2021 Spring Annual Meeting Abstract Submission

The Bengal You Haven't Heard Of: Pseudomyogenic Epitheliod Sarcoma-Like Hemangioendothelioma

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A 28 year old male with no medical history presented to the dermatology clinic with growing, painful bumps on the left upper abdomen for 1 year. These lesions were initially thought to be folliculitis by the primary physician and were treated with oral clindamycin to no avail. He has no personal or family history of skin cancer or other dermatoses. His review of systems was pan-negative.

Physical exam revealed three, small, tender flesh colored papules with underlying induration on the left upper abdomen. Only one lesion was readily visible on exam (Figure 1).

A differential diagnosis of painful skin nodules was raised (i.e. BENGAL): blue nevus, eccrine spiradenoma, neuroma, glomus tumor, angiolipoma, or leiomyoma. An 8 mm punch biopsy was performed on the largest available lesion.

Pathology (Figure 2) showed plump spindled and rhabdomyoblastic-like epithelioid cells arranged in fascicles and extending to deep margins. Cells were CD34 negative and Erg positive. Outside immunohistochemistry staining at Mayo Clinic showed FOS B positivity(1), confirming a diagnosis of pseudomyogenic epitheliod sarcoma-like hemangioendothelioma (PMHE).

The patient had CT /MRI imaging which localized the tumor to the soft tissue without underlying muscular involvement. Subsequent 2 cm margin wide local excision was performed by surgical oncology with simultaneous excisional biopsies of other two nodules. Results are pending at this time.

PMHE is an indolent, low-grade sarcoma which often presents as multi-centric painful nodules in young men(2). Recurrence rates are high after WLE(3). Less than 200 cases have been reported worldwide.

1. Alegría-Landa V, Santonja C, Jo-Velasco M, Kutzner H, Requena L. Cutaneous pseudomyogenic (epithelioid sarcoma-like) haemangioendothelioma FOSB immunohistochemistry demonstrating the *SERPINE1-FOSB* fusion gene. J Eur Acad Dermatology Venereol. 2017 Dec;31(12).

2. Hornick JL, Fletcher CDM. Pseudomyogenic Hemangioendothelioma: A Distinctive, Often Multicentric Tumor With Indolent Behavior [Internet]. Vol. 35, Am J Surg Pathol. 2011. Available from: www.ajsp.com

3. Requena L, Santonja C, Luis Martinez-Amo J, Saus C, Kutzner H. Cutaneous Epithelioid Sarcomalike (Pseudomyogenic) Hemangioendothelioma A Little-Known Low-Grade Cutaneous Vascular Neoplasm [Internet]. Vol. 149, JAMA Dermatol. 2013. Available from: https://jamanetwork.com/

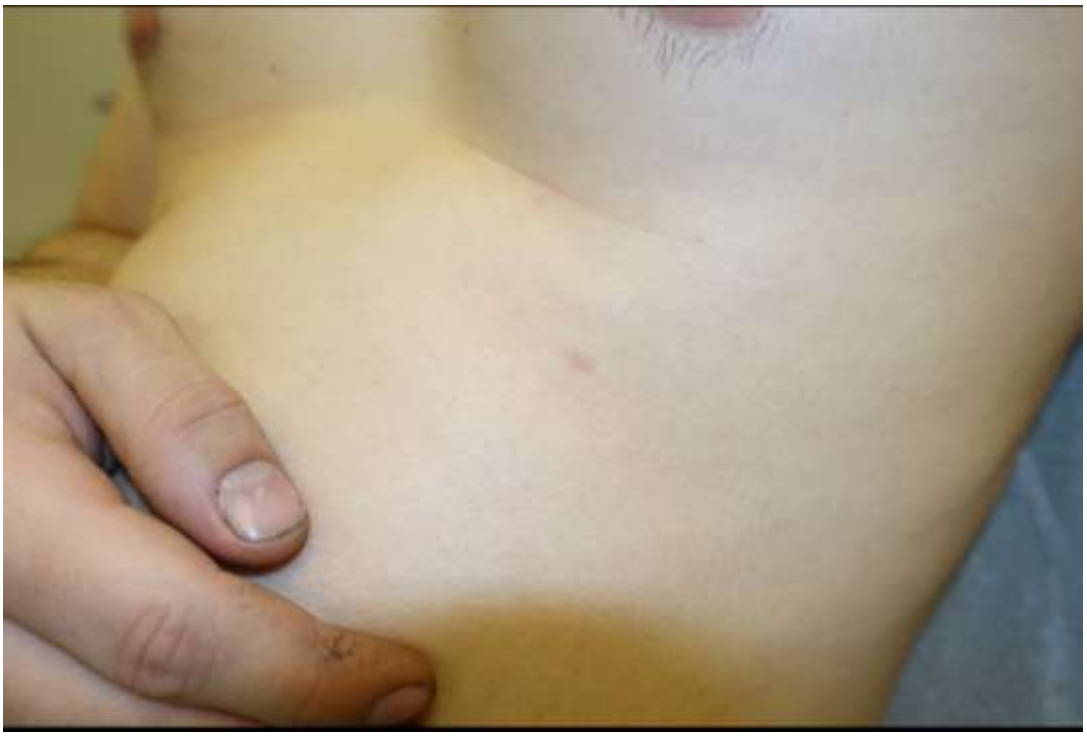
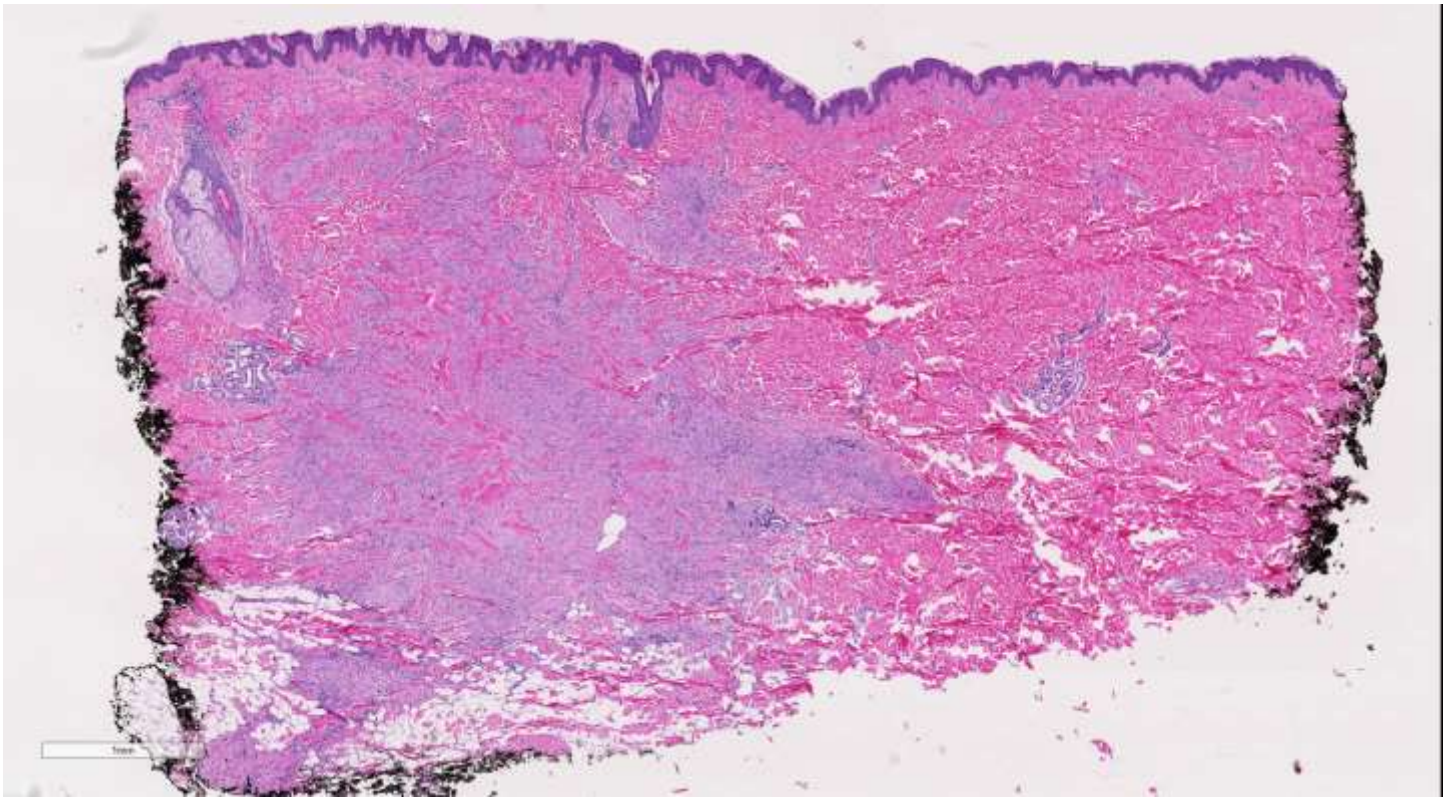


Figure 1: Small, tender, indurated, flesh-colored papule on the left upper abdomen.



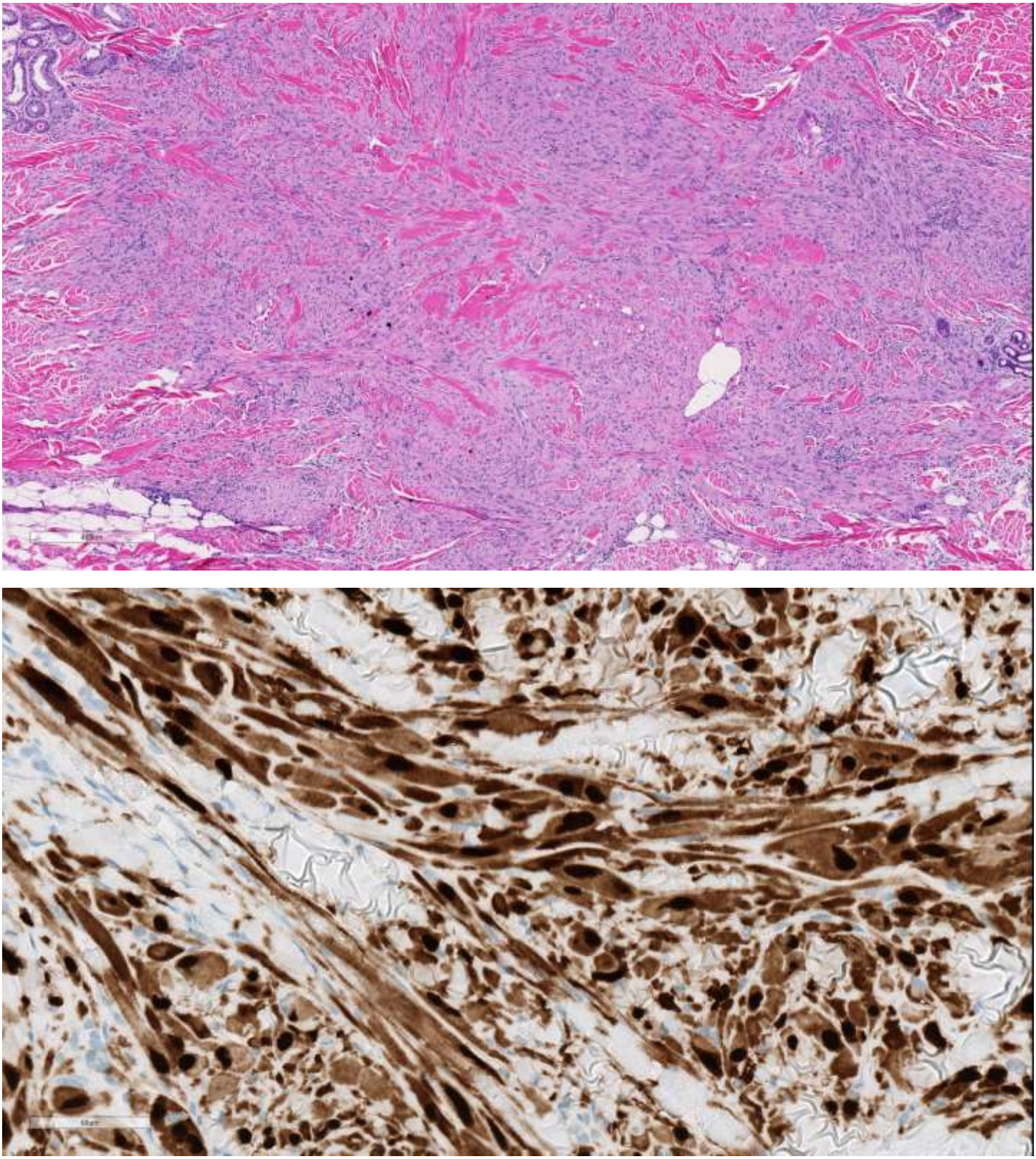


Figure 2a,b,c: A) Punch biopsy specimen showing a spindle cell mass invading the deep margin. B) Medium power image showing plump spindled and epitheliod fascicles. C) FOS B positivity IHC courtesy of Mayo Clinic.