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Synovial sarcoma of the limbs (about 20 cases)

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ABSTRACT

Evaluate retrospectively, in a homogeneous series of patients with synovial sarcoma of the limbs, the different steps of the management of this disease, the diagnosis and the therapeutic difficulties. This is a retrospective study of 20 cases of synovial sarcoma of the limbs treated in orthopedic surgery department at Ibn Sina university hospital and in the National Institute of Oncology Rabat, between January 2000 and December 2012. 20 cases of synovial sarcoma of the limbs were collected of which 12 were localized in the lower limb and 8 localized in the upper limb. The mean age was 45 years with a male predominance. The patients consulted after an average period of 18 months and tumoral syndrome was the constant reason for consultation. All the patients were treated surgically, except the 4 cases with lung metastases. After an average follow-up of 48 months, 10 patients are alive, 5 died and 5 are lost of view. Because of their rarity, the diversity of their histological types and their localization, synovial sarcoma presents problems at every stage of their management. Identification of specific chromosomal translocations is a great advantage in their diagnosis. Multidisciplinary approach is mandatory in their management.

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Introduction

The synovial sarcoma defined as primitive mesenchymal malignant tumor reproducing histological features of synovial tissue [1]. They represent 5 to 10% of all malignant tumors of the soft tissues. They present a problem in their diagnosis, their differential diagnosis and in their therapeutic care that requires a multidisciplinary collaboration.

Through a retrospective study of 20 cases of synovial sarcoma of the limb collected at Orthopedic Surgery department of Ibn Sina University Hospital between January 2000 and December 2012, we will discuss the various stages of the treatment of this tumor, show the diagnostic problems and finally discuss our results with a review of the literature.

Materials and methods

Our series includes 20 cases of synovial sarcoma of the limb. The average age was 45 years. The sex distribution showed a slight male predominance with 12 men for 8 women. The lower limb was the most frequent location with 12 cases versus 8 cases in the upper limb. Thigh and knee was the most common location.

The constant reason for consultation was the appearance of a swelling of the limb with gradual increase in size. The average duration of evolution before the first consultation was 18 months with extremes of one month to 42 months. The deterioration of the general condition has been reported in 3 cases. On physical examination, the most encountered physical sign was a painful soft single mass increasing in volume. All patients underwent standard radiography that showed opacity of soft tissues in 13 cases and calcifications in 10 cases. The ultrasonography of the soft parts was realized in 8 cases but the diagnosis of synovial sarcoma wasn't evocated. CT scan was performed in 14 cases and showed in all cases a lesion with dual component (fluid and tissue process) and osteolysis of the cortex. The MRI with gadolinium injection was performed in 12 of our patients, which allowed distinguishing the tumor from peri-lesional edema. It also specified the heterogeneous nature of the tumor and identified clearly the areas of necrosis. Angiography was performed in 6 cases and a Doppler ultrasound in 5 cases to explore the vascular axis, but no vascular invasion was found in any case. As part of a general assessment of expansion, thoraco-abdominal CT was done for all patients and showed lung metastases in 4 cases. The bone scans for all patients didn't show bone metastases. A surgical biopsy was performed in all cases. Histological study showed biphasic synovial sarcoma in 12 cases and monophasic in 8 cases. All patients underwent a conservative surgical treatment or amputation, except the 4 cases with lung metastases.

- Conservative treatment was used in 10 of our patients.
- Wide excision was performed in 6 cases and one patient was operated again for complementary resection.
- Radical resection was performed in 4 cases immediately removing adjacent structures of the tumor.
- Marginal excision wasn't performed in any case.
- Amputation was performed in 6 cases.
- 10 patients underwent additional radiotherapy.

Results

Our results were evaluated at an average of 48 months. 6 patients are alive without local or metastatic recurrence, 5 died and 5 are lost of view. We noted a local recurrence in 6 cases. The average time of appearance of these recurrences were 5 years. A second surgical excision was made for these recurrences. 4 patients presented lung metastasis.

Discussion:

The synovial sarcoma is a rare malignant tumor. It represents 5-10% of soft tissue sarcomas, with an incidence of approximately 2.75 \ 100000 \ year [2], this tumor can occur in all ages, but it seems with a high incidence among young adults males, as described in most series [3, 4, 5]. The synovial sarcoma can develop in any site where a synovial membrane, bursa, tendon, tendon sheath or a facial fascia exist. Most publications emphasize the frequency of locations in lower limb and especially the thigh and knee. As in the series of Gerner

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and al. among 34 cases of synovial sarcoma, 26 tumors involved the lower limb in 76% of cases [6].

Clinical latency is important because the tumor has an insidious and asymptomatic growth. The time of diagnosis is often long, the swelling is revealing in more than half of the cases [5]. The pain can be an early warning. On physical examination, the synovial sarcoma occurs most often as a single mass, soft slowly increasing in volume and becomes progressively painful with limits that are often clear and keep a mobility on the superficial levels, rarely on the deep ones [3, 7]. Conventional radiography remains essential during the initial assessment. It reveals an opacity associated or not to calcifications and eliminates a pseudo mass (exostosis, hypertrophic callus) or bone lesion extension into the soft tissues [8]. Ultrasound can differentiate between liquid or solid nature of the lesion, to clarify its superficial or deep location and its relationship to adjacent structures [9, 10]. MRI is the essential means of exploration. It permits to establish the loco regional extension and the follow up after treatment; however, this technique can identify morphological and signal characters towards the diagnosis of synovial sarcoma and to assess the prognosis.

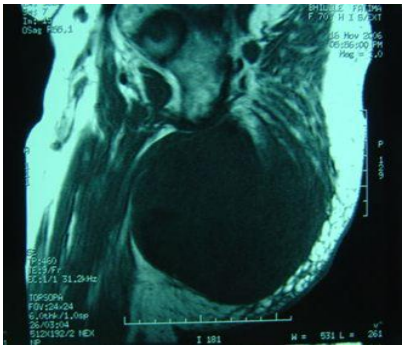

Figure 1: MRI of synovial sarcoma of buttock

Figure 2: Biopsy of synovial sarcoma of buttock

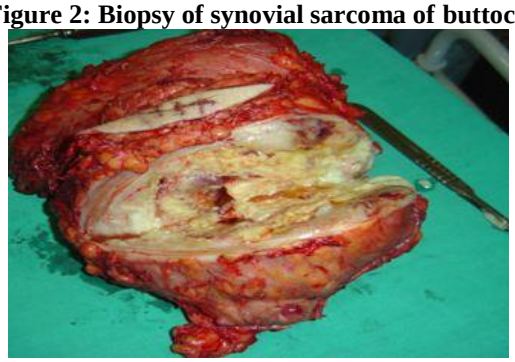

Figure 3: Resection of synovial sarcoma of thigh

It is also the most efficient way to detect local recurrences or residues after incomplete resection [11, 12, 13]. The biopsy should be done before any treatment attempt, the purpose of this biopsy is to get the diagnosis of certainty, a classification of the tumor and possibly assess the histological grade. All these elements allow an adequate therapeutic decision. Surgical biopsy is the gold standard, which provides good exposure of the tumor tissue and permits to collect a representative fragment of the tumor. It relates to all tumors whose diameter exceeds 5 cm and all the deep tumors regardless of their size [14, 15, 16]. At the histological level, we could classify as biphasic, monophasic, or calcifying synovial sarcoma. Immunohistochemistry currently plays an important role in the diagnosis and classification of soft tissue sarcomas [17]. The cytogenetic and molecular biology confirm the diagnosis of synovial sarcomas allowing detection of a translocation between chromosome X and chromosome 18 [18, 19, 20, 21]. The principles of surgery of synovial sarcomas join those of any soft part sarcoma. It is important to know the local expansion mode of synovial sarcomas of the limbs to understand the principles that govern the surgical excision [22, 23]. These sarcomas of soft tissue grow by centrifugal pushes involving in periphery of the tumor a compression of adjacent normal tissue , from where the appearance of a peripheral pseudo capsule. This pseudo capsule is often vague and very variable thickness thus does not represent a reliable and satisfactory limit to carry out a complete excision with a safety margin [16.23, 24.25]. The resection margins are then evaluated according to the classification of MUSCOLOSKELETAL TUMOR SOCIETY and are classified as radical, wide, marginal and intra lesional. The surgical resection is common to all soft tissue tumors. The anatomical surgical confrontation defines the quality of resection according to the criteria of ULCC (R classification ULCC in the fourth edition TNM) [26].

R0: healthy margin microscopic
R1: existence of a microscopic residue
R2: existence of a macroscopic residue

Surgical resection followed by complementary radiotherapy is the standard treatment of loco regional soft tissue sarcomas localized to the members and operable immediately. For Some authors [27, 28, 29], the abstention of adjuvant radiotherapy is possible for superficial tumors, smaller than 5 cm and low histological grade, for which a surgery with healthy limits of section could be realized [30]. The benefit of adjuvant radiotherapy in terms of local control of soft tissue sarcomas of the limbs after R0 resection was clearly demonstrated retrospectively [31]. If marginal surgery, irradiation of the tumoral bed with large safety margins improves local control rate in this indication. External radiation therapy is most often used, but some teams use intraoperative brachytherapy with good results and little late complications [32, 33]. Chemotherapy of sarcomas uses two major drugs: doxorubicin, and ifosfamide. The current challenges are to improve the combination of these drugs, particularly by modulating their doses. The role of adjuvant chemotherapy after resection of soft tissue sarcomas is uncertain and controversial [34, 35]. The present meta-analysis Sarcoma Meta-Analysis Collaboration [36] suggests that the use of an optimal schema based anthracycline / ifosfamide significantly prolongs survival [36]. However, the analysis did not include the data of the most important negative test, which, up to now has only been reported in the form of summary [37]. Overall, despite the latest positive meta-analysis, it is difficult to recommend adjuvant chemotherapy as standard practice for all patients with synovial sarcoma of the limbs. If there is an advantage of survival for adjuvant chemotherapy with doxorubicin, it seems weak, not exceeding 5 percent absolute increase in the rate of survival at 5 years [36]. In metastatic situation, the poly chemotherapy has the greatest place, which allows, in case of remission to operate the patient with prolonged survival [38].

Conclusion:

Synovial sarcoma is an uncommon tumor of the soft parts. Its histological diagnosis remains difficult. The Treatment is multidisciplinary consisting in a collaborative approach between (orthopedist, oncologist, histopathologist, psychotherapists .....) and it requires a radical excision as soon as possible. The modern imaging, the chemotherapy and the radiotherapy allow considerable optimism.

The authors declare no conflict of interest

References:

[1] Russel W, Cohen J, Enzinger et AL. A Clinical and pathological staging system for soft tissue sarcomas. Cancer 1977; 40: 1562-70.

[2] Hampole Mk, Jackson B; Analysis of 25 cases of malignant synovioma. Can med.Assoc.j, 1968 Nov 30; 99(21):1025-9.

[3] Hadju SI, Shiu M-H, Fortner J-G. : Tendinosynovial sarcoma, a clinic pathological study of 136 cases cancer 1977; 39, 1201-1217.

[4] Brodsky J, Hadju S, Casper E, Brennan M.: Tendino synovialsarcoma clinic pathologic features, treatment and prognosis. Cancer, 1992, 70, 484-489.

[5] Varela-Duran J, Enzinger FM. : Calcifying synovial sarcoma cancer, 1982, 50, 345-352.

[6] Gerner RE, Moore GE. : Synovialosarcoma. Ann Surg, 1975 Jan; 181(1):22-5.

[7] Haagensen CD, Stout AP. : Synovial sarcoma. Ann Surg. 1944 Dec; 120(6):826-42.

[8] Levi-Valensin G, Petit-Perrin M, Amouroux J, and Al: Synovialosarcome. Ann Radiol (Paris). 1979 Jan-Feb; 22(1):45-52.

[9] Breuil V, Flory P, Grimaud A, and Al: Les synovialosarcomes en milieu rhumatologique : a propos de 2 cas, 1996, 48,65-73.

[10] Vander W. Ultrasound in musculo-skeletal tumors. Radiol Clin North Am 1999; 4: 753-66.

[11] Emas B.E., Heelan R.T., Lane J. Soft-tissue sarcomas of the extremities. Am J Roentgenol 1988; 150: 615-20.

[12] De Schepper A.M., De Beuckeleer L., Vandevenne J., Somville J. Magnetic resonance imaging of soft tissue tumors. Eur Radiol 2000; 10: 213-23.

[13] Petasnick JP, Turner DA, Charters JR, Gitelis S, Zacharias CE. : Soft tissue mass of the locomotors system: Comparaison of MRI and CT Radiology, 1986, 160,125-133.

[14] Mankin H.J., Mankin C.J., Simon M.A. The hazards of the biopsy revisited. J Bone Joint Surg 1996; 78A: 656-63.

[15] Coindre JM. : Recommandations pour la prise en charge anatomopathologiques des sarcomes des tissus mous. Ann pathol. 1998, 18, 6,505-511.

[16] M.Mahfoud : cancer de l'os (tumeurs Malignes des membres)- édition 2009.

[17] Coindre J.M. Intérêt pratique de l'immunohistochimie dans le diagnostic des tumeurs des tissus mous. Rev Fr Lab 1994; 267: 41-8.

[18] Turc-Carel C., Pedeutour F., Durieux E. Characteristic chromosome abnormalities and karyotype profiles in soft tissue tumors. Curr Top Pathol 1995; 89: 73-94.

[19] Limon J, Mrózek K, Nedoszytko B, and al: Cytogenetic findings in two syovial sarcomas. Cancer Genet. Cytogenet, 1989, 38, 215-22.

[20] Noguera R, Lopez-Ginés C, Gil R, and al: Translocation (18) in a synovial sarcoma a new case. Cancer Genet Cytogenet, 1988, 33, 311-2.

[21] Sandberg AA, Bridge JA; Updates on the cytogenetics and molecular genetics of bone and soft tissue tumors. Synovial sarcoma,Cancer Genet Cytogenet. 2002 Feb; 133(1):1-23.

[22] Bowden L, Booher RJ. The principles and techniques of resection of soft parts of sarcoma 1958; Clin Orthop Relat Res. 2004 Sep ;(426):5-10.

[23] Enneking WF, Spanier SS, Malawer MM.The effect of the Anatomic setting on the results of surgical procedures for soft parts sarcoma of the thigh. Cancer. 1981 Mar 1; 47(5):1005-22.

[24] Bonvolot, D. Vanel.P.Terrier, C .le Pechoux.A.le Cerne : principes du traitement des sarcomes des tissus mous de l'Adulte EMC- Rhumatologie orthopédie (2004).

[25] Dirix L, Somville J, Van Oosterom A.: Diagnosis and treatment of soft tissue sarcomas in adults. Current Opinion Oncol 1996; 8: 289-98.

[26] Alvarenga J.C., Ball A.B., Fisher C., and al. Limitations of surgery in the treatment of retroperitoneal sarcoma. Br J Surg 1991; 78: 912-6.

[27] Y. Milbéo a, G. Kantor a, H. LAHARIE: Adjuvant radiation therapy for soft tissue sarcoma of the extremities: analysis of local control according to volume and dose Cancer/Radiotherapy 9 (2005) 293–303.

[28] Rydholm A, Gustafson P, Rooser B,Willen H,Akerman M, Herrlin K, et al. Limb-sparing surgery without radiotherapy based on anatomic location of soft tissue sarcoma. J Clin Oncol 1991; 9: 1757–65.

[29] Geer RJ, Wodruff J, Casper ES, Brennan MF. Management of small soft tissue sarcoma of the extremity in adults. Arch Surg 1992; 127: 1285–9.

[30] Standards. Options et Recommandations pour la prise en charge des patients atteints de sarcomes des tissus mous (http://www.fnclcc.fr/fr/ sor/ pdf/rapport_integral/ sarcome_integrale_0595.pdf).

[31] Khanfir K, Alzieu L, Terrier P, Le Pechoux C, Bonvalot S, Vanel D, et al. Does adjuvant radiation therapy increase loco regional control after optimal resection of soft tissue sarcoma of the extremities Eur J Cancer 2003; 39:1872–80.

[32] Alektiar KM, Velasco J, Zelefsky MJ, and al. Adjuvant radiotherapy for margin-positive high-grade soft tissue sarcoma of the extremity. Int J Radiat Oncol Biol Phys 2000; 48:1051–8.

[33] S. Heymanna, G.-M. Junga, P. SIMON: Late outcome of 89 patients with soft-tissue sarcomas treated by surgery and three different radiotherapy schedules Cancer/Radiotherapy 11 (2007) 443–451.

[34] Blay, JY, Le Cesne, A. Adjuvant chemotherapy in localized soft tissue sarcomas: still not proven. Oncologist 2009; 14:1013.

[35] Schuetze, SM, Patel, S. Should patients with high-risk soft tissue sarcoma receive adjuvant chemotherapy?. Oncologist 2009; 14:100

[36] Pervaiz, N, Colterjohn, N, Farrokhyar, F, et al. A systematic meta-analysis of randomized controlled trials of adjuvant chemotherapy for localized resectable soft-tissue sarcoma. Cancer 2008; 113:573.

[37] Bramwell, V, Rouesse, J, Steward, W, et al. Adjuvant CYVADIC chemotherapy for adult soft tissue sarcoma--reduced local recurrence but no improvement in survival: a study of the European Organization for Research and Treatment of Cancer Soft Tissue and Bone Sarcoma Group. J Clin Oncol 1994; 12:1137.

[38] J. Fayette a, J.-Y. BLAY: Chemotherapy of sarcomas: Optimization of existing drugs and new molecules Cancer/ Radiotherapy 10 (2006) 72–77