SKIN

BRIEF ARTICLE

Angiosarcoma Clinically Mimicking a Targetoid Hemosiderotic Hemangioma

Hannah Nam, BA[1], Jordan Lim, MB BCh BAO[1], Alexandra Flamm, MD[1]

[1]Penn State Health, Hershey Medical Center, Department of Dermatology, Hershey, PA

ABSTRACT

Angiosarcoma is a rare malignant entity that typically presents as a rapidly progressing ecchymotic or cellulitis-like lesion with subsequent bleeding or ulceration. Atypical presentations have been described, such as cases mimicking rosacea, making the diagnosis more challenging. Due to the high rate of local recurrence, metastasis, and disease-specific mortality, early identification and treatment is critical. Here we describe a 69-year-old man with a vascular targetoid lesion clinically consistent with a targetoid hemosiderotic hemangioma (THH); however, biopsy was consistent with an angiosarcoma. Early diagnosis of this lesion resulted in prompt wide local excision and no detection of metastasis. This case report highlights the importance of thorough evaluation of new onset vascular lesions and the unique THH-like presentation of an angiosarcoma.

INTRODUCTION

Angiosarcoma is a rare, aggressive soft-tissue sarcoma of endothelial cell origin with increased likelihood for local recurrence, metastasis, and poor prognosis. Angiosarcomas can affect any organ in the body; however, nearly 50% of all cases are of its cutaneous form often of the head and neck[1].

The five year survival of cutaneous angiosarcoma ranges from 10-30%[2]. The variation in clinical presentation and the lack of disease awareness contribute to the difficulty in diagnosis[3]; however, early detection and intervention is critical for improved prognosis. The size of lesion at presentation is an important prognostic factor for cutaneous angiosarcoma[3], and a delayed diagnosis may lead to substantial challenges in treatment[4]. Perez et al. reported that a tumor size less than 5cm was associated with a significantly improved overall and recurrence-free survival compared to a tumor size greater than 5cm[5]. Despite early detection, the neoplasm can often be more advanced than is apparent from initial physical exam[2]. This case report emphasizes the value of early clinical suspicion for atypical, rapidly changing vascular lesions.

CASE REPORT

A 69-year-old white man with no history of skin cancer presented with a 2cm vascular targetoid lesion with a central 5mm firm dermal purple nodule and surrounding ecchymosis on his right flank (Figure 1). The patient noticed the lesion approximately three months prior which then progressively grew. No palpable axillary or inguinal lymph nodes were noted. Review of systems was

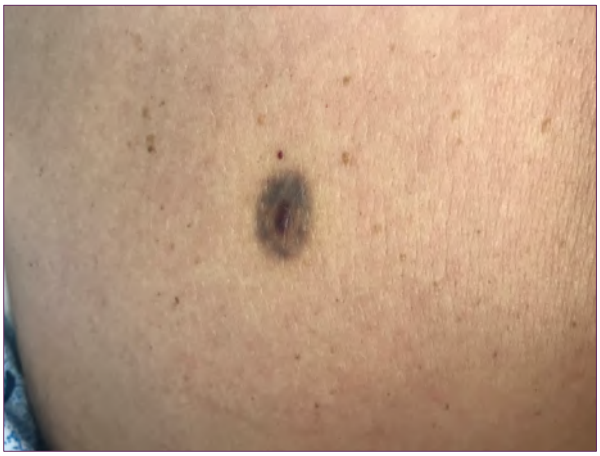

Figure 1. 2cm targetoid vascular lesion on right flank

unremarkable. A 5mm punch biopsy of the lesion was performed.

Histopathologic evaluation revealed vascular proliferation within the superficial to mid dermis composed of enlarged and pleomorphic endothelial cells. Many of the endothelial cells formed small irregular ectatic vascular units, with crack-like spaces. Scattered mitotic figures along with single scattered enlarged endothelial cells with background hemorrhage were observed (Figure 2). The endothelial cells were highlighted on CD31 immunostaining and negative with SOX-10, Melan-A, and HHV-8 immunostaining (Figure 3). Mib-1 immunostaining revealed an increased proliferation index. This was consistent with a diagnosis of an angiosarcoma.

A full body PET scan demonstrated no areas of concern for metastasis. A wide local excision with 2.5cm margins followed by reconstruction with a posteriorly-based advancement flap was completed. The tumor was completely excised with negative margins. This case was discussed at a multidisciplinary tumor board where surgical treatment alone was determined to be sufficient for this localized, small lesion. Monitoring will consist of PET scans every six months for two years

DISCUSSION

Cutaneous angiosarcoma is an aggressive malignant lesion frequently presenting as a raised purplish-red papule on the head and neck. Early presentations can often be dangerously mistaken as a benign vascular lesion, leading to delayed diagnosis, delayed treatment, and worse prognosis[6]. Histological examination of angiosarcoma include irregular vascular channels and atypical endothelial cells that may appear normal, pleomorphic, and hyperchromatic[7]. Immunohistochemistry may be positive for CD34 and CD31[7].The local recurrence rates of cutaneous angiosarcoma after surgical excision have been reported to range from 35-86%[8]. A retrospective study conducted at the Mayo Clinic reported a 5-year-recurrence free survival of 16%, a 5-year local regional control of 18%, and a 5-year overall survival of 38% among patients treated for cutaneous angiosarcoma [9]. Another study conducted among patients diagnosed with angiosarcoma of the face and scalp reported an even higher recurrence rate of 93.8%[8]. Imaging including MRI, CT, and PET scan is crucial for staging and management of angiosarcoma. Due to its rare incidence, the optimal treatment is not currently established[9]. Wide local excision with negative margins is the gold standard; however, currently, there are no specific margins recommended by the NCCN guidelines. The multifocal nature of the lesion and propensity for tissue infiltration often result in positive resection margins, recurrence, and metastasis of the tumor[10]

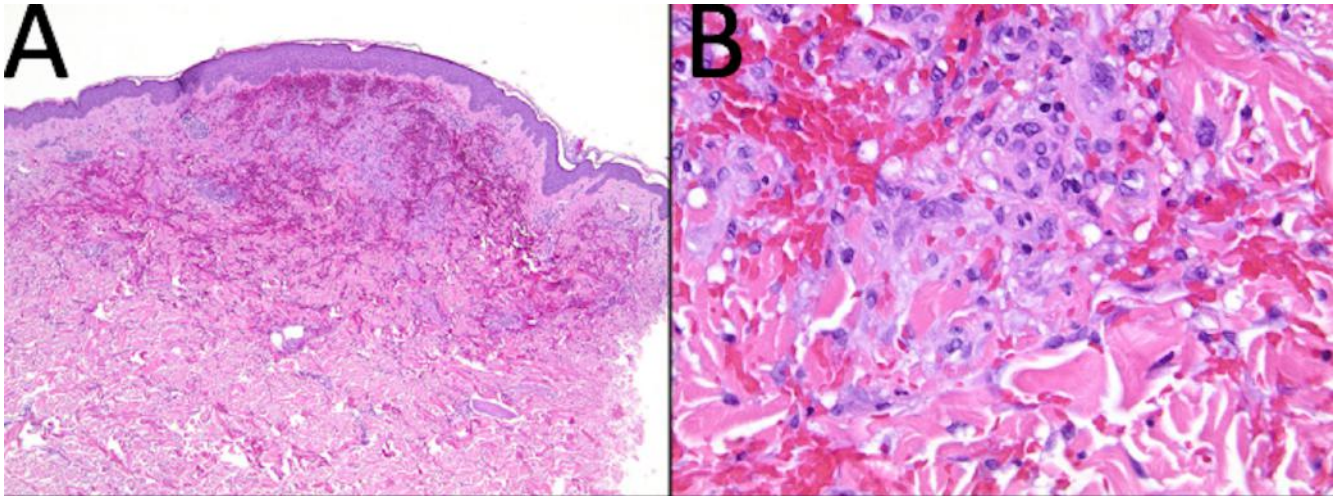

Figure 2. 5mm bunch biopsy of right flank. H&E stain **A)** 4x showing an atypical vascular proliferation and background hemorrhage within the dermis **B)** 40x highlighting the atypical endothelial cells demonstrating mitotic figures, arranged singly and forming small irregular vessels

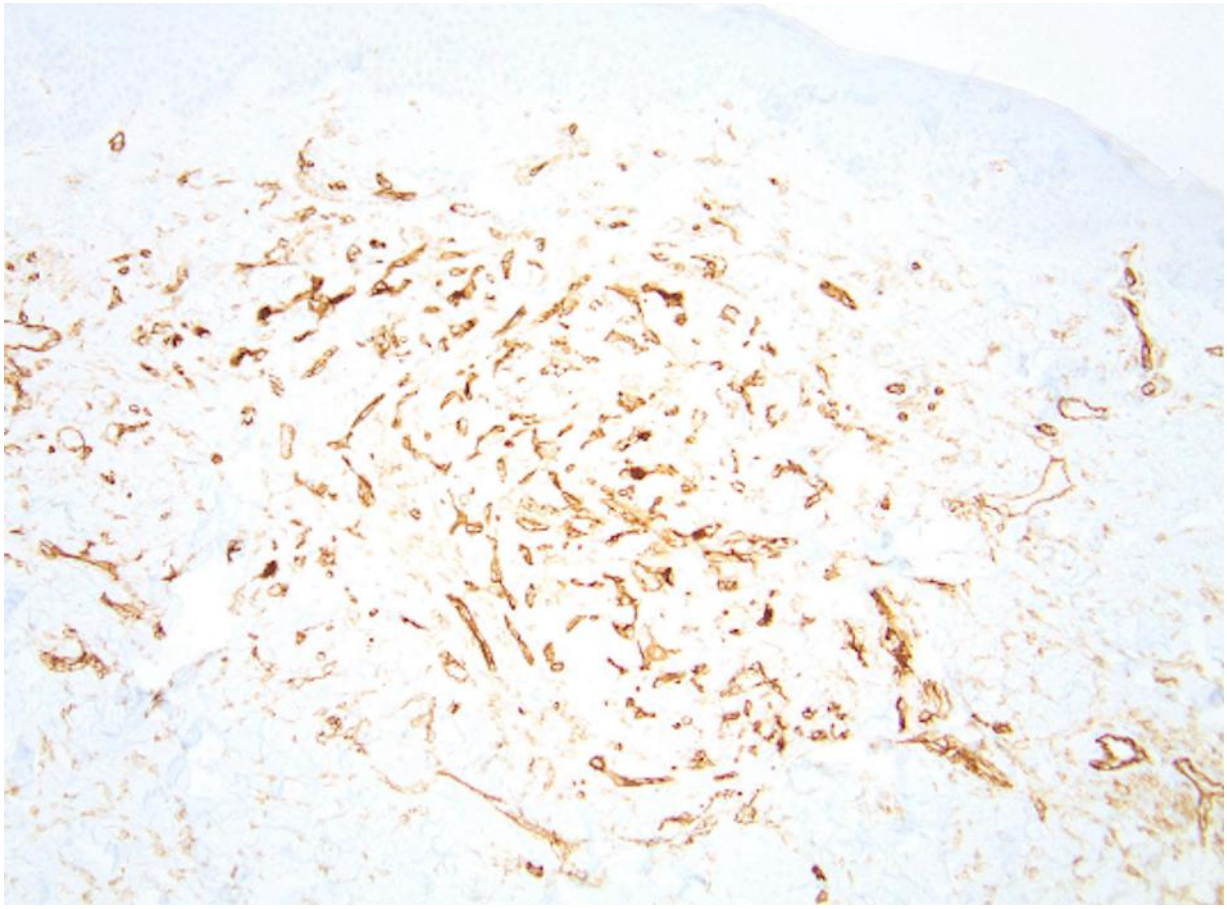

Figure 3. CD31 Staining highlighting the atypical endothelial cells and vessels (10x)

and an additional peripheral ecchymotic ring. THH lesions typically occur on the proximal extremities and trunk. Several studies have reported a cyclical variation in palpability, In

In contrast, targetoid hemosiderotic hemangioma (THH), also known as hobnail hemangioma, is a benign vascular lesion that often presents as a solitary annular violaceous papule surrounded by a pale ring size, and color of the lesions, which contributes to the challenge in diagnosis[11]. Histological findings of THH include non-circumscribed dermal vascular proliferations that are usually found within the superficial and mid dermal layers of the skin[11]. The vascular channels are lined by a single layer of "hobnail-like" endothelial cells. Excision of a THH lesion is mostly curative, and malignant transformation, recurrence or systemic metastases have not been reported[12].

CONCLUSION

In this case we demonstrate a unique clinical presentation of angiosarcoma in which a targetoid vascular lesion was identified, a finding more commonly associated with THH. The wide variety of clinical presentations of THH and angiosarcoma can make initial diagnosis challenging; however, early clinical suspicion is critical due to their vastly different prognoses.

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Corresponding Author:
Alexandra Flamm, MD
Assistant Professor of Dermatology & Pathology
Department of Dermatology
Penn State Hershey Medical Center
500 University Drive
Hershey, PA 17033
Phone: 717-531-8307
Fax: 717-531-4702
Email: aflamm@pennstatehealth.psu.edu

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