



DEPARTMENT OF PATHOLOGY

## Case of the Week

### Thoracic Pathology: Sarcomatoid squamous carcinoma arising in Marjolin's ulcer

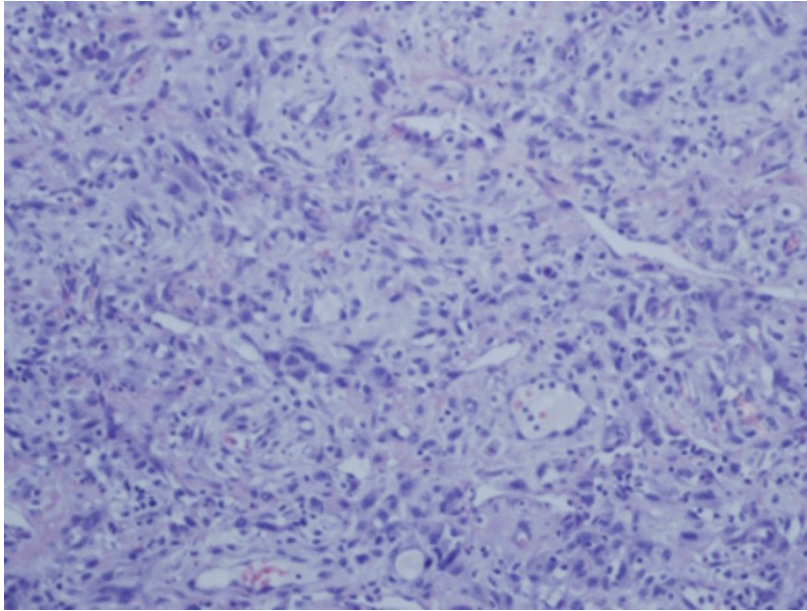
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November 24, 2015

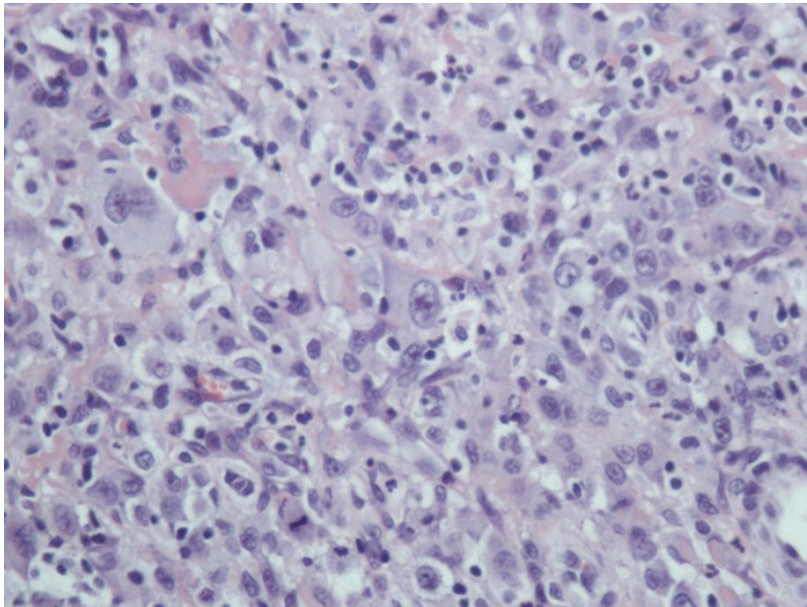
#### History

75 year old female with past history of pulmonary tuberculosis diagnosed 20 years previously (complicated by empyema, status post left pneumonectomy and open pleural drainage with Eloesser flap thoracostomy window), now presents with fever, exertional chest pain and WBC count 16000. She had multiple biopsies of the Eloesser flap over the last 19 years revealing granulation tissue but was lost to follow up in the past 4 years. Chest CT revealed soft tissue thickening in the Eloesser flap area extending into the mediastinal cavity, measuring 8 x 4 cm. Multiple biopsies from the Eloesser flap and chest cavity soft tissue thickening were obtained.

**Mediastinal Biopsy:**



*Figure 1. 200x magnification*



*Figure 2. 400x magnification*

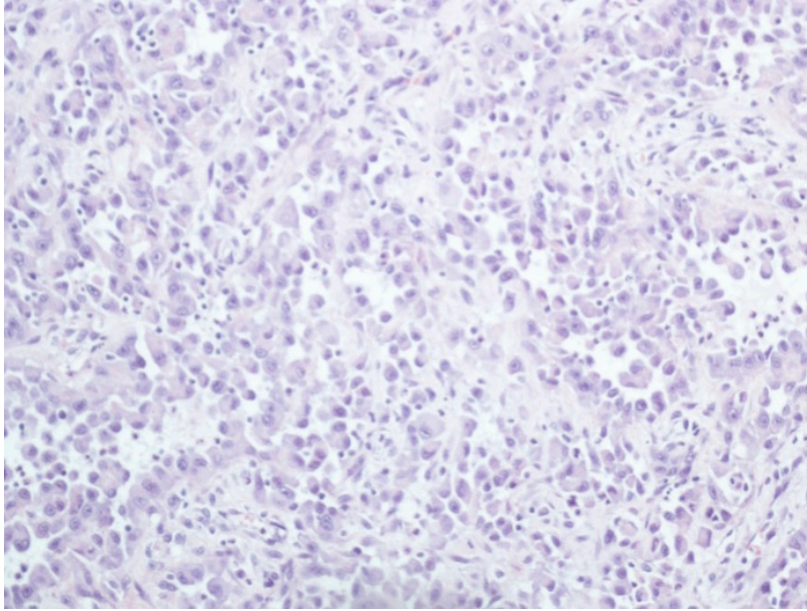


Figure 3. 200x magnification

### Figure 1, 2 and 3

*Figures 1-3.* Fig. 1: H&E stain, 200x magnification: Diffuse sheets of polygonal to spindle cells interspersed by inflammatory cells. Fig. 2: H&E stain, 400x magnification: Sheets of highly pleomorphic cells with abundant cytoplasm and brisk mitotic activity. Fig. 3: H&E stain, 200x magnification: Area with pleomorphic cells arranged in gland or slit like architecture with hobnailing.

### Immunohistochemical Stains:

Pleomorphic cells were diffusely positive for vimentin, CAM5.2 and p63. Immunohistochemical stains for pancytokeratin AE1/AE3, EMA, desmin, smooth muscle actin, myogenin, HMB45, S-100, calretinin and CD34 demonstrated negative expression.

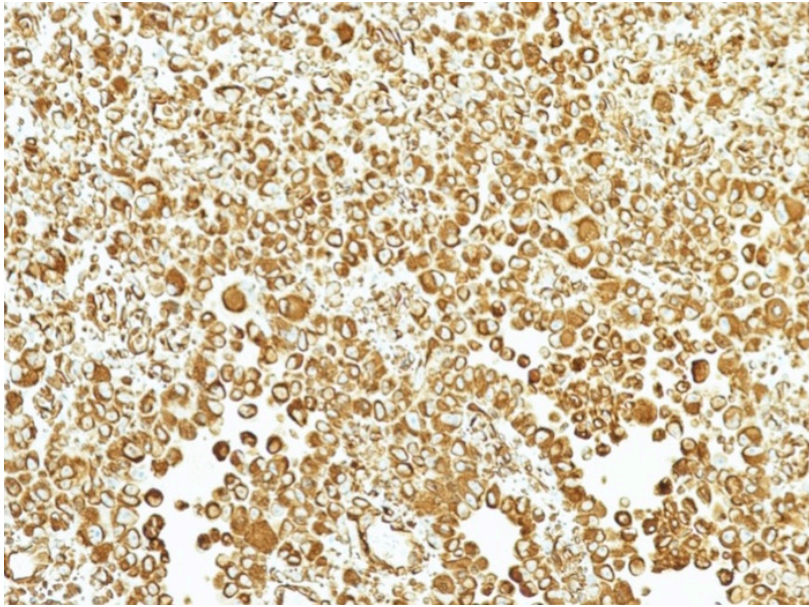


Figure 4. Vimentin

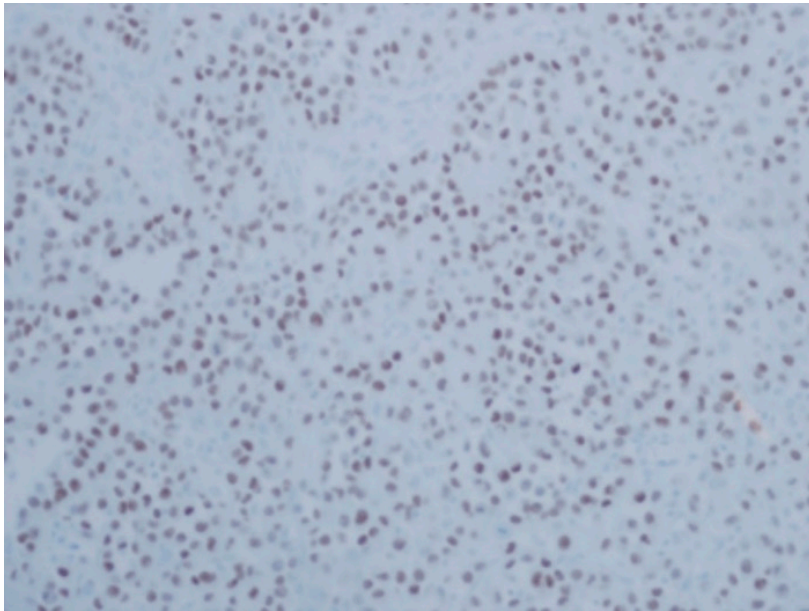


Figure5. p63

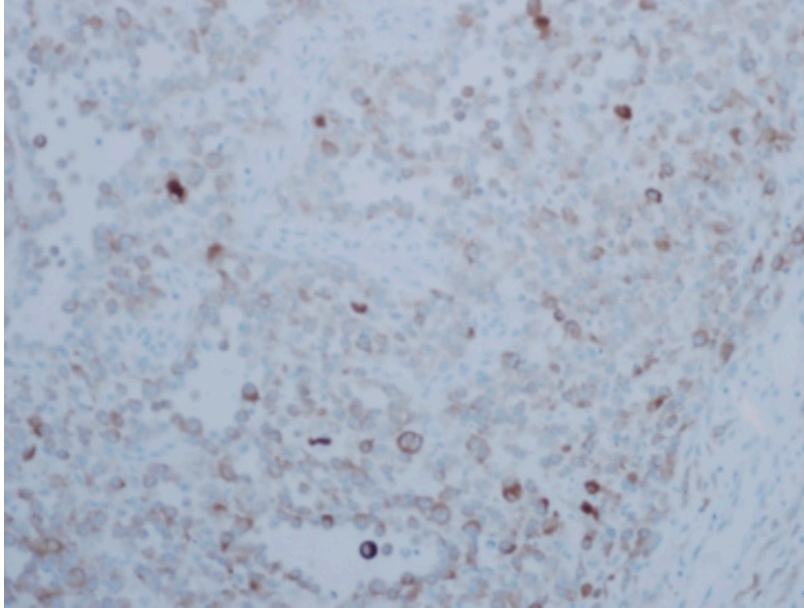


Figure 6. Cam5.2

#### **Figure 4-6**

*Figures 4-6.* Fig. 4: Vimentin Immunohistochemistry stain, 400x magnification: Diffuse, strong cytoplasmic staining. Fig. 5: p63 Immunohistochemistry stain, 200x magnification: Positive nuclear expression. Fig.6: Cam5.2 Immunohistochemistry stain, 200X magnification: Diffuse cytoplasmic staining.

Biopsy from the area closer to the eloesser flap showed invasive squamous carcinoma along with atypical spindled and polygonal cells (Fig.7). Multiple other biopsies showed granulation tissue with scattered clusters of atypical spindle and polygonal cells.

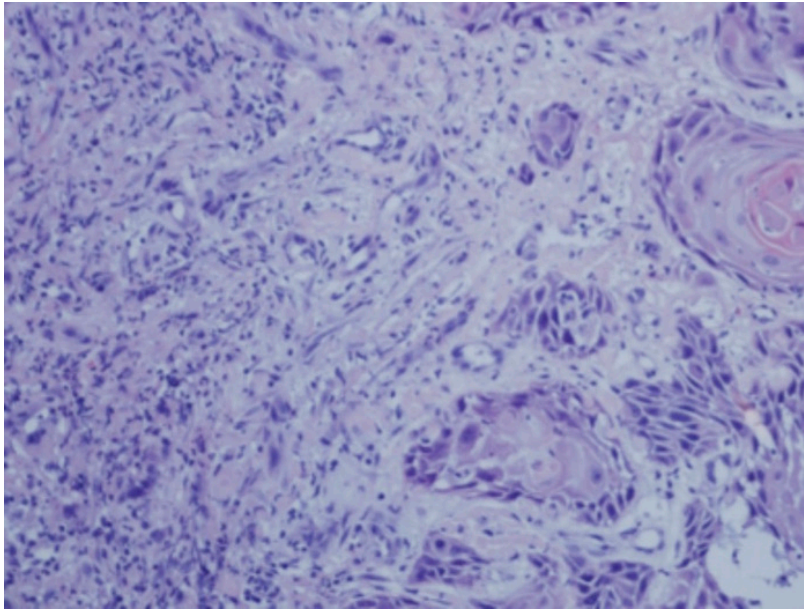


Figure 7.

## Diagnosis

Sarcomatoid squamous carcinoma arising in Marjolin's ulcer

## Discussion

Marjolin's ulcer (MU) is a term that can be used to include squamous cell carcinoma (SCC), basal cell carcinoma (BCC) and malignant melanoma (MM) that develop in chronic wounds, first described in the 1st century AD by Aurelius Cornelius Celsus. The term can also be used to describe malignant degeneration in burn scars, chronic venous insufficiency ulcers, pressure ulcers, vaccination sites, urinary fistulas, frostbite, snakebites, osteomyelitis, pilonidal abscesses, hydradenitis suppuritiva, herpes zoster, skin graft donor sites, knife wounds and gunshot wounds.<sup>1</sup> The most commonly involved sites are the feet (49.4%), hands (10.8%) and thigh (4.8%). Patients often present during cycles of non-healing ulcers, followed by skin rupturing, bleeding, itching and scratching, severe pain, discharge and foul odor. The mean latency interval for Marjolin's Ulcer-SCC is reported to be up to 32 years, with an average presentation at 52 years old. Marjolin's Ulcer-SCC has a male gender predominance (3:1), while non-MU-SCC do not (1.1-1.7:1). Marjolin's Ulcer-SCCs compared to non-Marjolin's Ulcer-SCCs are more aggressive and have a higher recurrence rate, higher metastatic rate (27.5-40% v 2-23% respectively) and poorer 5 year survival (43-58% v 61.5% in poorly-differentiated and 94.6% in well-differentiated non-MU-SCC). The standard treatment of Marjolin's Ulcers involves a wide local excision, with 2-4cm free resection margin compared to 4-6mm in non-Marjolin's Ulcer-SCCs.<sup>2</sup>

Sarcomatoid carcinomas are poorly differentiated epithelial malignancies. These carcinomas are thought to have derived from primary epithelial tumors which undergo divergent mesenchymal differentiation as they develop. The sarcomatoid components can show osteoblastic, chondroid, or skeletal or smooth muscle differentiation.<sup>3</sup> They most commonly arise in the head and neck, lung and bladder. They can have variable morphology ranging from spindled with storiform pattern to marked epithelioid with single cell infiltration in collagenous matrix.<sup>4</sup>

While some cases may have obvious areas of traditional in-situ or invasive carcinoma, others with only pleomorphic or spindled malignant cells may be difficult to distinguish from true sarcoma, melanoma or even reactive spindle cell lesions, particularly on light microscopy in small biopsy specimens.

Immunohistochemistry for markers of epithelial differentiation can be critical for making the diagnosis, especially on biopsy specimens. Traditional markers for epithelial neoplasms, such as cytokeratin and epithelial membrane antigen (EMA) may be lost in sarcomatoid carcinomas, and have been shown to only be positive in 29% and 47% of cases respectively. Staining for p63 however, is retained in most cases of sarcomatoid carcinomas (63%) including those that have lost expression of other epithelial markers like EMA and pancytokeratin.<sup>4</sup> Sarcomatoid carcinoma is usually positive for vimentin.<sup>4,5</sup>

## References

1. Iqbal FM, et al. Marjolin's ulcer: a rare entity with a call for early diagnosis. *BMJ Case Rep* (2015). doi: 10.1136/bcr-2014-208176
2. Fazeli MS, et al. Marjolin's ulcer: clinical and pathologic features of 83 cases and review of literature. *Medical Journal of the Islamic Republic of Iran* (2013). Vol. 27, No. 4, 215-224.
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4. Viswanathan S et al. Sarcomatoid ( Spindle cell) carcinoma of the head and neck mucosal region: A clinicopathologic Review of 103 cases from a tertiary referral cancer centre. *Head Neck Pathol*. 2010 Dec;4(4):265-275
5. Lewis Jr JS, et al. Alternative epithelial markers in sarcomatoid carcinomas of the head and neck, lung, and bladder – p63, MOC-31, and TTF-1. *Modern Pathology* (2005) 18, 1471-1481.