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Malignant Fibrous Histiocytoma

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Abstract: Malignant fibrous histiocytoma is the most common soft tissue sarcoma in adults but is rare as a primary cutaneous tumor. It has a male predilection with a peak incidence between the fifth and sixth decades of life. Malignant fibrous histiocytoma predominantly occurs in the extremities. We report a case of malignant fibrous histiocytoma presenting in the hallux of the right foot.

Keywords: Tumor; Fibrous histiocytoma; Soft tissues

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1. Introduction

Soft tissue sarcomas are rare tumors, accounting for 1% of all malignant tumors in adults. Among them, 70% are located in the extremities (49% in the lower extremities, and 19% in the upper extremities), while the remaining 30% are located in the trunk, abdomen, or retroperitoneum. Malignant fibrous histiocytoma (MFH) is a histological subtype of sarcoma and is considered the most common adult soft tissue sarcoma [1,2]. It was first described in 1964 by O'Brian and Stout. MFH commonly occurs in men over 50 years of age, and the incidence ratio of men to women is 2:1.1 [2]. Its origin is uncertain. Clinically, it manifests as a multilobulated tumor, with areas of hemorrhage and necrosis, similar to other high-grade sarcomas [3-5]. It is difficult to distinguish MFH from other sarcomas and carcinomas histologically. Hence, immunohistochemistry techniques are used for definitive diagnosis. The most important prognostic factors are age, depth of the lesion, histological variant, location, size, and the presence or absence of metastases [2]. In general, it is a tumor with a poor prognosis due to its high aggressiveness and recurrence rate [6].

The treatment of choice for MFH is surgery. Complete excision with oncological surgical margins and adjuvant radiotherapy is the most accepted combination [6].

2. Case

A 78-year-old Argentinean male with a medical history of hypertension consulted our department for a painful lesion in the right hallux that had been present for a year. He reported an increase in size in the past 3 months. Otherwise, he had no relevant family history.

On dermatological examination, an erythematous-violaceous, foul-smelling lesion measuring 3 × 4 cm was observed, with hemorrhagic and necrotic areas and raised infiltrative border extending from the dorsal aspect of the distal phalanx toward the plantar aspect of the right hallux (**Figures 1**–**4**).

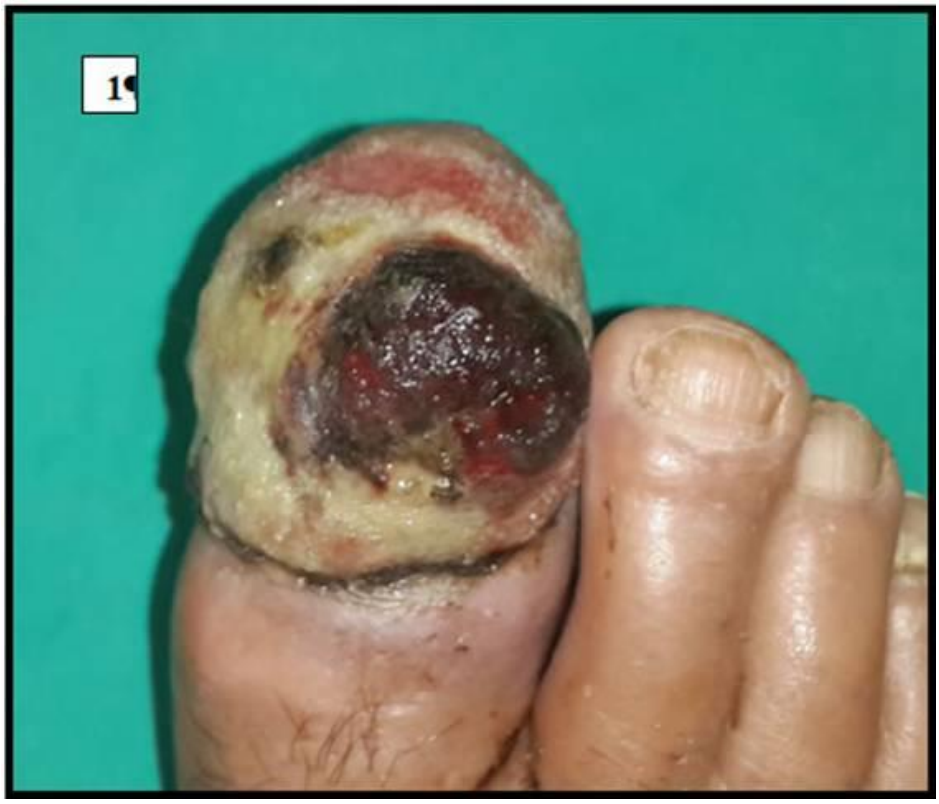

Figure 1. Angiomatous lesion of the hallux of the right foot

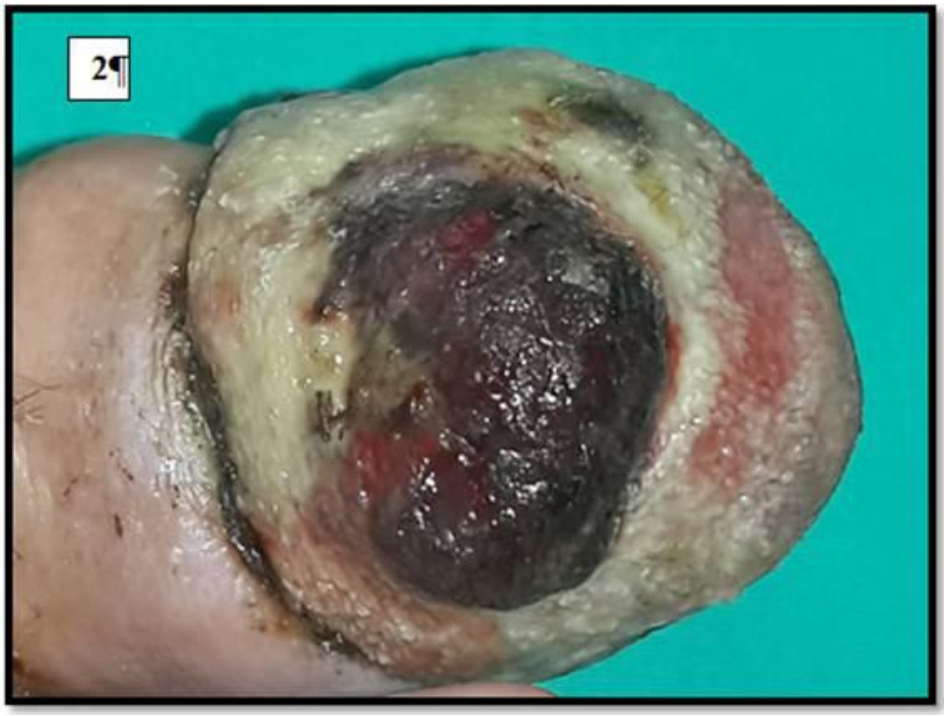

Figure 2. Erosive lesion with purulent discharge on the right hallux

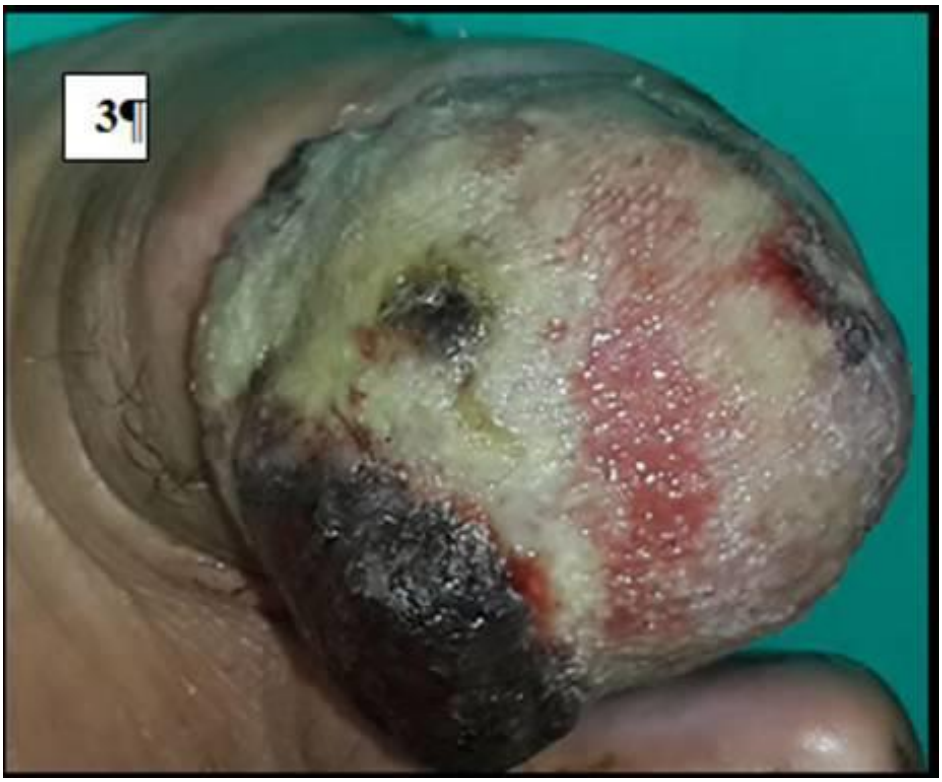

Figure 3. Raised infiltrative edge extending to the plantar aspect of the right hallux

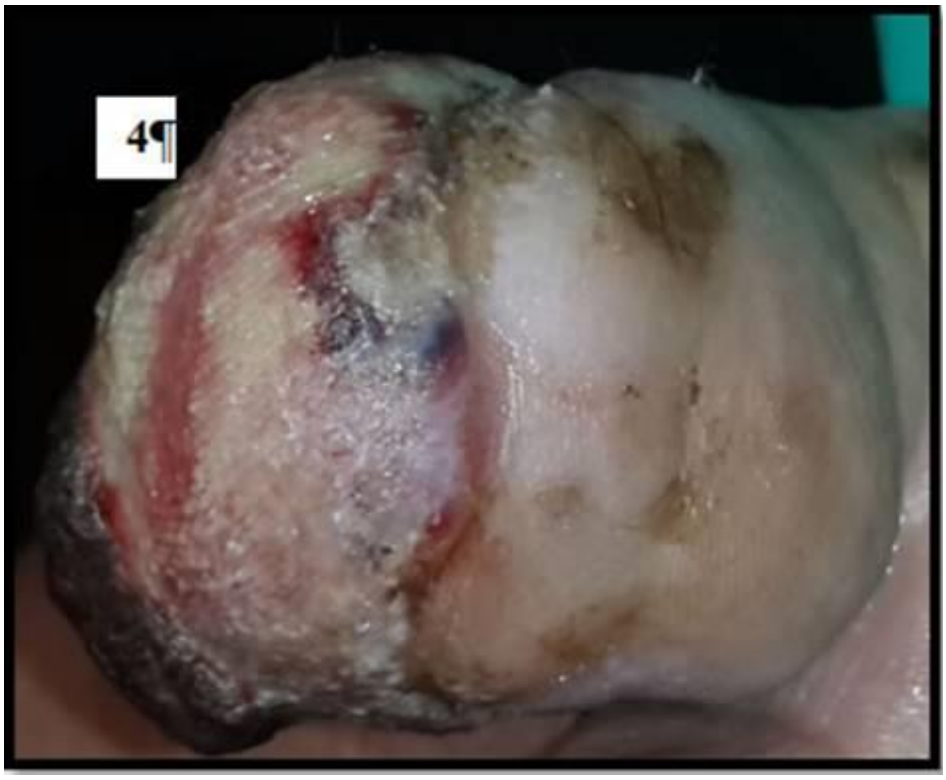

Figure 4. Raised infiltrative edge extending to the plantar aspect of the right hallux

The skin biopsy performed by our department was reported as inconclusive, so we decided to consult the Traumatology Department. A foot X-ray was requested, which revealed an osteolytic image in the right hallux. Due to the size of the tumor and its macroscopic characteristics (infiltrated edges, invasive tumor), we decided, in conjunction with the Traumatology Department, to perform tumor resection with amputation

of the hallux and a safety margin up to the middle 1/3 of the first metatarsal for histopathological study of the specimen.

The pathological report was as follows: proliferation of spindle-shaped or tapered cells arranged in thin fascicles with a swirling arrangement in some areas on microscopic examination with hematoxylin and eosin (H&E) staining. In addition, it showed positive immunohistochemistry staining for Vimentin and CD68. The histological image was consistent with MFH (**Figures 5**–**7**).

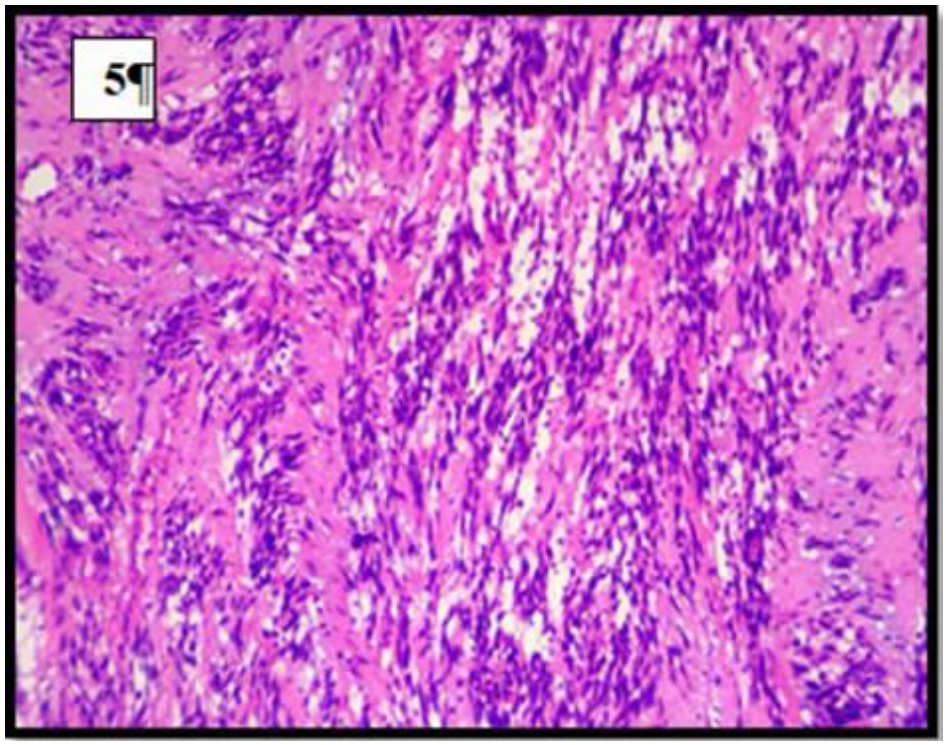

Figure 5. Proliferation of spindle cells arranged in thin fascicles on hematoxylin and eosin staining

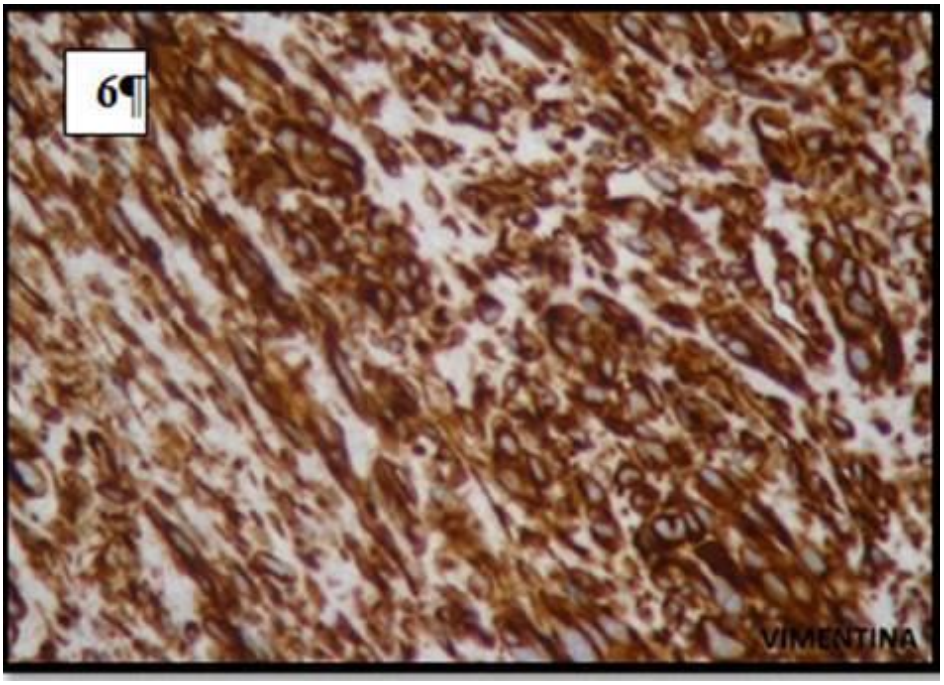

Figure 6. Vimentin-positive on immunohistochemistry staining

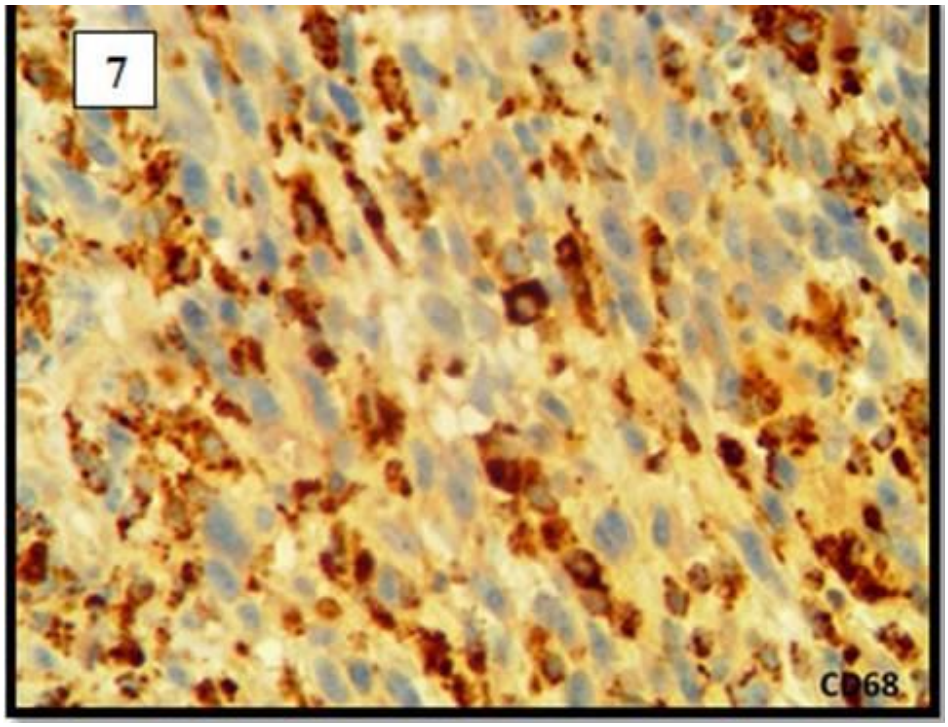

Figure 7. CD68-positive on immunohistochemistry staining

In view of these results, the patient was referred to the Oncology Department for follow-up and treatment. Computed tomography (CT) of the chest, abdomen, and pelvis was performed, showing no evidence of distant disease. The surgical treatment was considered sufficient and strict follow-up was indicated. The patient did not show up for subsequent check-ups.

3. Discussion

MFH is the most common soft tissue sarcoma in adulthood [4,6]. It is predominant in males, with a male to female ratio of 2:1, and after the fifth decade of life. However, there have been cases reported in adolescents and young adults [1,4]. In 70%–75% of cases, extremities are involved, with the lower limbs being the most affected. Other locations include the trunk and retroperitoneum [2]. MHFs often arise in previously irradiated areas, such as the head, neck, and breast [6], and the tumors typically arise in deep fascia and skeletal muscle. MFH was described by O'Brien and Stouten 1964. Since then, there has been controversy around the origin of the tumor cells. Several researchers have suggested that it originates in primitive mesenchymal stem cell with differentiation into fibroblasts and histiocytes [5].

3.1. Diagnosis and classification

Classically, MHF has been a diagnostic challenge due to its clinical and histopathological diversity. The advent of immunohistochemistry techniques has made it possible to precisely delimit the diagnosis of this pathology and to avoid overdiagnosis due to its similarity to other undifferentiated tumors. The diagnostic classification of MFH has been modified and expanded since its first description in the 1960s [1].

In 2002, the World Health Organization (WHO) redefined it within the malignant fibrous tumors of bone and soft tissue as undifferentiated pleomorphic sarcoma, which can present as three tumor phenotypes: (i) high-grade undifferentiated pleomorphic (or storiform) sarcoma; (ii) undifferentiated pleomorphic sarcoma with giant cells; and (iii) inflammatory undifferentiated pleomorphic sarcoma [1,3,9].

In the 2020 update, it is classified as a tumor of uncertain differentiation and defined as an undifferentiated sarcoma.

The diagnosis of MFH is based on clinical and histopathological findings [6,8]. Clinically, it usually presents as a rapidly growing tumor, initially palpable as a subcutaneous tumor without skin involvement. In more advanced cases, it may invade the skin, manifesting as a multilobulated, infiltrative tumor, with areas of necrosis and hemorrhage. Its histopathological features include the presence of pleomorphic spindle cells that infiltrate deep into the dermis and may be accompanied by necrotic tissue and infiltration of lymphoid cells [1,2,6].

As mentioned above, due to their wide pleomorphism, they are often mistaken for other mesenchymal tumors (liposarcomas and fibrosarcomas). Hence, immunohistochemical studies must be performed for a definitive diagnosis. A number of studies have shown that this tumor is positive for Vimentin, actin, CD68, alpha-1 antitrypsin, and alpha 1-antichymotrypsin [1,5,7,9].

3.2. Differential diagnoses

Its differential diagnoses include other variants of soft tissue sarcomas, such as liposarcoma, myxofibrosarcoma, rhabdomyosarcoma, and synovial sarcoma, in addition to dermatofibrosarcoma protuberans, atypical fibroxanthoma, and amelanotic melanoma [5,7,9,10].

3.3. Treatment

The treatment of choice for MFH is surgery with wide surgical margin confirmed by the pathologist. The success of surgery, in many cases, depends on the location of the tumor and its proximity to vital organs. For local recurrences, the treatment of choice is also surgery.

Adjuvant radiotherapy is useful in cases of tumors adjacent to important anatomical structures, in which surgical resection does not achieve adequate surgical margins [6,8].

The use of chemotherapy in the treatment of MHF is controversial, as no significant differences have been observed in the outcomes of patients treated with chemotherapy. Further studies are needed to evaluate the aforementioned therapeutic options [1,6,8,9].

3.4. Evolution and prognosis

MHF is an aggressive tumor, with high degree of malignancy and local recurrence despite the treatments mentioned above [3].

Local recurrence is more frequent in tumors more than 5 cm in size and in cases where surgical margins are inadequate [8].

The most common sites of distant metastases are the lungs, followed by lymph nodes, liver, bone, and brain [1,5].

Factors associated with poor prognosis include positive surgical margins, head and neck involvement, tumor size more than 5 cm, deep-seated tumors, and advanced stage at diagnosis [8-10].

4. Conclusion

The interest of this case lies in the presentation of an uncommon pathology as a primary cutaneous lesion and in an unusual location such as the acral region. Given the diagnostic, clinical, and histopathological challenges of this entity, we resorted to interconsultations with the Traumatology Department and Oncology Department for appropriate multidisciplinary management.

Disclosure statement

The authors declare no conflict of interest.

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