region is the most affected in the head and neck area.

There are different theories regarding the aetiopathogenesis of SS. This tumour sometimes occurs in sites not related to the synovium such as parapharyngeal, retropharyngeal or hypopharyngeal regions, tongue, cheek or buccal pad, therefore, the histogenesis remains controversial. According to Leader et al., it is currently accepted that SS arises from undifferentiated or pluripotent mesenchymal cells with dual differentiation capacity, both epithelial and mesenchymal. This is why this Author suggests classifying SS as carcinosarcoma. This theory is in contrast with others describing a clear link with joints as beginning sites of these sarcomas.

Can we confirm the role of temporomandibular joints (TMJ) in the development of SS located in the parotid gland? The absence of SS arising in the other major salivary glands could be related to their distance from any facial joints. The optimal treatment of these tumours seems to be multimodal. Radical surgery represents the first approach. Post-operative radiation treatment has been found to improve the prognosis in head and neck localizations rather than in the extremities.

Chemotherapy with ifosfamide has been investigated especially in the hypopharynx and larynx where complete excision is sometimes not possible. Recently, some Authors suggested the possible role of epidermal growth factor receptor (EGFR) and human epithelial growth factor receptor 2 (Her-2/neu) in the carcinogenesis of SS thus suggesting that the anti-EGFR monoclonal antibody may play a role in the therapeutic approach.
Conclusions
Since synovial sarcoma of the parotid gland represents a rare entity, the diagnosis and clinical management can be a challenge. On account of the loco-regional and systemic spread, a multimodality therapeutic approach, as well as long-term follow-up are mandatory. The volume of the disease seems to be the only clear prognostic factor, such as the histopathological pattern. Parotid gland localizations of SS seem to be less aggressive, having a better prognosis compared with the other head and neck localizations.

References