

Exposition summary during meeting

Introducción:

Synovial sarcoma constitutes 3-10% of all sarcomas.

Men are affected more frequently than women with percentages of 2:1 or 3:2

Despite their name, these tumors do not originate from synovial tissue, but rather from pluripotential mesenchymal cells located both near and far from the joint surfaces.

This type of tumor rarely appears in synovial membranes; only 10% affect joint spaces.

Between 3 and 10% of synovial sarcomas appear in the head and neck.

The first case in this location was described in 1954 by Jernstrom, and it was a hypopharyngeal tumor.

Patients with synovial sarcomas of the head and neck have between 25 to 30 years, with a range of 7 to 63 years.

In the head and neck, the hypopharynx is the most frequently affected area.

Other locations, in the head and neck, described in the literature are the parapharyngeal space, oral cavity, tongue, parotid gland, temporomandibular joint, nasosinusal region, nasopharynx, oropharynx, etc. Although any area can be affected.

The size of these tumors can vary at the time of diagnosis between 1 and 12 cm.

They are generally solitary and firm tumors, with smooth and shiny cut surfaces, well circumscribed, spherical or micronodular, with pseudocapsule and a shiny grayish-yellow color.^{1,6} There is often focal calcification and it may have cystic or hemorrhagic foci

Although most metastases are through the blood, 10 to 20% of patients have lymphatic metastases.

Clinically, synovial sarcomas of the head and neck usually appear as a painless, slow-growing mass. Only in 20% of cases does pain appear

Most synovial sarcomas can clinically resemble a fibrosarcoma or rhabdomyosarcoma, behaving like a mass that infiltrates soft tissues.

Both CT and Nuclear Magnetic Resonance (MRI) describe the lesion, in the head and neck, generally well delimited, solid, and sometimes with a hemorrhagic or cystic component, as well as calcifications. Both CT and MR images should include the entire neck, because 12.5% of patients with head and neck synovial sarcoma have regional lymph node involvement. In addition, a chest CT should be performed to detect the existence of pulmonary metastases, which are the cause of death in a significant number of patients. Histologically, synovial sarcomas are characterized by the presence of a biphasic pattern consisting of a stromal background composed of spindle cells with an appearance similar to fibroblasts, firmly compacted, and with a relatively monotonous appearance. On this background the epithelial component is arranged, usually as glandular formations, compact nests, or cleft spaces. The spindle cell areas are reminiscent of fibrosarcoma histology. Synovial sarcomas can be highly vascular and may be histologically similar to a hemangiopericytoma. The relative proportion between spindle cells and epithelial cells varies from case to case and within the same lesion.

There are monophasic forms of synovial sarcoma, which present only a spindle cell or epithelial histological pattern. The metastases of the biphasic forms can have a monophasic or biphasic pattern, while the metastases of the monophasic forms are always monophasic. Cytogenetic studies of synovial sarcoma have been performed, demonstrating a specific reciprocal translocation on chromosome X and 18 in more than 90% of cases.

The treatment for synovial sarcoma is surgical. A resection with safety margins must be performed, often including the adjacent tissues, such as the peritumoral musculature.

Routine radical neck dissection is not considered necessary if there are no adenopathies

The recurrence rate of synovial sarcomas in the head and neck, after local excision, varies

between 21% and 56%, so postoperative radiotherapy is indicated. Adjuvant chemotherapy can reduce or delay the appearance of distant metastases.

Survival of synovial sarcoma of the head and neck, at 5 years is 36% to 54%, and at 10 years it is 20%, due to the appearance of metastases. The prognosis of synovial sarcoma is determined by a series of factors: histologically, calcifying synovial sarcomas have a better prognosis than the rest according to their largest diameter and extension, tumors smaller than 5 cm and that do not invade adjacent structures have a better prognosis; depending on the location, the head and neck have a better prognosis than the extremities

clinical case

A 32-year-old male patient, who came to the consultation for a tumor in the left cheek that took up the entire cheek of approximately 2 years of evolution that began as a cyst according to the non-painful patient, with no medical or surgical history.

On physical examination, it has a hard-elastic consistency with a size of approximately 5 cm to 4cm is not attached to deep planes and with free neck examination without cervical adenopathies.

Presents an ultrasound that reports a delimited solid mass that is adhered to the muscular wall with no other characteristics.

The CT shows a solid mass well delimited with the muscular planes and without invasion of the maxillary sinus, the bony walls being free of lesions.

The differential diagnosis includes carcinosarcoma for the classic type, adenocarcinoma metastasis for the epithelial monophasic variant, and spindle cell carcinoma and sarcomas such as fibrosarcoma, leiomyosarcoma, malignant tumor of the peripheral nerve sheath (neurofibrosarcoma) and hemangiopericytoma for the fibrous monophasic variant.

Surgical treatment with preservation of the facial nerve was proposed to maintain mimic muscle function. Adjuvant radiotherapy was recommended, which was performed.

Currently with 2 year and 6 months of follow-up, without evidence of disease.

Conclusions

Synovial sarcomas are extremely rare. This can lead to misdiagnosis if there is no accurate histopathology and the possibility of support with immunohistochemical techniques. We believe that, as far as possible, surgical resection should be aggressive, accompanied or not by lymph node dissection, as the case may be, and complemented with radiotherapy and according to protocols.