

Case report

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Received on: 31/01/2020

Approved on: 13/11/2020

Study conducted at the Universidade
de Mogi das Cruzes, Mogi das Cruzes
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Financial support: None.

Conflict of interest: None.



Undifferentiated pleomorphic sarcoma: a case report

Sarcoma pleomórfico indiferenciado: Case report

DOI: <https://www.dx.doi.org/10.5935/scd1984-8773.20201242522>

ABSTRACT

Undifferentiated sarcomas are neoplasms of soft tissues derived from the mesoderm. We report a case of undifferentiated pleomorphic sarcoma, its dermoscopy, staging, and therapeutic approach.

Keywords: Dermoscopy; Sarcoma; Soft Tissue Neoplasms

RESUMO

Os sarcomas indiferenciados são neoplasias de tecidos moles derivados do mesoderma. Relatamos um caso de sarcoma pleomórfico indiferenciado, sua dermatoscopia, seu estadiamento e sua abordagem terapêutica.

Palavras-chave: Dermoscopia; Neoplasias de Tecidos Moles; Sarcoma

INTRODUCTION

Sarcomas are tumors derived from the mesoderm. They are divided into two types: bone and soft tissue sarcomas, which include skin tumors.¹ Soft tissue sarcomas affect one to five individuals/ 100,000/ year, mainly over 55 years old.² Its main subtypes are gastrointestinal, liposarcoma, and leiomyosarcoma.² There are two types of soft tissue sarcomas that most commonly appear on the skin: undifferentiated pleomorphic sarcoma (previously called storiform-pleomorphic malignant fibrous histiocytoma) and myxofibrosarcoma (formerly called myxoid malignant fibrohistiocytoma).¹ Cutaneous sarcomas usually present as skin-colored nodules or subcutaneous masses, with rapid growth, most commonly in the limbs.¹

There is a histological grade (G) for soft tissue sarcomas (FNCLCC-France) in which three parameters receive a score: differentiation (1 to 3), mitotic index (1 to 3), and tumor necrosis (0 to 2). The sum of the numbers results in GX, G1 (2 to 3), G2 (4 to 5), and G3 (6 to 8). The American Cancer Society (ACS) has also established a TNM staging for this group of tumors depending on their topography: head and neck, trunk and extremities, thoracic and abdominal visceral organs, and retroperitoneum.³

Undifferentiated pleomorphic sarcoma (UPS) is the fourth most important soft tissue sarcoma. Regarding its epidemiology, it affects mostly men (two thirds) in relation to women, in the age group of 50 to 70 years (most affected age group).⁴ It is more frequent in the lower limbs, but 10% to 15% occur in the head and neck.¹ Regarding its evolutionary trend, it presents a local recurrence of 19% to 31%, and a metastasis index of 31% to 35% (with a frequency of 90% in the lungs, 85% in the bones, and 1% in the liver). Its five-year survival rate is 65% to 70%. The recommended treatment is surgical with 2 cm margins and adjuvant radiotherapy in selected cases (however, it is difficult to obtain free margins through surgery). For metastatic disease, chemotherapy is indicated (albeit with poor results) with anthracyclines (16% to 27% response) associated or not with ifosfamide. Doxorubicin is palliative. New studies have shown greater benefit with eribulin mesylate.⁵

CASE REPORT

An 88-year-old woman presented a nodular lesion with progressive growth in the left clavicle region for three months. Dermatological examination showed a violet, ulcerated tumor, with friable necrosis, 11 cm in the longest axis, in the left infraclavicular area (Figure 1). Polymorphic vessels characterized the dermoscopy without other specificities (Figure 2). We performed the lesion's incisional biopsy, whose anatomopathological examination revealed infiltration of the deep portion of the sample by large cell neoplasia, with a predominant fusiform shape, with cell nuclei of varying sizes and shapes, with irregular chromatin. Numerous mitosis figures, compatible with fusocellular and pleomorphic sarcoma, infiltrated the reticular dermis, histological grade 3 (Figure 3). The immunohistochemical panel showed a negative result for the expression of all the antigens surveyed (AE1/AE3, p63, 34BetaE12, protein S-100, SOX-100, melan-A, AML, desmin, CD3, and CD34), compatible with undifferentiated sarcoma. After anatomopathological examination, we opted for excision of the lesion with wide surgical margins and closure by primary intent of the surgical wound (Figure 4). We performed screening for lung, lymph node, and abdominal metastases with imaging tests, which was negative. The patient is being followed up, with no signs of local recurrence (Figure 5).

DISCUSSION

For TNM staging of this case, we adopted the American Joint Committee on Cancer (AJCC) 2018 reference for the trunk and extremities' soft tissue sarcomas. Table 1 represents the



FIGURE 1: Clinical image



FIGURE 2: Dermoscopy

T, and Table 2 shows the final staging (note that the G classification described in the introduction is considered). In both tables, the case reported is highlighted.

In the reported case, immunohistochemistry was essential since clinically, and even histopathologically undifferentiated pleomorphic sarcoma makes a differential diagnosis with:

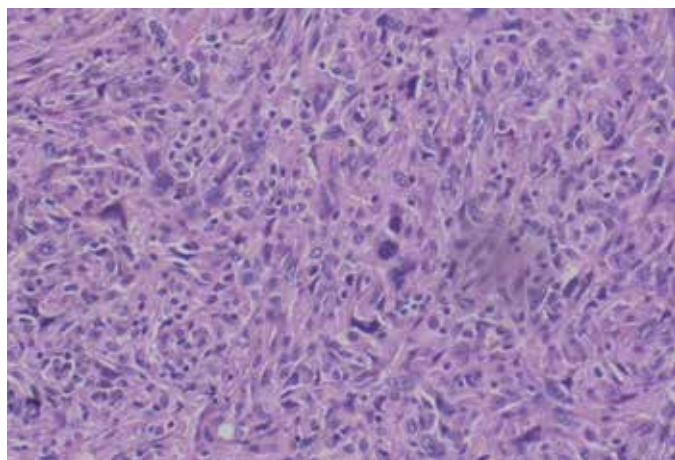


FIGURE 3: Histopathology



FIGURE 4: Wound after exeresis

- Melanoma (desmoplastic; frequent positivity for S100, HMW-MMA; rare positivity for SMA, desmin, factor XIIIa, laminin, and type IV collagen; and generally negative for HMB-45, Gp100, melan A/Mart1, tyrosinase, and transcription factor of microphthalmia).⁶

- Spindle cell squamous cell carcinoma (positive for AE1/AE3, vimentin, and CAM5.2, and vimentin).⁷

- Dermatofibrosarcoma protuberans (positivity for CD34 and vimentin; negativity for neuron-specific enolase, HMB-45, and S100 protein).⁸

- Atypical fibroxanthoma (exclusion diagnosis, negative for cytokeratins to exclude spindle cell carcinoma, spindle cells, S-100, and melanogenic markers such as melan-A and HMB-45 to exclude melanoma; desmin, actin, and H-caldesmon to



FIGURE 5: Final result

exclude leiomyosarcoma; and p63 and p40 to exclude squamous cell carcinoma.⁹

- Leiomyosarcoma (positivity for smooth muscle actin, desmin, and S100).¹⁰

- Undifferentiated pleomorphic sarcoma (positive for vimentin, factor XIIIa, CD68, CD10, CD34, CD 99; and negative for keratin, muscle, and melanocytic markers).¹¹

CONCLUSION

Table 1: Final TNM

T	Values (cm)
1	Until 5
2	5 to 10
3	10 to 15
4	Higher than 15

Table 2: G staging

Staging	TNM and G
IA	T1N0M0G1
IB	T2-4N0M0G1
II	T1N0M0G2-3
IIIA	T2N0M0G2-3
IIIB	T3-4N0M0G2-3
IV	N1M1

The case presented enriches the dermatologist's memory regarding the possible differential diagnoses among the rare mesenchymal tumors. We emphasized how essential the role of histopathology and immunohistochemistry is, reviewing the game

of markers with their diagnostic correlations in this mini-challenge and bringing the opportunity to review the staging and the conduct of soft tissue sarcomas. ●

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