Acta Otorrinolaringol Esp. 2010;61(5):389–391

Acta Otorrinolaringológica Española

www.elsevier.es/otorrino

CASE STUDY

Synovial sarcoma of the infratemporal fossa

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Received March 1, 2009; accepted October 30, 2009

KEYWORDS
Synovial sarcoma;
Infratemporal fossa;
Mesenchymal tumours of head and neck

Abstract
Synovial sarcoma is the fourth most common type of sarcoma. It is usually found in the knee or ankle joints, and is exceptional in the head and neck. Most cases are diagnosed in men between 20 and 40 years of age. Diagnosis is often casual due to the infrequent nature of this tumour and its non-specific clinical and radiological characteristics. Confirmation is therefore based on immunohistochemistry and electron microscopy techniques.
We report a case of biphasic sinovial sarcoma located in the infratemporal fossa treated at our hospital and we make a review of the literature.

PALABRAS CLAVE
Sarcoma sinovial;
Fosa infratemporal;
Tumores mesenquimales de cabeza y cuello

Sarcoma sinovial de fosa infratemporal

Resumen
El sarcoma sinovial representa el cuarto tipo de sarcoma más frecuente, situándose principalmente en las articulaciones de la rodilla y el tobillo, siendo excepcional la afectación de cabeza y cuello. Se da sobre todo en varones entre los 20 y 40 años. El diagnóstico suele ser inesperado por la rareza de la entidad y por las características inespecíficas clínico-radiológicas, basándose, por tanto, en técnicas inmunohistoquímicas y de microscopia electrónica.
Presentamos un caso de sarcoma sinovial bifásico, localizado en la fosa infratemporal, tratado en nuestro servicio, y revisamos la literatura.

Case study

A woman, 72 years old, attending consultation due to pain in the region corresponding to the right maxillary sinus, associated with asthenia, anorexia and weight loss (12 kg) of 6 months' evolution. The previous weeks, she had signs

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of inflammation with redness and swelling on the right cheek.

On examination, there was a slightly painful, oedematous hyperaemic skin area in the region of the right maxillary sinus and discreet inferior palpebral oedema. The rest of the ENT examination was normal.

A CT and MRI were performed, revealing a large 7x3 cm tumour mass in the right infratemporal area with a small necrotic peripheral region. The tumour was located in the infratemporal fossa, compressing the temporal bone and causing osteolysis in it and in the right maxillary sinus. The tumour produced a compressive bulge of the wall on the pterygoid muscles towards the back and downwards. There was no invasion or involvement of the cranial base and the brain, nor perilesional or cervical adenopathies (Figure 1).

Fine needle aspiration (FNA) was performed, with the consequent report of a suspected malignant densely cellular mesenchymal tumour, composed of spindle cells. Subsequently, a Tru-cut biopsy gave us the diagnosis of sinonasal hemangiopericytoma (hemangiopericytoma-like tumour).

Given the lack of final agreement among the different studies, a biopsy was obtained under general anaesthesia, yielding the following microscopic description: tumour with areas of small high-density cells of high mitotic activity along with other areas of extensive cell necrosis; in other areas, cell density decreased, showing a myxoid background and spindle cellularity. All areas presented hypervascularization with ectatic vessel lumens, adopting a hemangiopericytoma-like pattern. The tumour was finally classified as biphasic synovial sarcoma and then confirmed by immunohistochemical techniques, showing a notable vimentin expression in sarcomatoid areas and CD99 expression with membranous pattern.

Over the clinical course of the disease, the patient was admitted to the emergency room because the tumour extruded through the right buccal mucosa, protruding endo-buccally and even becoming visible from the outside due to its large volume (Figure 2).

After a thoracic-abdominal CT extension study showing no lesions suggestive of metastases and a negative bone scan, the patient underwent surgery through a transzygomatic preauricular subtemporal approach. Complete removal of the mass as needed was performed, with microscopic edges tumour-free. Radiotherapy was subsequently applied with adjuvant radical intent on the original tumour location and safety margins. The application of chemotherapy was discarded due to the general condition of the patient.

After 1 year and 2 months of controls without any signs of locoregional recurrence, the patient died due to the presence of multiple pulmonary and pleural metastases.

Discussion

Sarcomas are mesenchymal tumours that represent less than 1% of head and neck tumours. Synovial sarcoma (5%

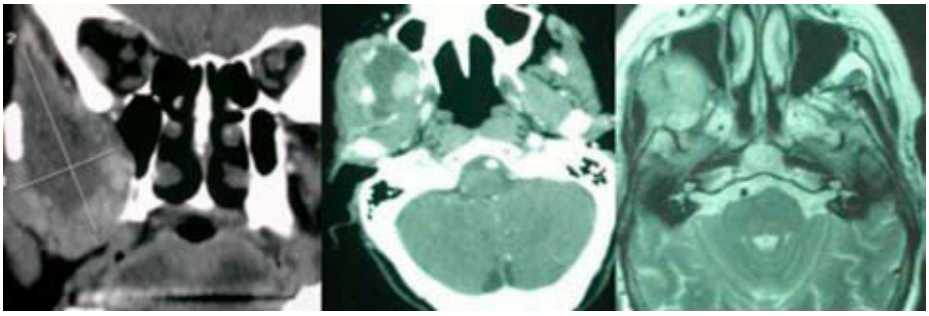

Figure 1 CT and MRI scans showing the tumour location in the infratemporal fossa.

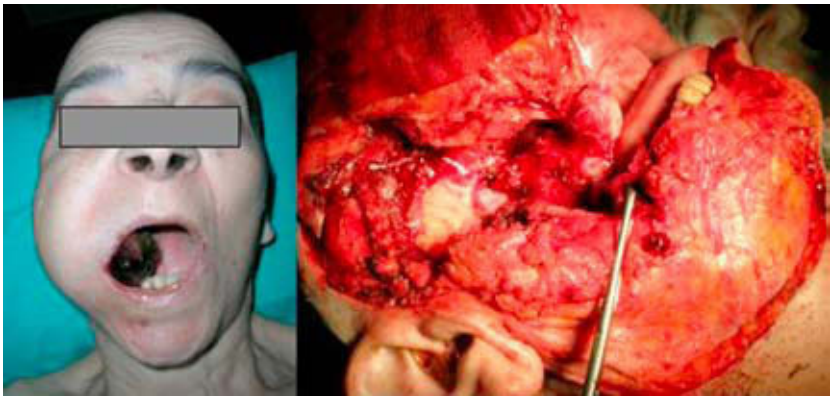

Figure 2 Preoperative and intraoperative images of the tumour.

to 10% of all cases of soft tissue sarcomas) is the fourth in frequency among soft tissue tumours after liposarcoma, malignant fibrous histiocytoma and rhabdomyosarcoma.[1]

A cervicofacial location for a synovial sarcoma is extremely rare, with this being the fourth case reported in Anglo-Saxon literature to our knowledge.[2] Its typical locations are the knees and ankles, often near the joints, and it occurs more often in adolescents and young adults.

Its growth is slow and the first manifestation is usually a painless juxtaarticular mass. Evidence of regional lymph node involvement strongly supports the diagnosis.

Various imaging studies report a soft tissue mass with inherent calcifications in 30% of cases, as well as compressions and bone invasions.[3-5]

Since these sarcomas do not originate in the synovium and many of them occur outside the joint, they do not have any relation with the synovium, either ultrastructurally or immunohistochemically. It has been suggested that synovial sarcoma arise from pluripotent mesenchymal cells.

There are two forms[4,6]:

- *Biphasic:* they are characteristic and distinctive, composed of epithelial cells forming solid cords surrounded by spindle cells that create a glandular or adenoid pattern. They are the most common.
- *Monophasic:* they can be composed of epithelial or spindle cells. The fibrous type may resemble fibrosarcoma. The monophasic epithelial type is similar to metastatic carcinoma and is very rare.

The poorly differentiated type contains cells similar to an intermediate form, with both epithelial and spindle cells. This type is more aggressive and has an increased ability to metastasize.

The anatomopathological differential diagnosis is primarily established with malignant hemangiopericytoma, fibrosarcoma and squamous cell and spindle cell carcinomas.[7]

When doubts remain regarding the anatomopathological diagnosis, it is possible to resort to electron microscopy and immunohistochemistry (based on the keratin and vimentin test). Most have a reciprocal translocation between chromosomes X and 18 (p11.2; q11.2) with involvement of SYT-SSX genes, which is very helpful for diagnosis.[7]

The most important factors determining prognosis are grade and size, age and gender (better for younger cases and for females), calcifications (better prognosis), and anatomical location (determines the type of treatment).

The recurrence rate is high and lymphatic metastases to bones and lungs are characteristic. The 5-year survival is 25% to 55%.[8]

Surgical treatment is preferred, with wide safety margins.[9] Radiotherapy is associated with high-grade tumours or when they are close to important neurovascular structures. If tumours are greater than 8cm in diameter, administration of chemotherapy and radiotherapy is considered.[8]

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