

## Fibrosarcoma Presenting as Marjolin Ulcer – A Rare Case Report

Dr. Agam Hans<sup>1\*</sup>, Dr. Maitrayee Roy<sup>2</sup>, Dr. Vijay S. Nijhawan<sup>3</sup>

<sup>1</sup>Junior Resident, Department of Pathology, MMIMSR, Mullana, Ambala, Haryana, India

<sup>2</sup>Associate Professor, Department of Pathology, MMIMSR, Mullana, Ambala, Haryana, India

<sup>3</sup>Professor & Head, Department of Pathology, MMIMSR, Mullana, Ambala, Haryana, India

DOI: [10.36348/sjpm.2023.v08i08.003](https://doi.org/10.36348/sjpm.2023.v08i08.003)

Received: 02.07.2023 | Accepted: 05.08.2023 | Published: 08.08.2023

\*Corresponding author: Dr. Agam Hans

Junior Resident, Department of Pathology, MMIMSR, Mullana, Ambala, Haryana, India

### Abstract

Marjolin ulcer is a rare malignancy that arises in previously traumatised or chronically inflamed skin, particularly after burns, with an average latency period of 36 years. Nearly 75-90% marjolin ulcers are diagnosed as squamous cell carcinoma but other neoplasms like basal cell carcinoma, melanoma, fibrosarcoma etc have also been rarely reported. In this case, a 52-year-old male presented with a painless bleeding mass over lower chest wall rapidly increasing in size from last 2 years. He had suffered burn at the same site 20 years ago. Biopsy was reported as spindle cell neoplasm. Wide local excision revealed a poorly circumscribed subcutaneous spindle cell neoplasm exhibiting variable cellularity, fascicular to storiform architecture, mild to moderate nuclear atypia and scattered gaping thin-walled blood vessels. The neoplastic cells showed diffuse strong staining with CD34 in the less cellular area and patchy positivity in the cellular more mitotically active areas. The tumor cells were immunonegative for EMA, p63, SMA, CD99 and STAT6. The final diagnosis rendered was Dermatofibrosarcoma Protuberans dedifferentiating in to fibrosarcoma. Although sarcoma presenting as a Marjolin ulcer is exceedingly rare, it should be kept in mind in differential diagnosis of spindle cell malignancy arising at a burn site.

**Keywords:** Marjolin ulcer; Fibrosarcoma, dermatofibrosarcoma protuberans; spindle cell neoplasm.

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### INTRODUCTION

Marjolin ulcer is a malignancy that arises in a previously traumatised or chronically inflamed skin with an average latency period for malignant transformation being 36 years [1]. Malignant transformation of an ulcer is most commonly associated with burns, but it has also been reported in many other types of non healing wounds, such as traumatic wounds, osteomyelitis, pressure sores, venous stasis ulcers, fistulas and chronic trophic ulcers in leprosy [2]. The most common site for these lesions are the extremities (~60%), followed by head and face (~30%) whereas the least common site is the trunk (~10%) [1]. Histopathologically, approximately 75-90% cases are squamous cell carcinoma (SCC). However other neoplasms such as basal cell carcinoma (BCC), melanoma and soft tissue sarcomas have also been rarely reported [3].

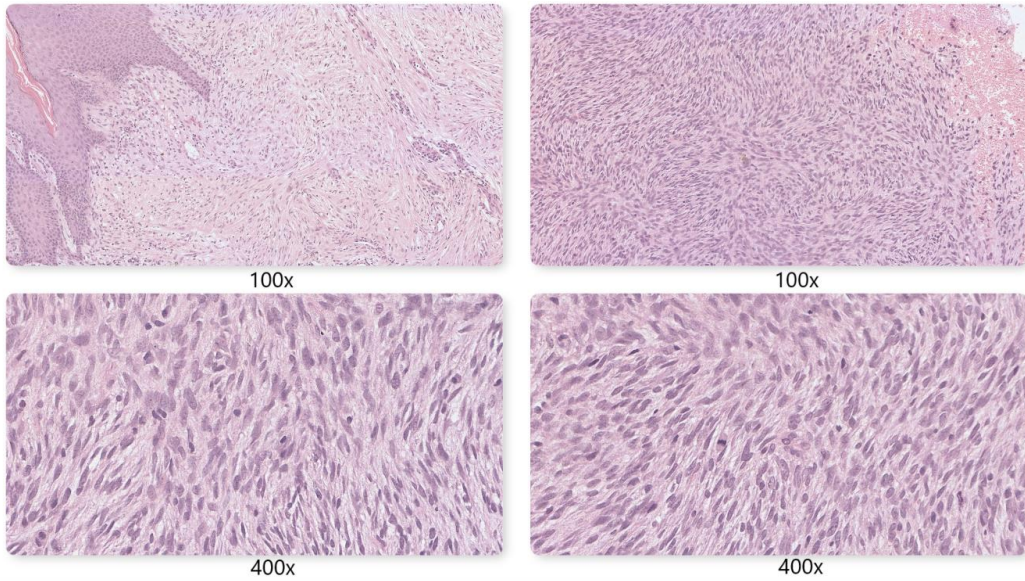
### CASE PRESENTATION

A 52-year-old male presented with a painless bleeding mass over lower anterior chest wall since two

years. The growth was non-tender but rapidly increasing in size. He had suffered burn at the same site 20 years ago. Punch biopsy done was reported as malignant spindle cell tumor (Figure 1). He underwent excision with split skin graft. A wide local excision specimen of fungating mass with overlying ulceration at multiple foci was received. The tumor measured 12x11x8 cm and was whitish, poorly circumscribed with epicentre in the subcutaneous plane (Figure 2). Microscopy revealed a poorly circumscribed spindle cell neoplasm involving the subcutaneous and dermis. The tumor was variably cellular, exhibit fascicular to storiform architecture, spindled nuclei with mild to moderate anisonucleosis, vesicular chromatin and 10 to 12 mitoses per 10 high power fields. Many gaping thin-walled blood vessels were present throughout the neoplasm (Figure 3). The resection margins were uninvolved. The neoplastic cells stained positive with CD34; the staining was diffuse and strong in the less cellular area and focal in the cellular more mitotically active areas. These cells were immunonegative for EMA, p63, SMA, CD99, S100 and STAT6 (Figure 4).

The final diagnosis rendered was dermatofibrosarcoma

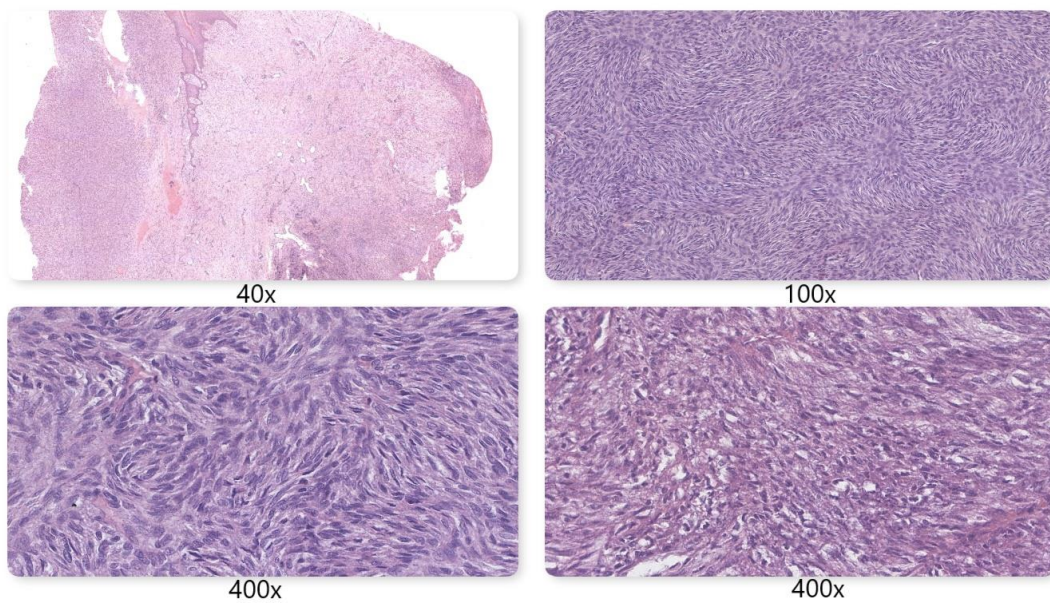
protuberans (DFSP) dedifferentiating into fibrosarcoma.



**Figure 1:** Punch biopsy showing A: Variably cellular spindle cell tumor in the dermis (H&E, 40x); B: Fascicular to storiform architecture (H&E, 100x); C & D: spindled nuclei with mild to moderate nuclear atypia, vesicular chromatin and moderate eosinophilic cytoplasm (H&E, 400x)

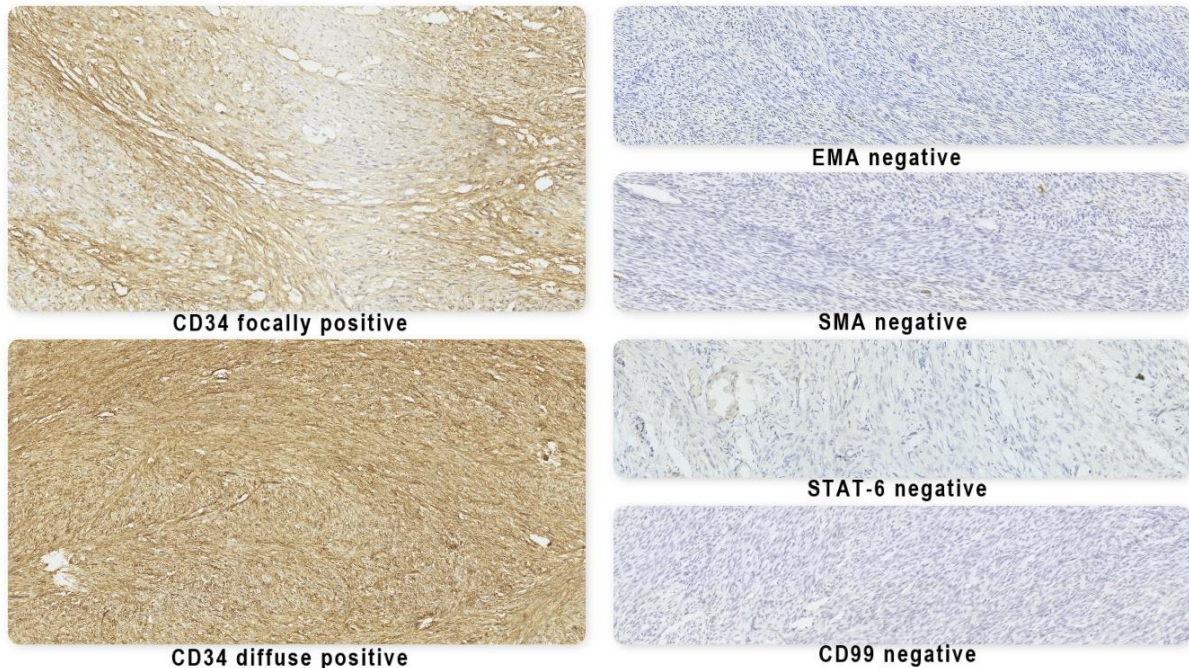


**Figure 2:** A: Mass at prior burn site in lower anterior chest wall; B: Wide local excision of fungating tumor mass





**Figure 3: Wide local excision showing A: spindle cell neoplasm in dermis and subcutis (H&E, 40x); B: Fascicular to storiform architecture (H&E, 100x); C & D: tumor cells with spindled nuclei with mild to moderate anisonucleosis, vesicular chromatin and brisk mitosis. (H&E, 400x)**



**Figure 4: Immunohistochemistry: Tumor cells are A, B: CD34 positive (diffuse in DFSP area and focal in fibrosarcoma differentiation area) (CD34, 100x); C: EMA negative (EMA, 100x); SMA negative (SMA, 100x); STAT-6 negative (STAT-6, 100x); CD99 negative (CD99, 100x)**

## DISCUSSION

Marjolin ulcer is a malignant degeneration arising from a chronically inflamed or traumatised skin usually after burns with incidence of around 2% in all burn scars [3, 4]. Various etiological factors have been suggested for malignant degeneration in burn scars. The loss of immune cells in epidermis along with loss of epithelial function (inhibition of claudins and cadherins) causes malignant cells to evade the immune system and cause metastasis. Increase in the mesenchymal markers like fibronectin, vimentin, laminin-4 have also been observed in cells of chronic burn scars. Although the most common cause for malignant transformation in Marjolin ulcer is burn scars, but non healing wounds, such as traumatic wounds, osteomyelitis, pressure sores, venous stasis ulcers, fistulas and chronic trophic ulcers in leprosy have also been reported in various articles across literature. Scarring from lupus, amputation stumps, frostbite, vaccination sites, skin graft donor sites, scars, urinary fistulas, and radiation have also been linked to Marjolin ulcer [2, 4, 5]. In most cases, the Marjolin Ulcer presents as a Squamous cell Carcinoma, followed by BCC, melanoma, sarcoma, squamobasal cell carcinoma but also a variety of rare tumours may also emerge in the chronic scars including fibrosarcoma, liposarcoma, dermatofibrosarcoma protuberans, and mesenchymal tumors [5-7]. Lower limbs constitute the most frequent site of Marjolin ulcers followed by head

and neck region, upper limbs and other body parts including thorax [5-7]. Based on the latency period of malignant transformation, Marjolin Ulcer can be divided into acute and chronic forms. Acute onset malignancies are referred to those malignancies that arise within one year of trauma whereas malignancies that arise after a latent period of more than one year are referred to as chronic forms of Marjolin Ulcer [3,4]. Post burn sarcomas and post burn carcinomas have a similar latency period of around 35 years, but acute onset malignancy has not been reported in post burn sarcomas. The earliest latent period described in literature for post burn sarcomas is 3 years [8]. In this case the microscopy of the lesion showed a well circumscribed variably cellular tumor with fascicular to storiform architecture having spindled nuclei with 10-12 mitosis/10HPF. The diagnosis was rendered as Malignant Spindle cell lesion. A panel of IHC markers was ordered for definitive characterization of tumor. The panel consisted of EMA, p63, SMA, CD99, S100 and STAT6 and CD34. The tumor cells were immunonegative for EMA, p63, SMA, CD99, S100 and STAT6 which ruled out synovial sarcoma, cutaneous spindle cell SCC, leiomyosarcoma, malignant peripheral nerve sheath tumor (MPNST) and solitary fibrous tumor (SFT). While CD34 was diffuse and strong positive in tumor cells in less cellular areas and focally positive in areas having high mitotic activity. So, the final diagnosis rendered was dermatofibrosarcoma protuberans (DFSP)

dedifferentiating into fibrosarcoma. The site of the lesion i.e., anterior thoracic wall as well as histopathologically Marjolin ulcer presenting as DFSP dedifferentiating into fibrosarcoma in a chronic scar makes this case unusual.

## CONCLUSION

Although sarcoma presenting as Marjolin ulcer is exceedingly rare, it should be kept in mind in differential diagnosis of spindle cell malignancy arising at a burn site. Amongst sarcomas, fibrosarcoma is the most common type of sarcoma in a scar site probably due to abundance of fibroblasts in the skin scar tissues [9].

## REFERENCES

1. Chalya, P. L., Mabula, J. B., Rambau, P., Mchembe, M. D., Kahima, K. J., Chandika, A. B., ... & Gilyoma, J. M. (2012). Marjolin's ulcers at a university teaching hospital in Northwestern Tanzania: a retrospective review of 56 cases. *World journal of surgical oncology*, 10(1), 1-8.
2. Pavlovic, S., Wiley, E., Guzman, G., Morris, D., & Braniecki, M. (2011). Marjolin ulcer: an overlooked entity. *International Wound Journal*, 8(4), 419-424.
3. Oruç, M., Kankaya, Y., Sungur, N., Özer, K., Işık, V. M., Ulusoy, M. G., ... & Koçer, U. (2017). Clinicopathological evaluation of Marjolin ulcers over two decades. *The Kaohsiung Journal of Medical Sciences*, 33(7), 327-333.
4. Bazaliński, D., Przybek-Mita, J., Barańska, B., & Więch, P. (2017). Marjolin's ulcer in chronic wounds—review of available literature. *Contemporary Oncology/Współczesna Onkologia*, 21(3), 197-202. doi: 10.5114/wo.2017.70109.
5. Ochenduszkiewicz, U., Matkowski, R., Szynglarewicz, B., & Kornafel, J. (2006). Marjolin's ulcer: malignant neoplasm arising in scars. *Reports of Practical Oncology and Radiotherapy*, 11(3), 135-138.
6. Gamatsi, I. E., Bailie, F. B., Srinivasan, J. R., & McCulloch, T. A. (2000). Malignant melanoma in a skin graft: burn scar neoplasm or a transferred melanoma?. *British journal of plastic surgery*, 53(4), 342-344.
7. Kowal-Vern, A., & Criswell, B. K. (2005). Burn scar neoplasms: a literature review and statistical analysis. *Burns*, 31(4), 403-413.
8. Çeliköz, B., Demiriz, M., & Selmanpakoğlu, N. (1997). A shorter lag period of mesenchymal malignancy on Marjolin's ulcer. *Burns*, 23(1), 72-74.
9. Fazeli, M. S., Lebaschi, A. H., Hajirostam, M., & Keramati, M. R. (2013). Marjolin's ulcer: clinical and pathologic features of 83 cases and review of literature. *Medical journal of the Islamic Republic of Iran*, 27(4), 215-224.