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Case Report

Synovialosarcoma of the hand About 03 cases.

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ABSTRACT:
Synovialosarcoma is a rare tumor. It accounts for 8-10% of malignant soft-tissue tumors and around 10% of malignant soft-tissue tumors in the hand (2,4,8,9,11). Its prognosis is the most severe of all soft-tissue sarcomas, with around 50% recurrence within two years and an estimated survival of 50% at 5 years, and 25% at 10 years (1,9). Conservative surgery of the hand and wrist is indicated according to tumor extension and affected compartment. **Material and methods**: The authors report three cases of synovialosarcoma of the hand. The patients were 28, 32 and 37 years old. Two patients were female. Diagnosis was late in all cases. Extension work-up included standard chest X-ray, MRI and thoraco-abdomino-pelvic CT scan. In all three cases, the clinical presentation was different:

Keywords: Synovialosarcoma, malignant soft-tissue tumors, thoraco-abdomino-pelvic CT scan

CASE REPORT:
A: The first patient, Miss A, aged 28, presented with a 2cm swelling on the palmar surface of the right hand (right-handed patient), which had appeared 6 months previously. The CT scan showed a tumor opposite the palmar surface of the diaphysis of the 4th metacarpal (fig 1). Extension workup revealed no other localization. A palmar biopsy showed a monophasic synovialosarcoma with an R1 resection margin, i.e. microscopically incomplete. As the palmar resection was incomplete, a dorsal amputation was performed, removing the 4th metacarpal bone (Fig. 2), followed by reconstruction of the 4th radius with a cortico-cancellous tibial bone graft (Fig. 3). Adjuvant chemotherapy was performed. Local and pulmonary metastases appeared one year later, resulting in the patient's death.

Fig1: Horizontal section through the diaphysis of the metacarpals.

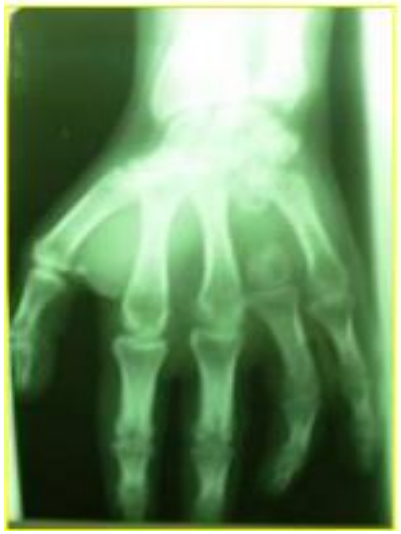

Fig 2: Rx after completion of dorsal resection

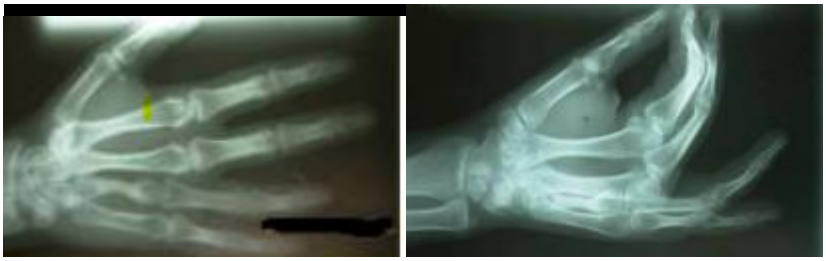

Fig3: Rx at 6 months after tibial cancellous bone graft reconstruction

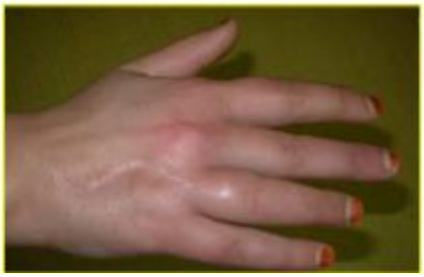

Fig4: Aspect of the dorsal surface

B- The second patient, Mr. E, a 32-year-old man, presented 6 months after the onset of swelling on the palmar surface of the 4th finger of the right hand, misdiagnosed as villonodular tenosynovitis after cytopuncture. The patient initially underwent no surgery. After 1 year, and in view of the onset of pain, an MRI scan was carried out, which revealed a tumor that was a priori aggressive and spread opposite the palmar surface of the PPI of the 4th finger (Fig6). Given the aggressive nature of the tumor on MRI, the patient underwent surgery. Tumor excision was performed via the palmar approach, preserving the collateral pedicles, with resection of the A3 pulley and conservation of the 2 flexor tendons. The anatomopathological diagnosis of monophasic synovialosarcoma was made with wide R0 resection margins, i.e. margins of healthy tissue. Adjuvant chemotherapy was performed.

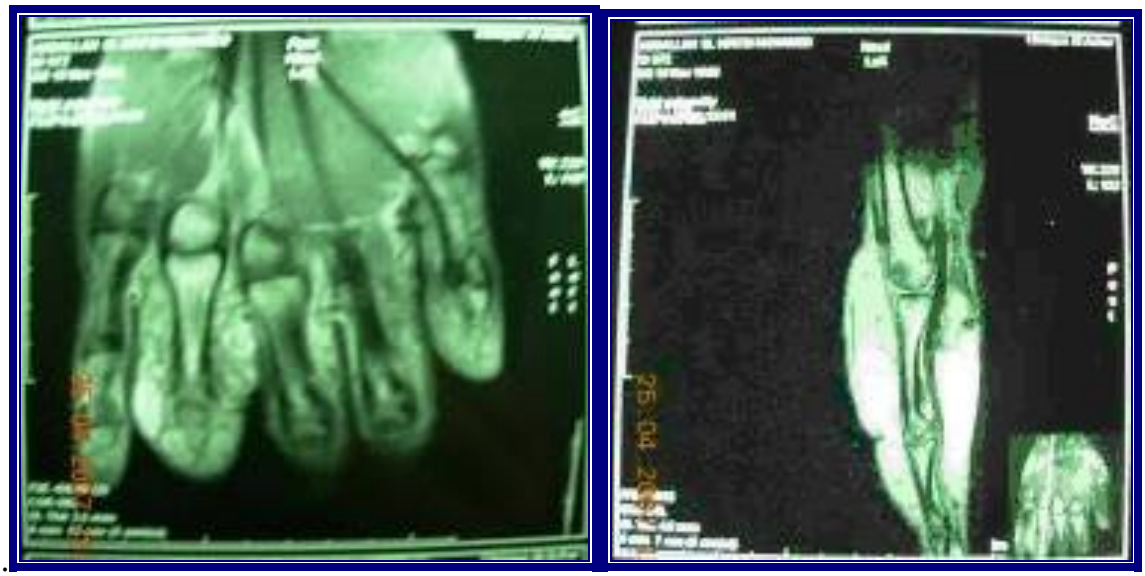

Fig6 : MRI

The functional and overall result was satisfactory, with full mobility and no local recurrence.

C- The third patient, Mrs. K, aged 37, presented 7 months after the appearance of a swelling opposite the palmar surface of the metacarpophalangeal joint of the thumb, extending into the first interosseous space (Fig7) of the right hand. A palmar biopsy was performed, preserving the pedicles, and the anatomopathological result was in favor of a biphasic synovialosarcoma with healthy R0 resection margins (Fig8) Adjuvant radiotherapy was performed.

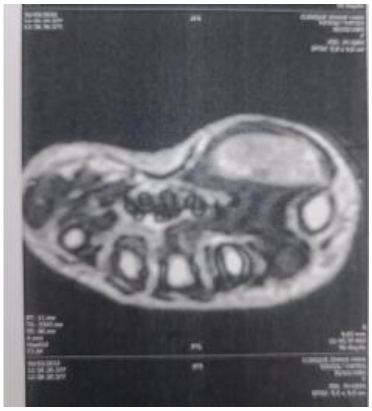

Fig7 :MRI

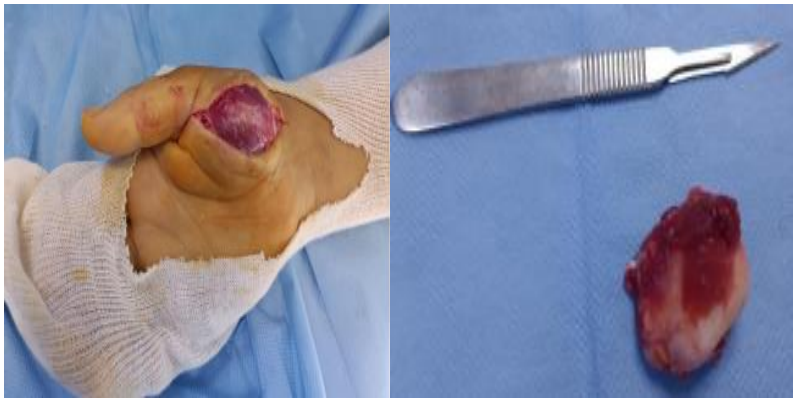

Fig 8: Peri-operative aspect

The functional and overall result was satisfactory, with full mobility and no local recurrence.

3. Results (Summary table of the 3 cases):

Age	28	32	37
Sex	F	H	F
Delay symptoms/diagnosis	6 months	6 months	7 months
Location	palmar surface 4th	palmar side 4°finger	1° comissure

	metacarpal		
Anatomopathology	monophasic	monophasic	biphasic
of synovialosarcoma			
Follow-up	36 months old	12 months	36 months old
Resection	Exeresis biopsy	Cytopsy prior to biopsy Biopsy Exeresis	Biopsy and excision
Recurrences	Local and pulmonary	No recurrence	No recurrence
Neo-adjuvant	lung	no	no
Adjuvant	no	Yes	no
Adjuvant chemotherapy	Yes	no	yes

DISCUSSION:

Epidemiology:

Synovialosarcomas are rare tumors, and published series on this tumor in the wrist and hand are few in number (8,9,11,13,23). Nevertheless, this tumor is considered by most authors to be the most common soft-tissue sarcoma of the hand and wrist (2,4,24). It most often affects young people, between the ages of 15 and 40 (4,11,18). Diagnosis may take several years (18,28) and is often unrecognized (2,9,12,13,29).

Anatomopathology:

The anatomopathological definition of synovialosarcoma corresponds to: a malignant spindle-cell mesenchymal tumor with variable epithelial differentiation, including glandular structures, and which has a chromosomal translocation t(x,18) (p11q12). (9,16)

- Four morphological forms have been described (9,12,13,16):
- biphasic;
- monophasic with spindle cells;
- monophasic with epitheloid cells;
- poorly differentiated with round cells.

The monophasic form is the most frequent (9,16).

Diagnosis is morphological, aided by immunohistochemistry: cells express markers such as EMA, AE/1, AE/3, Vimentin and CD99. Currently, fusion transcript research shows a chromosomal translocation t(x, 18) (p11,q12) specific to synovialosarcoma (9,16).

Prognosis:

Local recurrence within two to three years of diagnosis is common (50%), and lymph node or distant metastases (lung, bone) occur in 40% of cases. In various series, survival at 5 years is 50% and 25% at 10 years (1,9).

Good prognostic factors are :

1. young age.
2. tumor diameter: tumor size is a major prognostic factor. Tumors < 5cm have a better prognosis (3,4,9,12,16).
3. less than 10 mitoses per 10 fields, corresponding to tumor aggressiveness (Grade)
(4,16).
4. absence of intra-tumoral necrosis
5. macroscopically and microscopically complete resection: authors insist on the importance of carcinological resection with healthy resection margins (type R0 according to UICC) of the tumor to avoid recurrence (2,4,17,18,26).
6. The biphasic morphological form has a better prognosis than the spindle-cell monophasic form, which has a better prognosis than the epithelioid-cell monophasic form, while the poorly differentiated round-cell (rhabdoid) form has a worse prognosis (4,9,16).

TREATMENT:

Surgically, extended resection is often difficult, due to the complexity of hand anatomy(5,30,31,32) and the technical difficulties of hand reconstruction (flaps, tendon transfers, tendon and nerve bone grafts). From a medical standpoint, synovialosarcomas are not very chemosensitive; however, in the case of large, extensive tumors, neoadjuvant chemotherapy may be indicated to limit the need for resection and thus improve functional sequelae (7,14,15,17,22). With regard to adjuvant chemotherapy, the "standard" management that emerges

from the various series seems to suggest adjuvant chemotherapy only for patients with high-grade tumors measuring more than 5 cm; it would reduce the incidence of local recurrence and metastases, thus improving recurrence-free survival and overall survival (7,14). As far as radiotherapy is concerned, numerous retrospective studies and two randomized trials (one of external radiotherapy and one of brachytherapy) have shown that adjuvant radiotherapy after complete surgery reduces the risk of local relapse in limb sarcomas. Thus, the combination of surgery and post- or pre-operative radiotherapy is part of standard treatment, with a local relapse rate of less than 25% and, above all, a very low amputation rate (7, 9, 10,17,25).

CONCLUSION:

The treatment of synovialosarcoma is multidisciplinary. Its management is based on evolving concepts (5,19). The contribution of adjuvant treatments, in particular radiotherapy and chemotherapy, means that surgery can be directed towards more conservative rather than radical procedures, in order to preserve function, avoid recurrence and improve survival (20,28,30,33).
Based on the literature and our own experience, we recommend the following course of treatment:

Good Prognosis Tumor:

-Wide resection,
± Multi-tissue reconstruction
+ Radiotherapy

Tumor with poor prognosis:

Neoadjuvant chemotherapy.
-Wide resection ,
± Multi-tissue reconstruction
+ Radiotherapy.

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