World Cancer Research Journal WCRJ 2022; 9: e2238

# HIGH-GRADE UNDIFFERENTIATED PLEOMORPHIC SARCOMA IN THE GLUTEUS WITH A V-Y FLAP: A CASE REPORT

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**Abstract – *Objective:*** *Undifferentiated pleomorphic sarcoma (UPS) is rare and affects mainly the elderly. Due to the low incidence and heterogeneous variety of this sarcoma, little is known about the UPS regarding clinical behavior, treatment strategies and prognosis. The objective is to describe the case of a young patient, with a UPS measuring 14 × 11.2 × 8.2 cm, with a 6-month evolution and no metastases.*

***Case Report:*** *A female patient, 34 years old, reported the presence of nodulation in the left gluteal region for 6 months with progressive growth associated with pain, weight loss of 10 kg in the last 2 months. An anatomopathological study showed a malignant neoplasm of spindle cells and high-grade pleomorphic sarcoma on immunohistochemistry. Neoadjuvant chemotherapy and complete resection of the lesion with subsequent adjuvant radiotherapy were performed.*

***Results:*** *Since undifferentiated pleomorphic sarcoma has a better prognosis when diagnosed early and in a small size, it is crucial to immediately recognize and assess any enlarged mass and nodule, even though it is a rare diagnosis.*

***Conclusions:*** *Due to the high rate of recurrence and metastasis, monitoring should be carried out regularly for years after surgical excision.*

**KEYWORDS:** *Pleomorphic sarcoma, High grade, Case report.*

## INTRODUCTION

Soft tissue sarcomas represent less than 1% of all neoplasms. Among them, undifferentiated pleomorphic sarcoma (UPS) is one of the most common types, with an incidence of approximately 0.08–1/100,000. It usually occurs in adults between the fifth and seventh decades of life, with a slight predominance in males, and tends to occur in the extremities, retroperitoneum, viscera, and head and neck[1,2]. Owing to the low incidence and heterogeneous variety of this sarcoma, little is known about UPS regarding clinical behavior, treatment strategies, and prognosis[1].

In this report, we present the case of a female patient with high-grade undifferentiated pleomorphic sarcoma in the left gluteal region, measuring 14 × 11.2 × 8.2 cm, with muscle involvement and without the presence of metastases. Neoadjuvant chemotherapy was performed with subsequent resection of the tumor and a myocutaneous flap in V-Y associated with post-surgical adjuvant radiotherapy.



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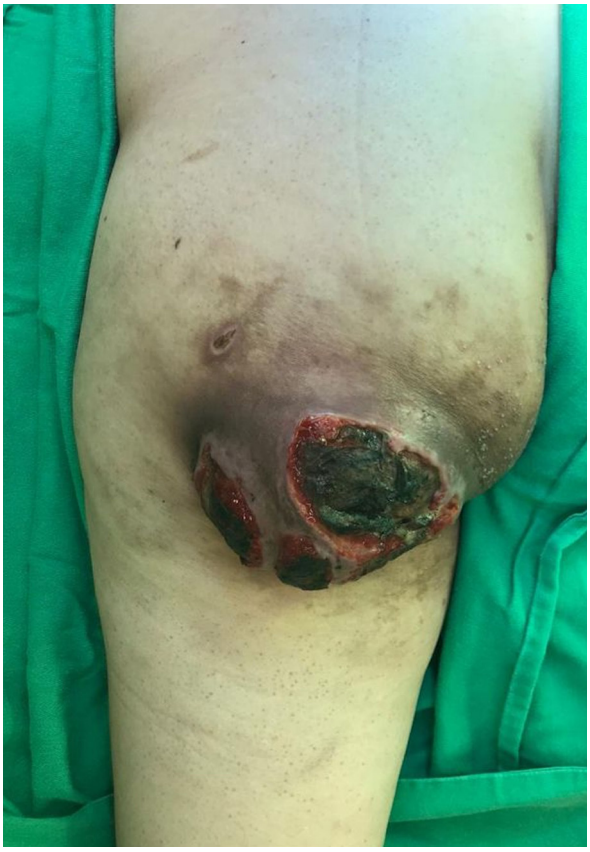


**Figure 1.** Nodular lesion in the left gluteus with inflammatory signs and areas of necrosis.

### *Ethics Committee and Informed Consent*

This article was registered under CAAE 45326321.8.0000.5362 and approved by the Ethics Committee of the proposing institution under opinion No. 4,644,920. Informed consent form in the use of information from the medical records and images was signed by the patient.

### *Case report*

A female patient, 34 years old, reported the presence of nodulation in the left gluteal region for 6 months with progressive growth associated with pain, weight loss of 10 kg in the last 2 months, and a Karnofsky performance status (KPS) of 90% with no other symptoms. On physical examination, she presented with a nodular lesion in the left gluteus with inflammatory signs and areas of necrosis (Figures 1 and 2). Ultrasonography was performed in the region, which revealed a well-delimited hypoechoic formation, with involvement of deep planes, approximately 124 × 81 × 91 mm, with peripheral vascularization identified using Doppler ultrasound imaging.

Soft tissue and chest tomography were performed for staging in addition to biopsy of the lesion. Soft tissue examination revealed solid hypodense formation with heterogeneous contrast enhancement, lobulated contours, and skin and subcutaneous topography without clear cleavage, with an inferior border of the left gluteus maximus muscle, measuring approximately 117 × 77 × 37 mm. No significant changes were observed using chest computerized tomography. An anatomopathological study showed a malignant neoplasm of spindle cells and high-grade pleomorphic sarcoma on immunohistochemistry.

Neoadjuvant treatment was initiated with chemotherapy with doxorubicin and phosphamide, followed by a surgical plan. Two days after the completion of the first cycle, the patient sought emergency care due to malaise, hypotension, dyspnea, and fever. On admission, she presented with tachycardia, high fever, high blood pressure, and pancytopenia on laboratory tests. The patient was hospitalized for febrile neutropenia with a probable cutaneous focus, and antibiotic therapy, meropen-

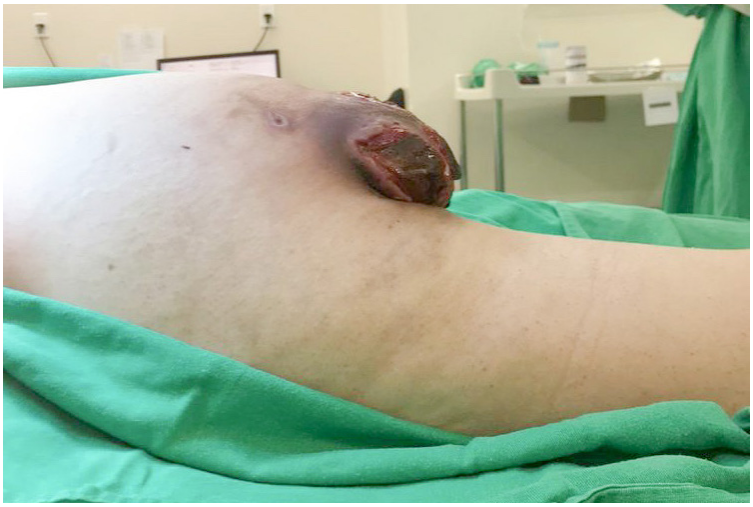
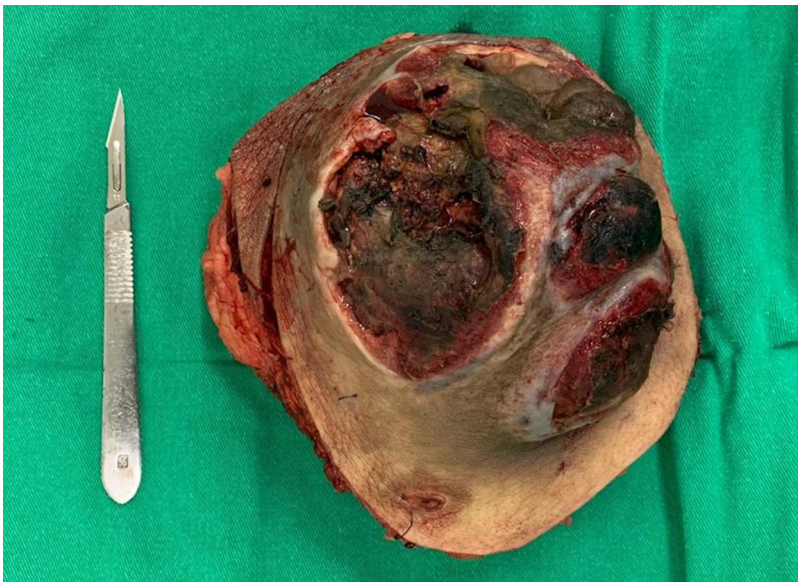


**Figure 2.** Nodular lesion in the left gluteus with inflammatory signs and areas of necrosis.

**Figure 3.** Tumor after excision.



em, and vancomycin were started, in addition to tranexamic acid, filgrastim, and two packed red blood cells. After 8 days of treatment, the patient's condition improved, with normalized leukocyte and platelet counts, and was then opted for surgery.

During the surgical procedure, after visualization of the involvement of the gluteus maximus muscle by the tumor, but without involvement of other muscles or sciatic nerve, the tumor was removed with a safety margin and a myocutaneous flap was performed in V-Y (Figures 3 and 4). The mass was sent for anatomopathological study, which revealed undifferentiated pleomorphic sarcoma with a size of 14 × 11.2 × 8.2 cm with a differentiation score of 3, mitotic rate of 3, and necrosis score of 1, resulting in a high-grade grade 3 sarcoma, according to the National Federation of French Cancer Centre's (FNCLCC) histological classification system. Free surgical margins with a minimum measurement of 2 mm were identified without angiolymphatic invasion. Due to the patient's good condition and an adequate flap, with no signs of ischemia, infection, or dehiscence, she was discharged on the seventh postoperative day and is currently undergoing adjuvant radiotherapy.

## DISCUSSION

Undifferentiated pleomorphic sarcoma, formerly known as malignant fibrous histiocytoma, is one of the most common soft tissue sarcomas in late adulthood, accounting for 20% of sarcomas. Approximately 25% of these sarcomas are due to previous exposure of the site to radiotherapy. Furthermore, UPS has been associated with certain syndromes such as Werner, Gardner, Li Fraumeni, and Von Recklinghausen[3]. However, none of these conditions were present in our patient.

Symptoms usually start with a palpable and painless mass, and as growth progresses, there may be symptoms of local mass effects, such as paresthesia, edema, and phlogistic signs[2,3]. Due to little symptomatology at the beginning, delay in diagnosis is extremely common. Primary in-

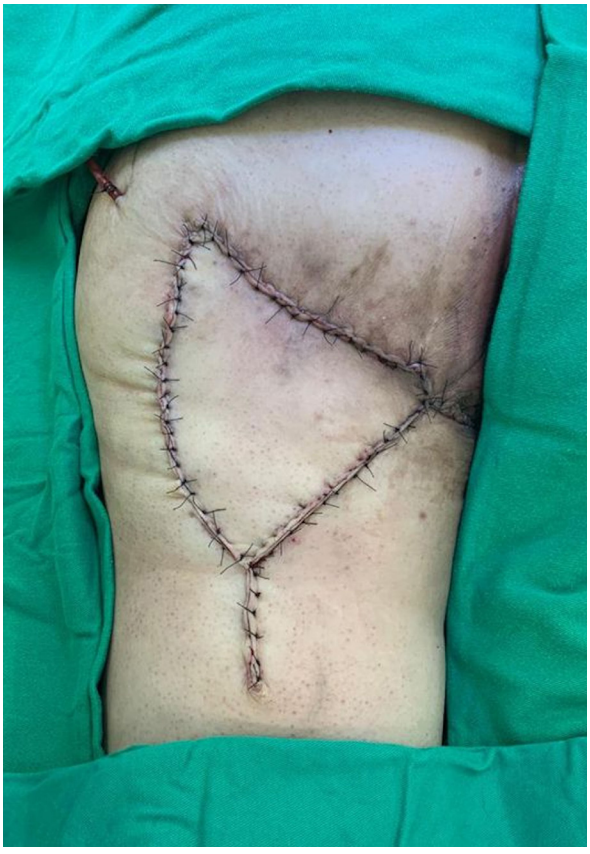


**Figure 4.** Final result with V-Y myocutaneous flap.

vestigation is through image examination to analyze the location, size, and invasion of adjacent structures[2]. However, these results do not diagnose UPS, which requires a biopsy. Histological diagnosis involves microscopy, molecular studies, and immunohistochemical techniques[3]. Regarding the analysis of our patient, an undifferentiated degree of differentiation (corresponding to a score of 3), mitotic rate greater than 20 (a score of 3), and presence of less than 50% of the area of necrosis (a score of 1) were found, resulting in a high-grade sarcoma. In addition, T3NXM0 anatomopathological staging was performed according to the American Joint Committee on Cancer (AJCC) and Union for International Cancer Control (UICC).

It is also recommended that all patients diagnosed with sarcoma undergo chest tomography to assess pulmonary metastasis and abdomen tomography to assess extrapulmonary metastasis to the abdomen and retroperitoneum[3].

Surgery is the standard treatment for all patients with complete excision and margins of at least 2 cm. Chemotherapy and radiotherapy can be used in treatment, mainly in deep high-grade lesions larger than 5 cm, but their impact on patient survival is still controversial[3]. In this patient, a neoadjuvant chemotherapy cycle was performed before the surgery, as well as adjuvant radiotherapy after surgery.

The patient prognosis is variable and includes factors such as size, degree, location, and inflammatory components. Despite complete excision, the recurrence rate can reach 51%[3]. According to a study carried out by Pezzi et al[4], the 5-year survival rate for high-grade tumors larger than 10 cm, as seen in our patient, is 41%. Furthermore, the metastasis rate in tumors between 10 and 15 cm is 49%, with 70–80% of these metastases being pulmonary[5].

Retrospective studies have determined that adequate resection margins during treatment are the most important prognostic factor for long-term survival, as local recurrence and distant metastases are significantly associated with higher mortality[2]. About 82% of patients in whom recurrence is identified also had associated pulmonary metastasis[6].

Because the primary objective of surgery is to achieve negative surgical margins, this procedure usually results in large soft tissue defects after resection that are not amenable to primary closure. Therefore, the use of similar local tissue is ideal for replacement, but donor availability can be a limiting factor. Traditional surgical procedures, such as the V-Y myocutaneous flap performed on the patient, are sufficient to satisfy both aesthetically and functionally. In addition, high-quality tissue is essential for adjuvant radiotherapy purposes[7].

## CONCLUSIONS

Since undifferentiated pleomorphic sarcoma has a better prognosis when diagnosed early and in a small size, it is crucial to immediately recognize and assess any enlarged masses and nodules, even though it is a rare diagnosis. In addition, growth is accelerated; in this case, in 6 months of evolution, the tumor reached a size of 14 × 11.2 × 8.2 cm. Local resection should be performed, and additional radiotherapy and chemotherapy may be necessary. Due to the high rate of recurrence and metastasis, monitoring should be performed regularly for years after surgical excision.

**Conflict of Interest:**
The authors have no conflict of interest to declare

**Funding:**
This research did not receive any specific grant from funding agencies in the public, commercial, or not-for-profit sectors.

**Ethical approval:**
This article was submitted and approved by the responsible Ethics Committee.

**Informed consent:**
Informed consent was obtained from the participant included in the study.

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