International Poster Journal IPJ of Dentistry and Oral Medicine

Int Poster J Dent Oral Med 2008, Vol 10 No 03, Poster 415

Sarcoma-NOS of the Face. Clinic and histopathology of a rare case of a Soft Tissue Sarcoma

Language: English

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Date/Event/Venue:
June 26th -30th, 2007
The 14th International Congress of the International Confederation for Plastic, Reconstructive and Aesthetic Surgery,
Berlin

Introduction

Epidemiology

Sarcomas are particularly rare malignancies that comprise less than 1% of all cancer diagnoses in the United States (1, 2). They are a heterogeneous group of seldom tumours that arise predominantly from the embryonic mesoderm (3). The crude incidence of soft tissue sarcoma in the European Union is 1.0-3.0/100 000 per year, the mortality 0.6-0.8/100 000 per year (4).
Among the different sarcoma groups the soft tissue sarcomas are the most frequent. In general, soft tissue sarcomas do not seem to result from malignant changes or the dedifferentiation of benign soft tissue tumours, and despite the variety of histologic subtypes. However they have many clinical and pathologic features in common (3). They are diagnosed at any age but are more frequent in older patients with a peak incidence at the age of 50 years (3). Adult soft tissue sarcomas are a heterogeneous group of tumours, including well-described subtypes by histological and genotypic criteria, and pleomorphic tumours typically characterized by non-recurrent genetic aberrations and karyotypic heterogeneity (5).
Approximately 5% of soft tissue sarcomas were located in the head and neck region. Beside neck with 31.5% face with 20.7% is the secondly most frequent localization (6). Currently, more than 50 histologic types of soft tissue sarcoma have been diagnosed by genetic and morphological Criteria The most frequently seen include liposarcoma, leiomyosarcoma, fibrosarcoma, synovial sarcoma and malignant fibrous histiocytoma (MFH) (3). Lately it turned out then due to increasingly detailed pathological investigation techniques (cytogenetic analysis, Immunhistochemistry etc.) that most MFH is nevertheless typable. Those MFH which remain unclassifiable with the help of new histological techniques also, are to be designated in the future as pleomorphic sarcoma NOS (7). Some other authors maintain that the actual WHO-Classification uses pleomorphic malignant fibrous histiocytoma (MFH) and pleomorphic sarcoma NOS (not otherwise specified) synonymously (8). Sarcoma NOS comprised all pleomorphic spindle-cell sarcomas that did not show evidence of specific differentiation by the criteria outlined above. These tumours were also negative for epithelial, melanocytic, and hematopoietic markers (9).Overall survival is approximately 50% at 5 years; the key determinant of survival is control of both local recurrence as well as distant dissemination (10).

Clinical features

Soft tissue sarcomas most commonly present as an asymptomatic mass. Soft tissue sarcomas grow in a centrifugal fashion and compress surrounding normal structures, but rarely does impingement on bone or neurovascular bundles produce pain, edema, and swelling(3). The type of surgical resection is determined by several factors, including tumour location, tumour size, the depth of invasion, the involvement of nearby structures, the need for skin grafting or autogenous tissue reconstruction, and the patient's performance status. Local therapy consisting of surgery, either alone or in combination with radiation therapy when wide pathologic margins are limited by anatomic constraints, is the approach taken in patients with small (less than 5 cm) primary tumours with no evidence of distant metastatic disease (3). Soft tissue sarcomas are generally surrounded by a zone of compressed reactive tissue that forms a pseudocapsule, which may mistakenly be used by inexperienced surgeons to guide resection (enucleation). Microscopic extensions of tumor beyond the pseudocapsule must always be considered when planning surgery and radiation therapy. Patients with microscopically positive surgical margins are at increased risk of local recurrence (3).

Material and Methods

Case Report and Follow-up

A 69-year-old man was referred for evaluation of a slow growing mass of 6 months duration involving the left side of the face. Clinical examination revealed an obviously visible, well-circumscribed, soft mass, measuring 4-5 cm in diameter, in the region under the left zygoma bone, and ventral the parotid gland. Some time ago a benign tumour has been already removed in the same region. (Fig. 1) Furthermore a Prostate carcinoma was to be mentioned in the medical history of the patient. There was however no history of trauma or infection, and no evidence of cervical lymphadenopathy. There was also no alteration of facial nerve function.
Magnetic resonance imaging revealed a well circumscribed lesion, within the left face half. Also the accomplished Positron Emission Tomography did not bring further realizations concerning the occult distant metastases.
Surgical removal started with an incision within the range of the old operation scar. The tumour was relatively well defined opposite the surrounding tissue with a thin cap. It could be released all-side from the surrounding tissue and removed completely. The thin cap could be also removed intact (Fig. 2, 3, 4, 5).
Since the operation the patient is in our more regularly recall. Post-operative progress was up to now 9 months after the operation uneventful. An offered adjuvant treatment with Radiatio was rejected by the patient. After 4 months CT scan and PET were accomplished. They brought no new findings regarding of a possible latent recurrence.

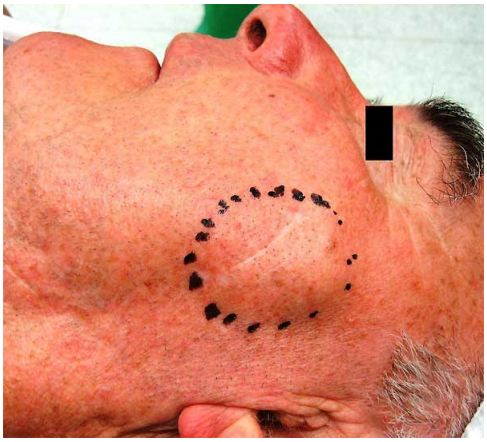

Fig. 1: Preoperative View with the Scar of the former operation.

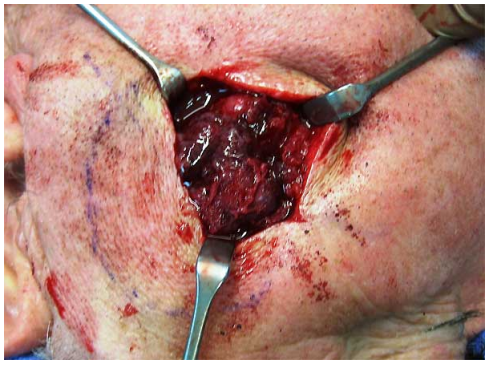

Fig. 2: Intra operative view after incision.

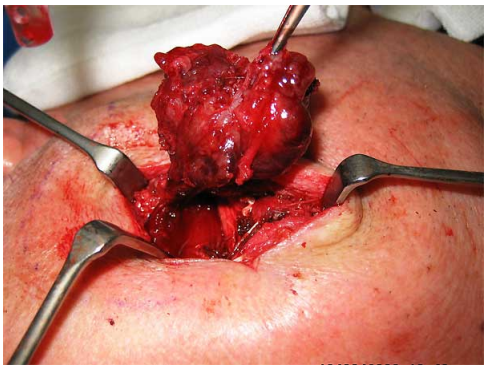

Fig. 3: Intra operative View directly before enucleation of the tumour.

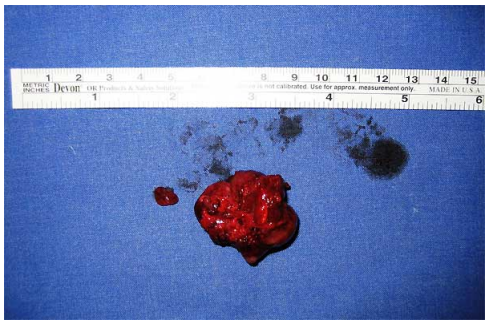

Fig. 4: Macroscopical view of the enucleated tumour.

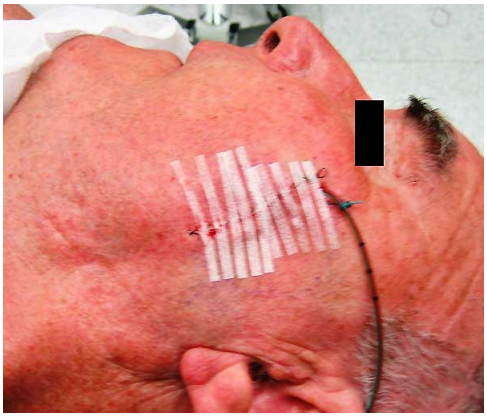

Fig. 5: Post operative view after suturing.

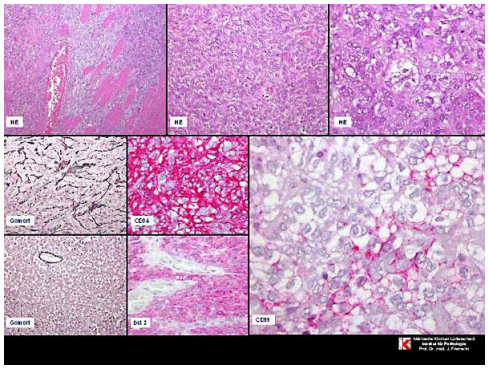

Fig. 6: Microscopic view of the Sarcoma NOS with infiltration. The surrounding muscle tissue, heteromorphic cytologic differentiation, different fibre content and uncharacteristic immune histochemical marker profiles.

Microscopic findings

The histological investigation revealed a tumour that was surrounded at least by a pseudo capsule and was composed of polygonal to spindle cells, which possess badly definable cytoplasm rooms and moderately chromatin rich nuclei. The resection margin were with all-side surrounding healthy tissue.
The tumour cells did not show a characteristic growth pattern and also with the help of supplementing immune-histochemical investigations a definite hitogenetically substantiated histopathological typing could not be accomplished (negative reaction with the anti-bodies against Myoglobin, Melan A, HMB45, MNF116, Lu5, CD117 and positive reaction only with Vimentin-, CD34-, BCL2- and CD99 antibodies.
Due to missing differentiation characteristics the tumour is classified as Sarcoma-NOS and because of missing necrosis and <9 mitosis per 10 HPF it has been classified as malignant grad 2 in the FNCLCC system (Fig. 6).

Conclusions

The overwhelming majority of cancers in the head and neck region (80%-90%) are of epithelial origin. Lymphoma is the next most common head and neck malignancy. Sarcomas of the head and neck are relatively rare (6). Soft tissue sarcomas in adults are uncommon, which partly accounts for some delays in diagnosis and complicates determination of optimal treatment approaches (11). Pathological assessment of soft tissue sarcomas has advanced considerably in the past 10-15 years These advances have led to greater appreciation of the distinct clinicopathological and biological features and natural histories that characterize the approximately 50 histological subtypes of soft tissue sarcoma (12). Our case of Sarcoma NOS will have to be assigned to the group of the Soft tissue sarcoma.
CT and MRI are the primary modalities used for lesion detection and local staging of soft tissue sarcomas PET can overcome limitations in MRI in quantifying biological activity and for whole body staging. Studies suggest that PET is reliable for separating high- from low-grade tumours, in determining biological activity of a tumour, and in predicting tumour necrosis after neoadjuvant therapy (12, 13).
Histologic grade, tumour size, and depth (superficial or deep) are well-established prognostic markers for local control and disease-free survival (13). Soft tissue sarcoma requires a multidisciplinary approach by an experienced team (4). The main therapy for soft tissue sarcomas of the head and neck is en-bloc resection with a margin of normal tissue 6. Some other groups favour a combined treatment approach with surgical extirpation additionally radiotherapy and/or chemotherapy (10).
Some authors minute, that preoperative or adjuvant chemotherapy are not standard practice Preoperative cytostatic therapy can be considered together with radiotherapy in patients with borderline resectable tumours. Adjuvant chemotherapy might improve distant and local control. Its impact on overall survival is still debated. It may be considered in younger patients with large and high-grade tumours (4).
However the therapeutic management of such diverse tumours is complex and depends on the stage, site, and histologic characteristics of the tumour. The most common site of metastasis is the lungs, metastasis generally occurs within two to three years after the completion of therapy (3).

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Poster Faksimile:

Sarcoma-NOS of the Face

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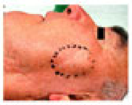

FIG.1: Preoperative View with the Scar of the former operation

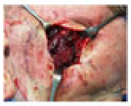

FIG.2: Intra operative view after incision

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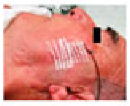

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Conclusion

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However the therapeutic management of such diverse tumours is complex and depends on the stage, site, and histologic characteristics of the tumour. The most common site of metastasis is the lungs, metastasis generally occurs within two to three years after the completion of therapy (3).

Abstract

Soft tissue sarcoma of head & neck are seldom. Particularly our knowledge concerning diagnosis, treatment modalities and pathologic features of the sub type sarcoma- NOS is already very limited.

A case of pleomorphic sarcoma-NOS of the face in adult, originating from the region between the zygoma bone and parotid gland is presented. The entire procedure of the diagnosis and therapy as well as the 11 months follow-up time is discussed.

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