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Disabling Giant Fibrosarcoma of the Shoulder Girdle at the Bamako Dermatology Hospital in Mali

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Abstract **Case Report**

Fibrosarcoma is defined by the World Health Organization (WHO) as a malignant tumour, characterized by the presence of intertwined bundles of collagen fibers, and by the absence of other types of histological structures, such as bone or cartilage. In this article, we describe the observation of a case of disabling giant fibrosarcoma at the shoulder girdle in order to highlight the clinical, Para clinical and therapeutic particularities. This is a 58-year-old male patient with a history of apparently good health. He reports that for 18 years he has had a pimple on his shoulder that has been growing until it reaches its current very large size and causing him a disability. After an unremarkable blood test, the patient was surgically treated by performing a wide excision with a good surgical margin and skin grafting. The surgical specimen weighed 3700 g. The patient was regularly followed up for 4 years without recurrence. Soft tissue sarcomas are a heterogeneous group of tumours that are not very common and histologically diverse, including fibrosarcoma, and whose treatment results in a wide surgical resection with free margins. Skin grafting provides a very satisfactory result face to huge generated loss of tissues while removing the handicap in patients with giant tumours in our resource-limited countries.

Keywords: Fibrosarcoma, Disabling, skin grafting.

INTRODUCTION

Sarcomas are malignant neoplasms originating from mesenchymal cells that can arise in any anatomical location [1].

Fibrosarcoma is defined by the World Health Organization (WHO) as a malignant tumour, characterized by the presence of intertwined bundles of collagen fibers, and by the absence of other types of histological structures, such as bone or cartilage [2].

Of all tumours, only 1% corresponds to primary sarcomas and, of these, 36% are fibrosarcomas [2].

Regarding its aetiology, association with some genetic disorders and radiation exposure has been observed [1].

Histologically, this tumour is monotopic and monostotic and occurs most frequently in the metaphyses of long bones, such as the distal femur and proximal tibia, as well as in the skull, maxilla, and vertebrae. It has many histological similarities with malignant fibrous histiocytoma and may appear in a patient with Paget's disease or a giant cell tumour [3].

The degree of cellular differentiation and mitotic activity may be stratified into four levels (tumour grade) and used to predict the clinical behavior of a tumour [4]. They are generally radioresistant tumours and surgery is the treatment of choice [3]

In this article, we describe the observation of a case of disabling giant fibrosarcoma at the level of the shoulder girdle in order to highlight the clinical, paraclinical and therapeutic particularities.

OBSERVATION (PATIENT AND METHOD)

This is a 58-year-old male patient with a history of apparently good health. He reports that for 18 years he has had a pimple on his shoulder that has been growing until it reaches its current very large size and causing him a disability. The mass began to ulcerate for which he undertook traditional treatment without success after which he consulted in a third level hospital from where he was referred to us for better management.

On physical examination we found a large mass made of multiple tumour nodules at the level of the shoulder girdle reaching the pectoroscapulohumeral region with isolated ulcerations and secretion of bad odour (Figure 1). A CT scan was requested (Figure 2), which informed an infiltration of the adjacent muscles in particular the pectoralis major, the biceps and the deltoid. The largest nodular mass measured 162 X 96 cm. There were axillary lymph nods, the largest of which measured 15 mm in the short axis. There was no secondary localization. An excisional biopsy was performed preoperatively and informed us that the fragments examined were the site of a fasciculate tumour proliferation made of spindle cells with hyper chromatic nuclei containing abnormal mitoses (one per 5 fields at high magnification). The epidermis was atrophic.

After an unremarkable blood test, the patient was surgically treated by performing a wide excision with a good surgical margin. The surgical specimen (Figure 3) weighed 3700 g which was entirely returned for study to the anatomy pathology department which concluded that the tumour corresponded to a fibrosarcoma made of bundles of spindle cells containing atypical and abnormal mitoses and that the resection limits were healthy (Figure 4).

After the excision, a partial skin graft was performed which, after healing, gave a satisfactory result from an aesthetic and functional point of view (Figure 5). The patient was regularly followed up for 4 years without recurrence.

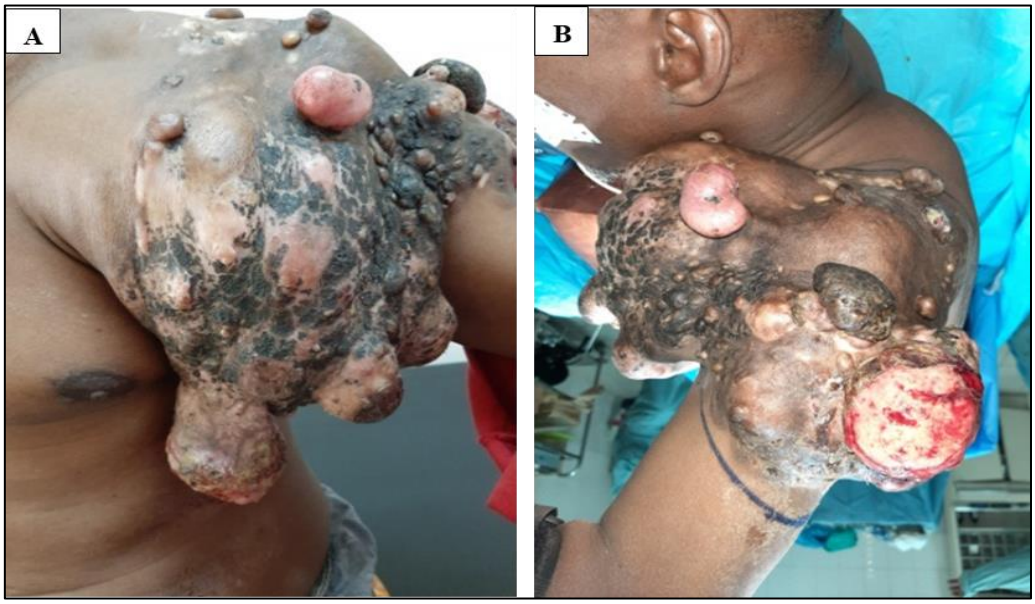

Figure 1: Clinical appearance of the tumour in frontal (A) and lateral (B) view respectively

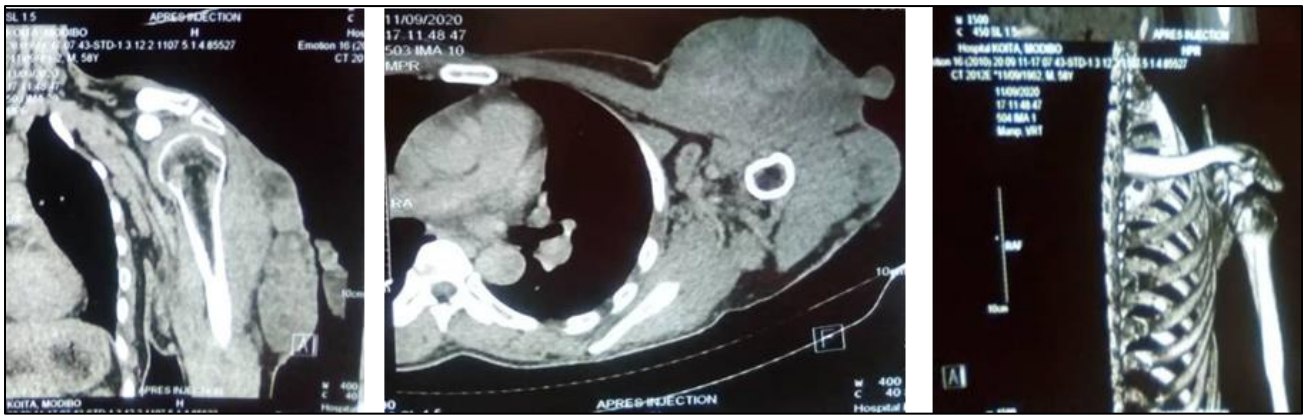

Figure 2: CT scan of tumour appearance

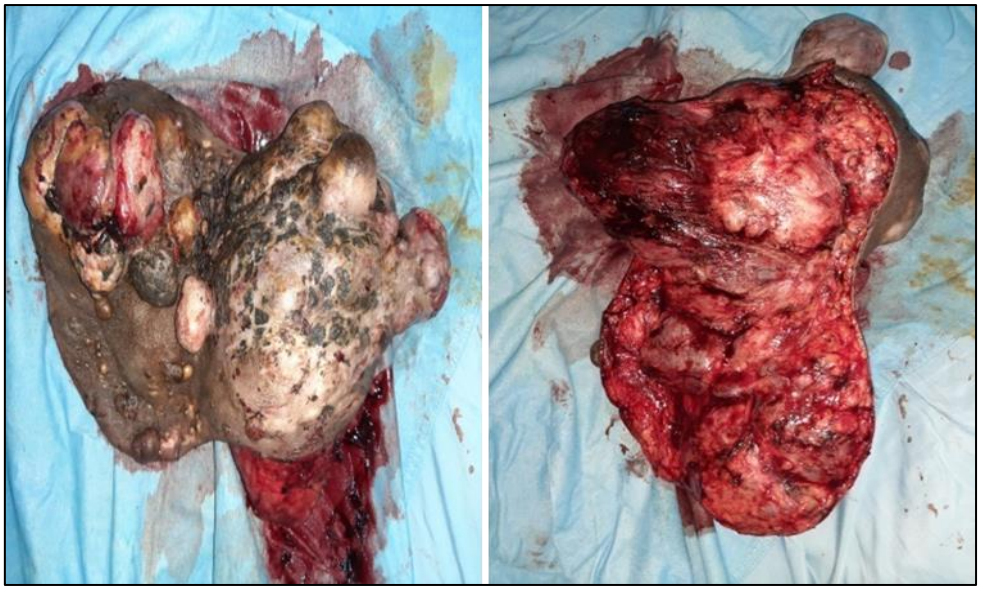

Figure 3: Tumour surgical specimen, upper and lower aspect respectively from left to right

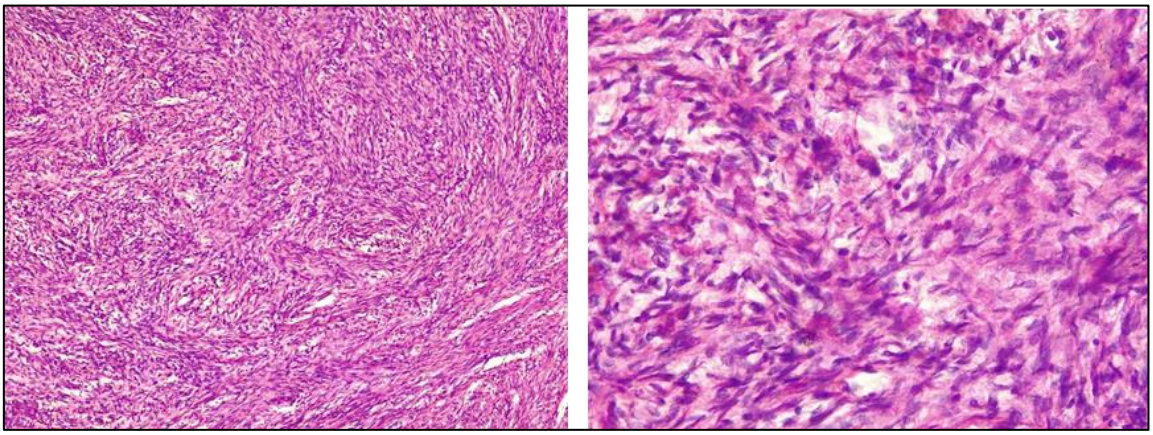

Figure 4: Histological study result of the surgical specimen at X10 and X40 objectives from left to right

Figure 5: Immediate postoperative results (A), at day 28 (B) and at one year (C)

DISCUSSION

Soft tissue sarcomas (STS) are a diverse group of malignant tumours arising from common and specialized extra skeletal connective tissue. These cancers constitute a rare entity, representing approximately 1% of all malignant tumours [5].

Soft tissue sarcomas have different cellular origins and therefore different diagnoses; however they are grouped together due to similarities in their histological appearance, clinical presentation and natural history [6].

Adult soft tissue sarcomas are a heterogeneous group of rare and histologically diverse tumours that arise primarily from the mesenchyme, which comes from the mesoderm and embryonic disk, and can therefore originate in the gastrointestinal mesenchymal tissues, the extremities (59%), trunk (19%), retro peritoneum (15%), head and neck (9%) and more rarely the tract [7].

More than 50 histological types have been identified but the most common are malignant fibrous histiocytoma (28%), leiomyosarcoma (12%), liposarcoma (15%), synovial sarcoma (10%) and malignant peripheral nerve sheath tumours (6%) [7, 8].

Fibrosarcoma presents on average at age 45 years (range 16 - 85 years), it presents as a tumour in 98% of cases and causes pain in 17%, the average size is 6 cm at the time of diagnosis, its most frequent location is in the lower limbs (58%), trunk (22%), upper limbs (18%), head and neck (2%) (38) [9].

According to Diarré T *et al*., the most aged patients were affected by the expansion of fibrosarcomas as it was found in the literature [5].

The most frequent location is in the root of the limbs (more common in the lower limbs), trunk and neck. It rarely appears in the abdomen, pelvis, retro peritoneum or viscera [10].

Tumours in the distal extremities are often small when discovered, while tumours in the proximal extremities and retro peritoneum may be voluminous at the time of diagnosis; they grow centrifugally, compressing surrounding structures, but rarely affect bone or neurovascular bundles, causing pain, oedema and inflammation [11].

The typical clinical picture is a circumscribed mass of firm consistency, slow-growing and painless [12].

In the extension study, in all cases of soft tissue sarcomas a chest CT should be performed [13].

The treatment of this entity is surgical, taking into account current oncological standards and its therapeutic approach should be multidisciplinary, in order to achieve a correct functional and aesthetic rehabilitation of the patient [1]. Surgery is the treatment of choice [3].

Edward H et al found that primary treatment consisted of local excision in 93 patients, amputation of the extremity in 10, radiation in 6, and one patient was left untreated. Of the 93 patients who had a local excision, 44 (47%) had recurrences one or more times, and 13 of these underwent amputation for recurrent tumour. Adjunctive radiation after local excision was given to 13 patients [4].

Treatment is usually extended surgical resection with free margins. Occasionally, amputation may be required if the bone is affected [14].

CONCLUSION

Soft tissue sarcomas are a heterogeneous group of tumours that are not very common and histologically diverse, including fibrosarcoma, and whose treatment results in a wide surgical resection with free margins. The closure of the generated loss of substance poses a huge problem, but fortunately skin grafting provides a very satisfactory result while removing the handicap in patients with giant tumours in our resource-limited countries.

Conflict of Interest: The authors declare that they have no conflict of interest in relation to this article.

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