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Original Research Article

Pleiomorph Sarcome of the Thigh

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Abstract: This article focuses on pleomorphic sarcomas, which are malignant mesenchymal tumors with a complex genetic background underlying their morphological pleomorphism. These are poorly differentiated tumors that can maintain different lines of differentiation, sometimes correlating with clinicopathological or prognostic characteristics. Accurate diagnosis in this group of tumors relies on adequate sampling due to their heterogeneity and evaluation by both microscopy and large immunohistochemistry panels. Molecular analyzes have a limited role in their diagnosis as opposed to translocation-related sarcomas, but they may provide important the anotic and prognostic information in the future.

Keywords: Thigh-Surgery-Tumor-Sarcoma-Resection.

INTRODUCTION

A pleomorphic sarcoma is an aggressive, rapidly growing soft tissue tumor that mainly affects the lower limb and thigh.

It can be differentiated into lipo- or fibro-sarcoma, but also undifferentiated. The undifferentiated form is the most common and represents 10% of all soft tissue tumors2, while the liposarcomatous form is the rarest.

Our article concerns the case of a 58-year-old patient, followed in the Orthopedic Department of Ibn Sina Hospital in Rabat, for a large tumor of the anterointernal part of the right thigh and for whom the treatment was consisted of surgical resection of the tumor after histological evidence was obtained and which was in favor of a pleiomorphic sarcoma of the thigh.

Pleomorphic sarcomas encompass a group of mesenchymal malignancies associated with complex genetic characteristics lacking defining alterations.

They can have a wide range of histologic lineages, including myogenic, lipogenic, neurogenic, or none and mainly affect adult patients aged over 50 years.

Morphologically, they exhibit variable levels of pleomorphism, although pleomorphism is not always a feature of malignancy.

Prognostic stratification can be assessed with the French three-level classification system FNCLCC, taking into account morphological characteristics, mitotic activity and tumor necrosis.

MATERIEL AND METHODS

Our article concerns the case of a 58-year-old patient, F.B, followed in the Orthopedic Department of Ibn Sina Hospital in Rabat, for a large tumor of the anterointernal part of the left thigh and for whom the treatment was consisted of surgical resection of the tumor after histological evidence was obtained and which was in favor of a pleomorphic sarcoma of the thigh.

The follow-up of this patient was long-term. He presented with a progressive increase in the volume of his right thigh over a period of 4 months and whose pain gradually worsened.

There was no vascular compression and the extension assessment did not find any pulmonary, hepatic, bone or other metastases.

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The first surgical stage consisted, after having had a complete radiological assessment, of a tumor biopsy, which was in favor of an undifferentiated pleomorphic sarcoma. The second surgical stage consisted of oncological resection of the tumor.

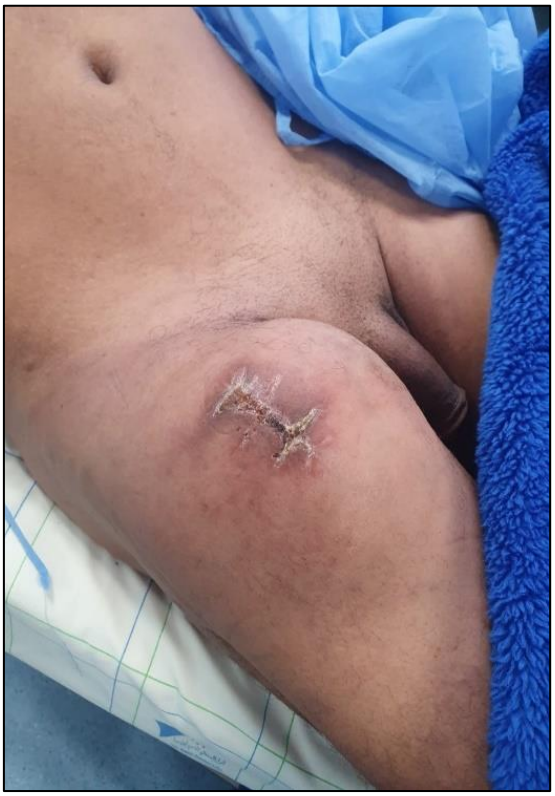

Figure 1: Macroscopic aspect of the thigh

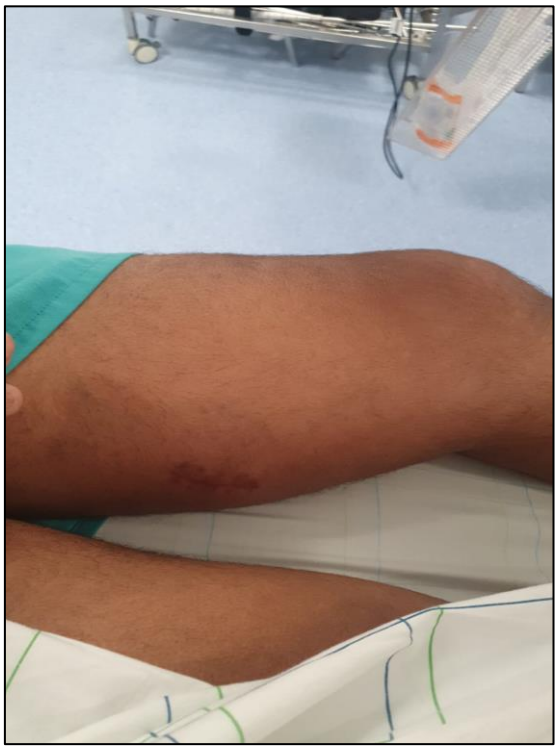

Figure 2: Other picture of the macroscopic aspect of the thigh

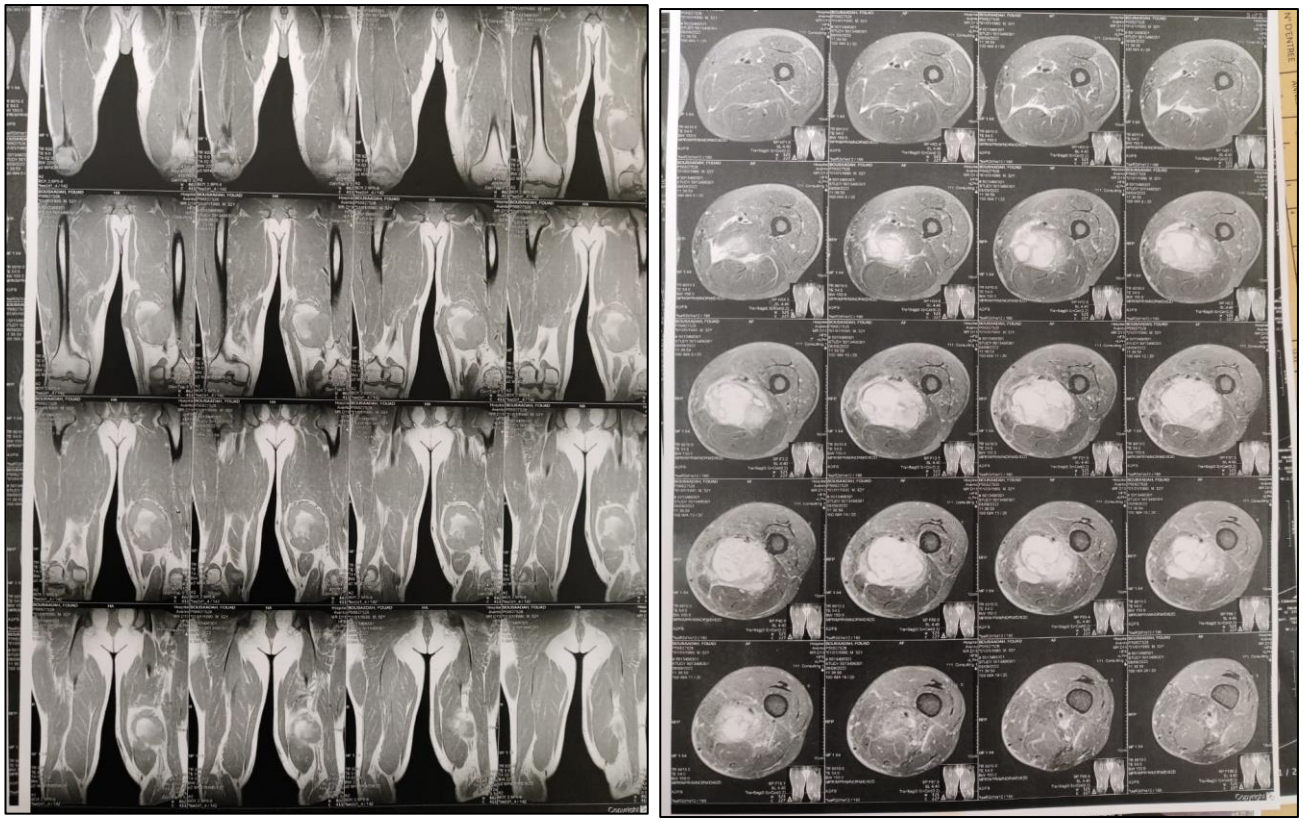

Figure 3: Radiological aspect of the thigh tumor based on the MRI (transversal and frontal incidences)

Shwanomme

TECHNIQUES : Examen réalisé par une IRM 1,5 Tesla type Siemens Avanto dernière génération en coupes coronales et axiales T1 et T2 Fat Sat, puis sagittales T2 Fat Sat, et T1 injecté dans les trois plans de l'espace

RÉSULTATS :

- Volumineuse masse inter-musculaire de la partie inférieure de la lige des adducteurs, aux contours polycycliques bien circonscrits, sans afférence ou efférence évidentes au niveau de ses pôles, de signal hétérogène mixte avec

- un contingent tissulaire périphérique en isosignal T1, hypesignal T2 et diffusion à b1000 avec restriction de l'ADC (témoignant d'une cellularité élevée), rehaussé de façon modérée diffuse.
- un contingent central infiltrant partiellement le tissu périphérique, en hypersignal T1 hypersignal intense T2 non effacé par la saturation de graisse ou en IP OP, non rehaussé.

- Elle mesure 12,2 x 9 x 8cm (H x E x AP)
- Elle refoule et comprime les structures adjacentes:

- les muscles sans les envahir: les adducteurs en haut et en dedans, le gracile en dehors (avec remaniement du trajet de biopsie de ce dernier en son sein)
- les vaisseaux fémoraux en avant et en dedans. L'artère fémoral est perméable et la veine fémorale n'est pas visualisé au contact de la masse sur le 1/3 inférieur de la cuisse sur environ 7 cm (compression ou envahissement ?) puis elle présente un aspect thrombosé au niveau du creux poplité (à confronter aux données doppler)

- Liseré graisseux de séparation avec le fémur. Pas de lésion osseuse suspecte
- Pas d'autre atteinte

CONCLUSION: Processus tumoral inter-musculaire du 1/3 inférieur de la loge des adducteurs gauches, dont les critères IRM ne sont pas spécifiques, doute sur une thrombose veineuse fémorale inférieure en amont d'une compression de la masse.

Figure 4: Radiological conclusion of the MRI

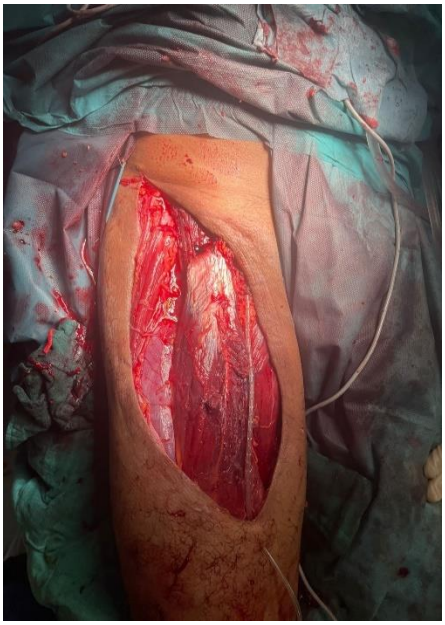

Figure 5: Aspect of the thigh after the tumor resection

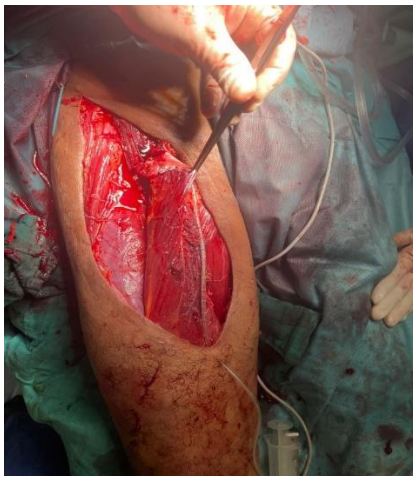

Figure 6: Other picture of the skin incision

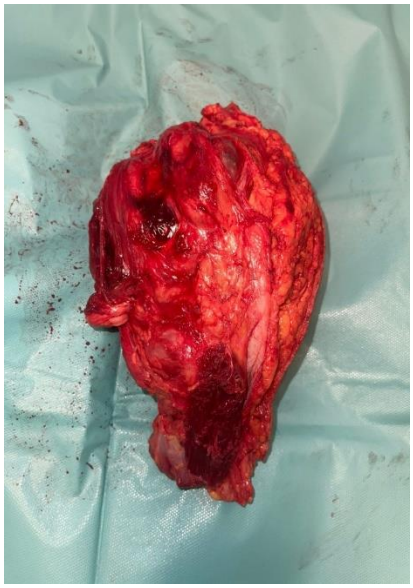

Figure 7: Macroscopic aspect of the tumor

RESULTS

After the surgical procedure, the patient remained under surveillance in the department for 96 hours; he received a transfusion of three packed red cells with an exit control hemoglobin of 10.1. Furthermore, the redon drain brought back 140 cc in 48 hours.

The postoperative course was simple; the bandage was changed every 48 hours and was never soiled. After a multidisciplinary consultation meeting with oncologists and radiologists, the indication for adjuvant radiotherapy or chemotherapy was not retained.

With one year's follow-up and follow-up in consultation for this patient, the evolution was favorable; there was no local or distant recurrence. He was able to resume his professional activity as a taxi driver after 2 months.

DISCUSSION

Undifferentiated pleomorphic sarcoma (UPS) is a rare and aggressive cancer that develops in connective tissues, including muscles, tendons, fat, and fibrous tissues. The term “undifferentiated” indicates that tumor cells do not resemble normal cells in the body and lack specific differentiation toward a particular tissue type. “Pleomorphic” refers to the varying shapes and sizes of tumor cells, which can appear abnormal and irregular under a microscope.

Undifferentiated pleomorphic sarcoma usually occurs in the arms, legs, or trunk, although it can also be found in deeper tissues such as the retroperitoneum (the area behind the abdominal cavity). It most often affects older people, but can occur at any age.

Most undifferentiated pleomorphic sarcomas present as a rapidly growing mass sometimes associated with pain. The cause of the vast majority of tumors remains unknown to this day. However, up to 25% of tumors associated with prior radiotherapy are diagnosed as undifferentiated pleomorphic sarcomas.

The diagnosis of undifferentiated pleomorphic sarcoma is often first suggested after a small sample of tumor is removed during a procedure called a biopsy. In the biopsy report, your pathologist will provide your doctor with a list of possible diagnoses, including undifferentiated and dedifferentiated pleomorphic sarcomas. Often, a final diagnosis cannot be made until the entire tumor has been surgically removed and the pathologist has been able to examine the entire sample.

When examined under a microscope, undifferentiated pleomorphic sarcoma consists of large, very abnormal-looking tumor cells. Tumor cells are described as pleomorphic because they exhibit significant variation in cell size and shape. Mitotic figures (tumor cells divide to create new tumor cells) are frequently found, and atypical mitotic figures may also be found.

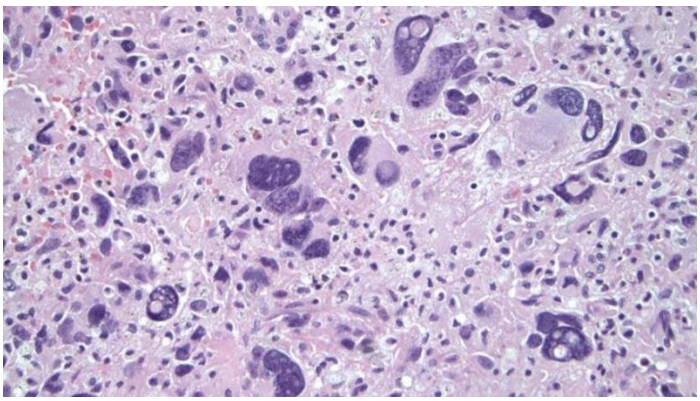

Figure 8: Histological aspect of the pleiomorph sarcoma

Immunohistochemistry is a test that allows pathologists to look for specific types of proteins inside cells. Pathologists use the results of this test to determine the function of the cell and where it originated in the body. When immunohistochemistry is performed on undifferentiated pleomorphic sarcoma, the tumor cells are usually only positive or reactive for non-specific cellular markers such as smooth muscle antigen (SMA), p16, p53. Tumor cells are generally negative for more specific markers such as desmin, ERG, caldesmon, S100, SOX-10, cytokeratin, p40.

At our current level of understanding, undifferentiated pleomorphic sarcoma has no known characteristic molecular changes. However, your pathologist may perform molecular testing on the tumor sample to rule out other sarcomas that may resemble undifferentiated pleomorphic sarcoma. A negative molecular test (e.g., with no identified translocation or amplification) is consistent with undifferentiated pleomorphic sarcoma. Pathologists test for these molecular changes by performing fluorescence in situ hybridization (FISH) or next generation sequencing (NGS). This type of test is more often performed on the biopsy specimen. If your pathologist is certain that the

tumor is an undifferentiated pleomorphic sarcoma, no molecular testing can be performed.

a) Sarcoma classification system of the French Federation of Cancer Centers (FNCLCC)

The sarcoma scoring system of the French Federation of Cancer Centers, or FNCLCC, is a system that pathologists use to score sarcomas, including undifferentiated pleomorphic sarcoma. The grade helps predict the likely behavior of the tumor, including how quickly it might grow and whether it might spread to other parts of the body.

The FNCLCC system assigns a score to the tumor based on three components:

Mitotic activity: This is the number of cells that are actively dividing in the tumor. Pathologists count the number of mitotic figures (dividing cells) in an area measuring 10 high-power fields under a microscope. The mitotic activity score can range from one to three, with a higher score indicating more frequent cell division and a potentially more aggressive tumor.

Necrosis: Necrosis refers to areas of the tumor where cells have died. The necrosis score also ranges from one to three, with a higher score indicating more extensive necrosis, which generally suggests a more aggressive tumor.

Differentiation: Differentiation refers to the extent to which tumor cells resemble normal cells. In the FNCLCC system, all undifferentiated pleomorphic sarcomas are assigned a differentiation score of three because the tumor cells do not resemble normal cells.

The scores for these three components are added together to give the tumor an overall grade, ranging from grade 1 (low grade) to grade three (high grade). A higher grade indicates a more aggressive tumor.

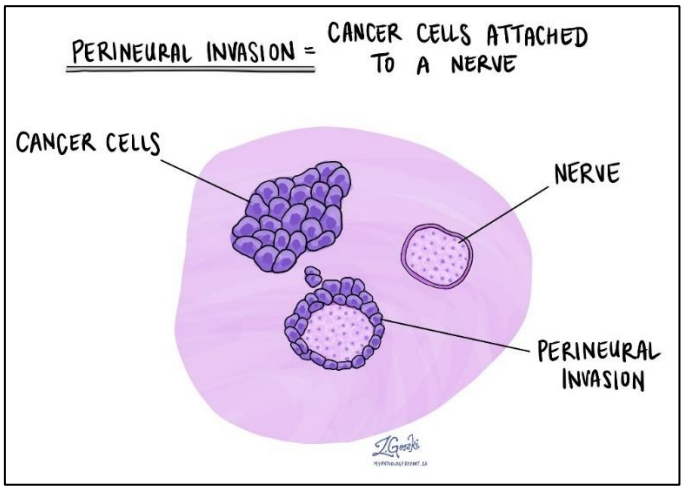

Figure 9: Neural invasion of a nerve by cancer cells

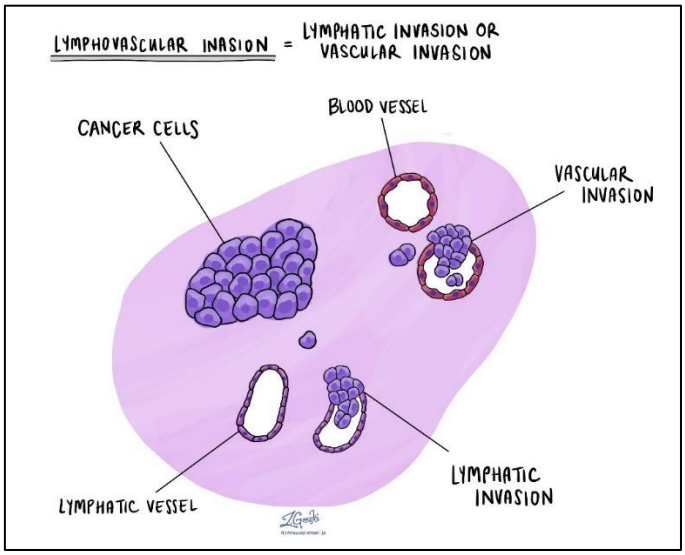

Figure 10: Lymphatic invasion by tumor cells

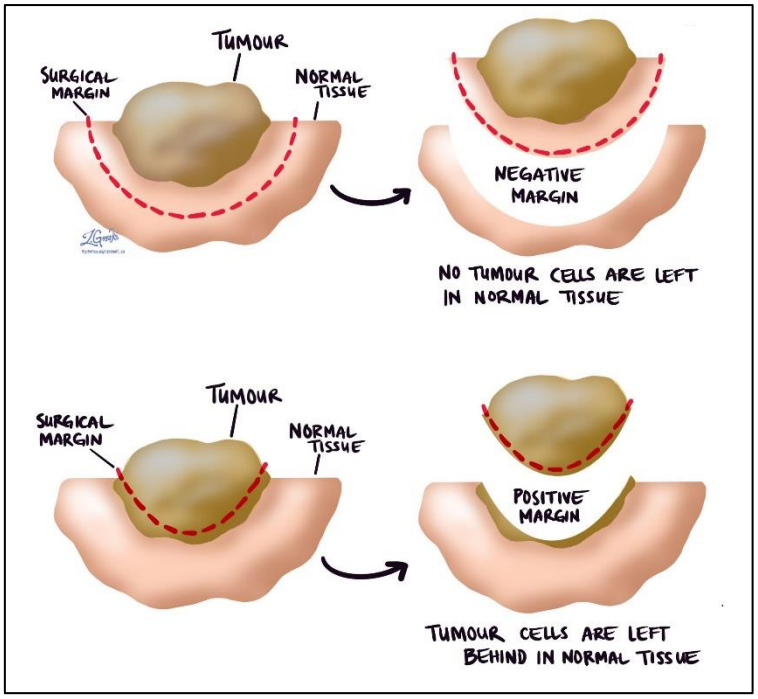

Figure 11: Margin of the surgical resection of the pleiomorph sarcom

CONCLUSION

A pleomorphic sarcoma, called malignant histocytofibroma until 20021, is a soft tissue tumor in the sarcoma family. It is an aggressive, rapidly growing tumor that mainly affects the lower limb and thigh. It can be differentiated into lipo- or fibro-sarcoma, but also undifferentiated. The undifferentiated form is the most common and represents 10% of all soft tissue tumors, while the liposarcomatous form is the rarest.

The standard treatment for pleomorphic sarcomas is surgery, based on complete resection of the tumor in a single block. Inoperable tumors can be treated with radiotherapy and chemotherapy.

Biopsy is crucial for treatment planning. It determines the histological type of the sarcoma. Following histological analysis and radiological assessment, the stage of the disease is determined, that is to say its extent. Several diagnostic tests are performed depending on the type of sarcoma. They include in particular a magnetic resonance (MRI), a chest computed tomography (scan), an abdominal-pelvic computed tomography (scan), a bone scintigraphy or sometimes a positron emission tomoscintigraphy (PET scan).

When a person is diagnosed with soft tissue or bone cancer, a specialized, multidisciplinary team establishes a personalized treatment plan with them. This treatment plan can vary depending on different elements, such as the type of cancer, its stage and the person's health. The treatment plan also takes into account the wishes and concerns of the affected person and those of their family.

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