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Orthopedic Surgery

Synovialosarcoma of the Right Ankle about a Case

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Abstract **Case Report**

Synovial sarcoma is a mesenchymatous tumor, despite its name; synovial sarcoma does not come from synovial tissue. We report the case of a 54-year-old woman presenting since 2 years a synovial sarcoma of the right ankle Diagnosis has been confirmed by immunohistochemistry. Treatment was based on surgery followed by radiotherapy. This case confirms the efficiency of this combined treatment.

Key words: Synovial sarcoma - Ankle – immunohistochemistry - surgery – radiotherapy.

INTRODUCTION

Synovialosarcoma is a mesenchymal tumor thatusually reaches the soft parts of the lower limbs, most often near tendon insertions orjoint capsules. Much more rarely, he meetsin the region of the head and neck [1, 2].Despite its name, synovial sarcoma does not developfrom the synovial tissue. As for most sarcomassoft tissues, the tissue origin of synovial sarcoma isstill unknown and the risk factors are not clearlyestablished; under these conditions, no screening programcan not be done or recommended [3]. Basic treatment includes surgical excisionbroadly whenever possible, supplemented by aexternal radiotherapy.

Chemotherapy keeps indicationsrestricted, it is mainly used in pediatrics. This case comesconfirm the effectiveness of combined surgical exeresisradiotherapy in adults.

OBSERVATION

A 54-year-old patient with no pathological history notables is place consult for ankle mass evolutionary right for 2 years, painless, gradually increasing of size and are not volume variation at the time of meals. Clinical examination revealed a painless, indurated, fixed mass with no ankle stiffness and no sensory motor disturbances (Fig. 1). The standard x-ray was unremarkable. ultrasound of the soft tissues had objectified a heterogeneous tissue lesion process contains multiple calcifications link a central vascularization look at a tumoral external malleol of the soft parts of the right ankle 50 x 45 x 65 mm in size (Fig 2) tke MRI: Tumor process in relation to the iso-intense right external malleolus in hyper-intense T1 in heterogeneous T2 after injection of igadolinum showing zones of necrosis. The lesion is well-limited, with infiltration of the subcutaneous tissue measuring 50 x 40 x 65 mm, and the extensor tendons and the anterior tibial pedicle (fig 3). In front of this mass whose aspect and the evolution have a malignancy, a biopsy was performed. Histopathological examination showed that it was a synovial sarcoma monophasic spindle cell grade II. In immunohistochemistry, epithelial cells were labeled with AE1/AE3 cytokeratins and membrane epithelial antigen (EMA). To remove any doubt about the histological aspect, the gene SYT-SSX1 fusion was sought and found by hybridizationin situ in fluorescence. The extension assessment, including a chest x-ray, an abdominopelvic ultrasound and a scintigraphy bone, no metastasis was found surgical treatment consists of a resection in monobloc of the tumor with an intraoperative extemporaneous exam to ensure the limits of excision (fig 4). The boundaries of the resection were in healthy tissue and there was no lymph node metastasis. They had thus found themselves in group I according to the IRS gradation (Intergroup study on rhabdomyosarcoma). She then benefited of Adjuvant External Radiotherapy at a Rate of 70 Gy Fractional over 6 weeks. Postoperative controls at 6 months and 1 not found as Clinical and Imaging recidivism.

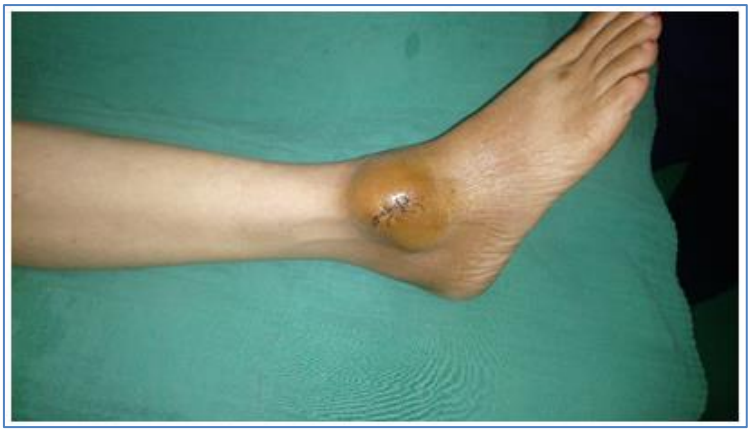

Fig-1: Preoperative image

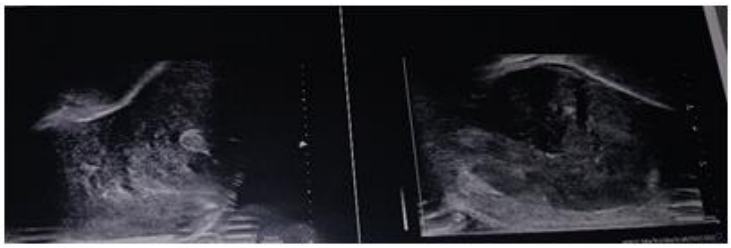

Fig-2: Heterogeneous tissue lesion process contains multiple calcifications link a central vascularization look at a tumoral external malleol of the soft parts of the right ankle 50 x 45 x 65 mm in size

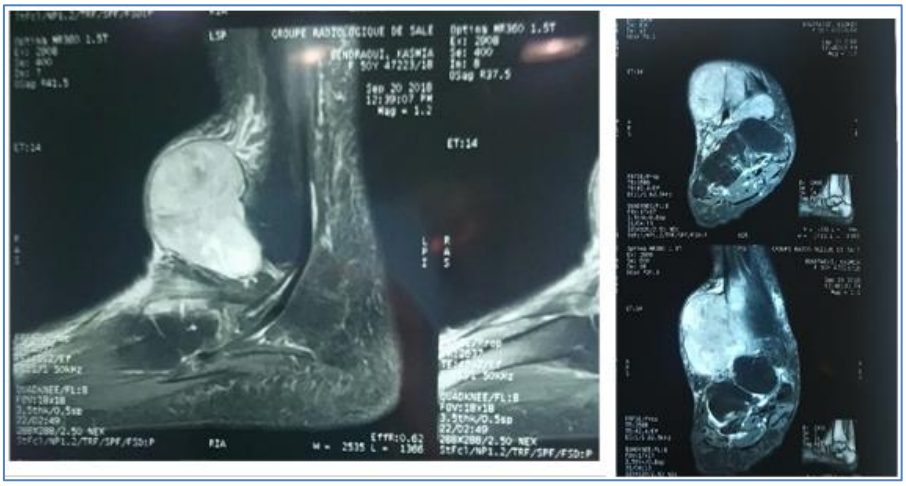

Fig-3: Tumor process in relation to the iso-intense right external malleolus in hyper-intense T1 in heterogeneous T2 after injection of igadolinum showing zones of necrosis. the lesion is well-limited, with infiltration of the subcutaneous tissue measuring 50 x 40 x 65 mm, and the extensor tendons and the anterior tibial pedicle

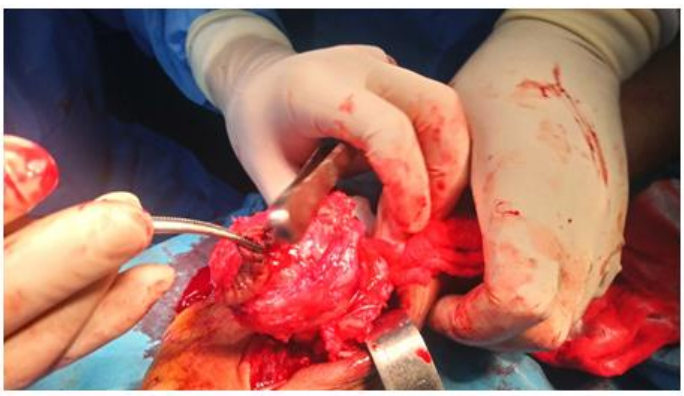

Fig-4: Resection in monobloc of the tumor with an intraoperative extemporaneous exam to ensure the limits of excision

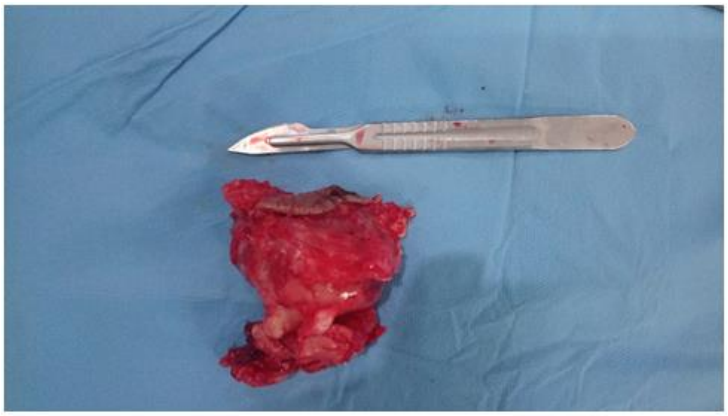

Fig-5: The tumor after exeresis

DISSCUSSION

Represents 6 to 10% of soft tissue sarcomas (1,000 / year in France), the fourth largest soft tissue sarcoma after liposarcoma, rhabdomyosarcoma and malignant fibrous histiocytoma. In fact, it is not derived from synovial tissue but consists of dedifferentiated mesenchymal cells. Concerns the young adult (15 to 45 years), of rather masculine sex. The tumor usually develops in the vicinity of tendons, bursae or joint capsules, and particularly in the lower limbs (preferential location in the knee [popliteal cavity], represents the most common malignant tumor of the foot and ankle between 6 and 45 years of age). Due to slow growth (2 to 4 years), a moderate size (<5cm) and a sometimes homogeneous and limited appearance, the tumor can be wrongly recognized as benign. The prognosis is aggravated by a high risk of local recurrences (50%) and metastases (40%) of which 16 to 25% are discovered in the initial assessment (lungs +++, ganglia, bones ...). Circumstances of discovery sometimes report a traumatic event that bleeds the mass.

Histology, immunohistochemistry and cytogenetics

On macroscopic examination, the tumor appears to be well limited, multilobulated, with 3 characteristic features: necrosis, haemorrhagic and liquid areas.We describe 3 types:Biphasic (20 to 30%), consisting of mesenchymal cells and epithelioids in variable proportion (up to an appearance of "glandular" tumor of the adenocarcinoma type);Monophasic (50 to 60%), consisting of single mesenchymal cells with a hemangioperiodtoid vascular component, a collagenous stroma and sometimes microcalcifications or areas of bone metaplasia (differential diagnosis of nerve sheath tumors ...);Little differentiated (15 to 25%), epithelioid morphology with significant mitotic activity and ranges of geographic necrosis, close to round cell tumors such as Ewing's sarcoma. The bi and monophasic forms are usually of intermediate grade (2/3) while the poorly differentiated forms are of high grade of malignancy (3/3).Immunohistochemistry thanks to its markers (keratin ...) and the detection of specific cytogenetic abnormalities (translocation T (X; 18); SYT-SSX) are of great help for the diagnosis. Certainty in case of doubt or small pathological specimen.

Imageries

On standard radiography, almost 50% of tumors are not visible because of their small size and localization (peripheral or axial). The mass syndrome can sometimes be observed, sometimes with microcalifications (30% cases) sometimes extensive, at the periphery of the tumor (differential diagnosis with villo-nodular synovitis), and in almost a quarter of the cases, bone abnormalities at contact (thickening or erosion of the cortex, micro geodic osteolysis).CT shows a heterogeneous mass of soft tissues, often multilobulated, of density less than or equal to the muscle, with necrosis or haematological plaques. The tumor limits are regular in 50% of cases and the enhancement after injection of iodinated contrast medium appears heterogeneous (> 90%). Calcifications are found in 30 to 40% of primary tumors but also in metastases (lung).MRI remains, as for all tumors of soft tissues, the reference examination of the locoregional extension assessment.

Treatment: It is based on surgical excision, the difficulty and quality of its margins depends essentially on the situation of the tumor. A local excision carrying the tumor, its pseudo-capsule and the peri-tumoral cap is conventionally carried out. Adjuvant treatments can be discussed in case of residual or metastatic tumor (chemotherapy) or in case of marginal excision (radiotherapy). Prognosis: the prognosis is reserved because of a high power of recurrences (20 to 40%) and of metastatic evolution (40 to 70%), in particular pulmonary (80%), ganglionic or more rarely bone.

CONCLUSION

Synovial sarcoma is an aggressive tumor witha high metastatic risk. The early diagnosis of thisrare entity is essential because it avoids mutilating surgeryand ensures a better prognosis for the patient.

RÉFÉRENCES

1. Russell WO, Cohen J, Enzinger F, Hajdu SI, Heise H, Martin RG, Meissner W, Miller WT, Schmitz RL, Suit HD. A clinical and pathological staging system for soft tissue sarcomas. Cancer. 1977 Oct;40(4):1562-70.
2. JULLIARD A. Tumeurs articulaires. La presse médicale. 1961;69:8.
3. Hampole MK, Jackson BA. Analysis of 25 cases of malignant synovioma. Canadian Medical Association Journal. 1968 Nov 30;99(21):1025.
4. Hadju SI, Shiu M-H, Fortner J-G. Tendinosynovial sarcoma, a clinic pathological study of 136 cases cancer. 1977, 39: 1201-1217.
5. Rajpal S, Moore RH, Karakousis CP. Synovial sarcoma. A review of treatment and survival in 52 patients. New York state journal of medicine. 1984 Jan;84(1):17.
6. Brodsky J, Hajdus S, Casper E, Brennan M. Tendinosynovialsarcoma clinic pathologic features, treatment and prognosis. Cancer. 1992, 70: 484-489.
7. Kampe CE, Rosen G, Eilber F, Eckardt J, Lowenbraun S, Foster J, Forscher C, Selch M. Synovial sarcoma. A study of intensive chemotherapy in 14 patients with localized disease. Cancer. 1993 Oct 1;72(7):2161-9.
8. Varela-Duran J, Enzinger FM. Calcifying synovial sarcoma. Cancer. 1982 Jul 15;50(2):345-52.
9. Brodsky J, Hajdus S, Casper E, Brennan M. Tendinosynovialsarcoma clinic pathologic features, treatment and prognosis. Cancer. 1992, 70, 484-489.
10. Varela- Dubran J, Enzinger F-M. Calcifying synovial sarcoma cancer. 1982, 50: 345-352. 138.