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Rapidly fatal metastatic cutaneous angiosarcoma initially mimicking a furuncle in a middle-aged male

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Abstract

Cutaneous angiosarcomas are rare but typically occur in three distinct clinical settings and are most commonly found on the scalp or face of elderly men. Positive prognostic factors include tumor size less than 5 cm, primary tumor location below the head, negative margins after excision, resectability, and younger age. Metastases drastically reduce survival and the most common metastatic site is lung. We present a 43-year-old man who had primary cutaneous angiosarcoma that initially mimicked a furuncle and eventuated in multiple metastases. The metastatic disease included brain involvement, which has rarely been reported, especially in a relatively young person without known predisposing conditions. This unique case also highlights the need for early diagnosis followed by advanced imaging, given the limitations of current therapies and high metastatic potential of angiosarcoma.

Keywords: cutaneous angiosarcoma, furuncle, chemotherapy, radiation

Introduction

Angiosarcoma (AS) is a rare vascular malignancy accounting for <2% of all soft tissue sarcomas. Although AS can originate from any anatomic site, >50% are primary to the skin, with the remainder arising in deep soft tissue and viscera [1-4]. Cutaneous angiosarcomas (cAS) typically occur in three distinct clinical settings: cAS associated with chronic lymphedema (Stewart-Treves syndrome), post-radiation cAS, and most commonly, idiopathic (sporadic) cAS of the head and neck. Idiopathic cAS classically presents as an asymptomatic enlarging bruise-like plaque on the scalp or face of elderly men, but it may be more nodular and resemble benign or malignant neoplasms or various inflammatory disorders [1]. The prognosis for AS is poor, with overall survival ranging from 10-54% depending upon the site of the primary [1-4]. Similar to other sarcomas, the most common site for metastases is lung, followed by liver, bone, soft tissue, and lymph nodes [2]. Brain metastases are rare, with the majority of cases related to primary cardiac AS. True brain metastases from cAS, as opposed to direct extension, are exceptionally rare and only a few isolated cases have been reported [5, 6].

Case Synopsis

A 43-year-old retired radar operator presented with a three-month history of an enlarging 3 cm violaceous nodule on his left cheek that he first noticed after cutting himself shaving (**Figure 1A**, **B**). His primary care provider initially suspected a furuncle because he had similar lesions in the past. Incision and drainage were performed, followed by a course of sulfamethoxazole/trimethoprim and a triamcinolone injection. When the lesion failed to respond, punch biopsies were obtained, which showed an infiltrative dermal proliferation comprised of pleomorphic endothelial cells that formed irregular anastomosing vascular channels and diffusely stained with CD31, consistent with cAS (**Figure 2**). While awaiting definitive excision, he developed severe headaches and left-sided weakness. A right parietal intraparenchymal hemorrhagic mass suggestive of metastatic AS was seen on head CT (**Figure 1C**) and pulmonary metastases were later identified. The patient underwent right parietal craniotomy and then resection of the enlarging 6.5 cm facial cAS, followed by dorsocervical flap reconstruction and

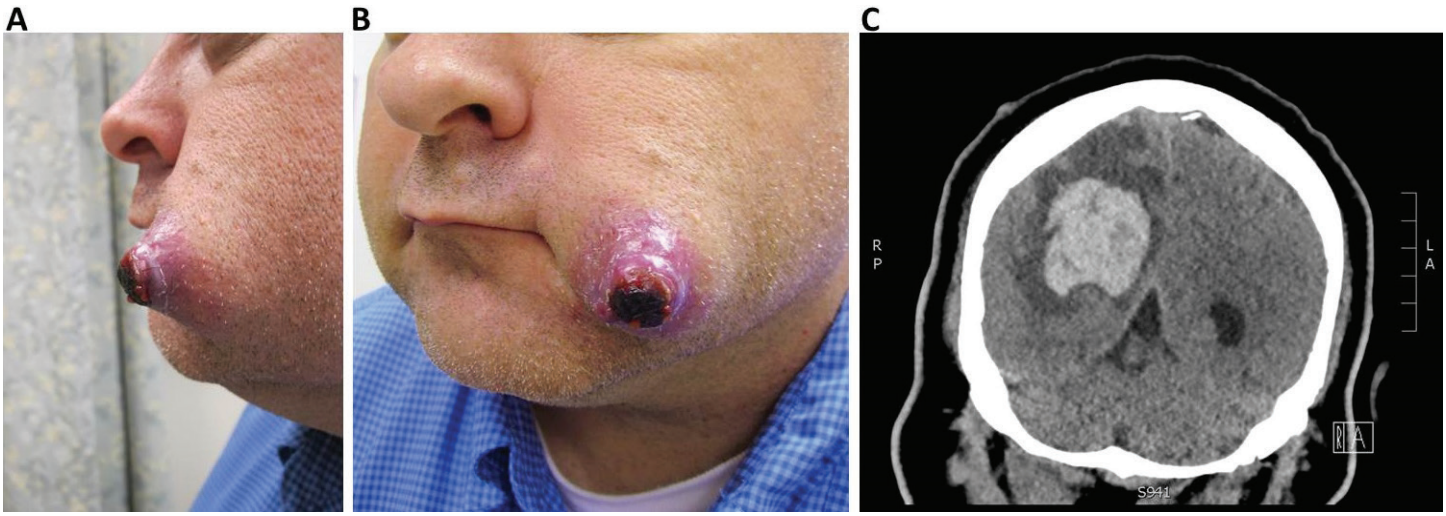

Figure 1. *Clinical Findings: A and B) Side and anterior views of the lesion after failed initial management, showing a protuberant 3 cm violaceous nodule with eschar on the left cheek. C) Head CT without contrast showing a 3.7 x 6.0 x 4.3 cm intraparenchymal hemorrhage of the right parietal lobe associated with surrounding edema and right-to-left midline shift.*

whole-brain radiation. The patient then received three cycles of paclitaxel but had metastases to his right atrium and expired nine months after his initial diagnosis.

Discussion

Most cAS arise in elderly men on chronically sun-damaged or, less commonly, prior irradiated skin with a median age at diagnosis of 71 years [3]. Other AS risk factors include chronic lymphedema and exposure to polyvinyl chloride, thorium chloride, arsenic, and radium, which are primarily associated with hepatic AS [4].

Contrary to other soft tissue sarcomas, histologic grading of AS is not useful in predicting outcome. Therefore, all AS are classified as high-grade malignancies [2]. Prognostic factors associated with worse clinical outcome include patient age >70, tumor size >5 cm, tumor stage III or IV, head and neck primaries, positive surgical margins, and failure to use multimodal therapy [1-4]. Median survival is approximately 25.2 months, but this is dramatically reduced to 2-6 months if metastases are present [2, 4, 6]. Since prognosis is closely linked to size and degree of invasion, early recognition is paramount, but diagnosis is often delayed because of lack of clinical suspicion and confusion by a broad differential diagnosis: furuncle, epidermoid cyst, squamous cell carcinoma, pseudolymphoma, cellulitis, erysipelas, fungal infection, atypical mycobacterial infection, rosacea, dermatitis, insect bite, or hematoma.

Treatment of choice is surgical excision, but cAS is notoriously ill-defined and multi-focal, making complete resection difficult; the benefit of Mohs is unclear. Therefore, adjunctive radiation, or radiation alone for poor surgical candidates is commonly performed [1-4]. Chemotherapies, primarily anthracycline, ifosfamide, and taxanes, are used in advanced disease but mostly as palliation. Currently, multiple trials are investigating therapies directed against VEGF or tyrosine kinases, but survival has not improved [2, 4].

Conclusion

This unique case describes a rapidly fatal cAS arising in a younger patient with no known predisposing risk factors that initially mimicked a furuncle. Owing to the high risk of metastatic disease, we concur with other authors and recommend advanced radiologic imaging upon definitive diagnosis [4].

Acknowledgments

The views expressed in this article are those of the authors and do not necessarily reflect the official policy or position of the Department of the Navy, Department of Defense, or the United States Government.

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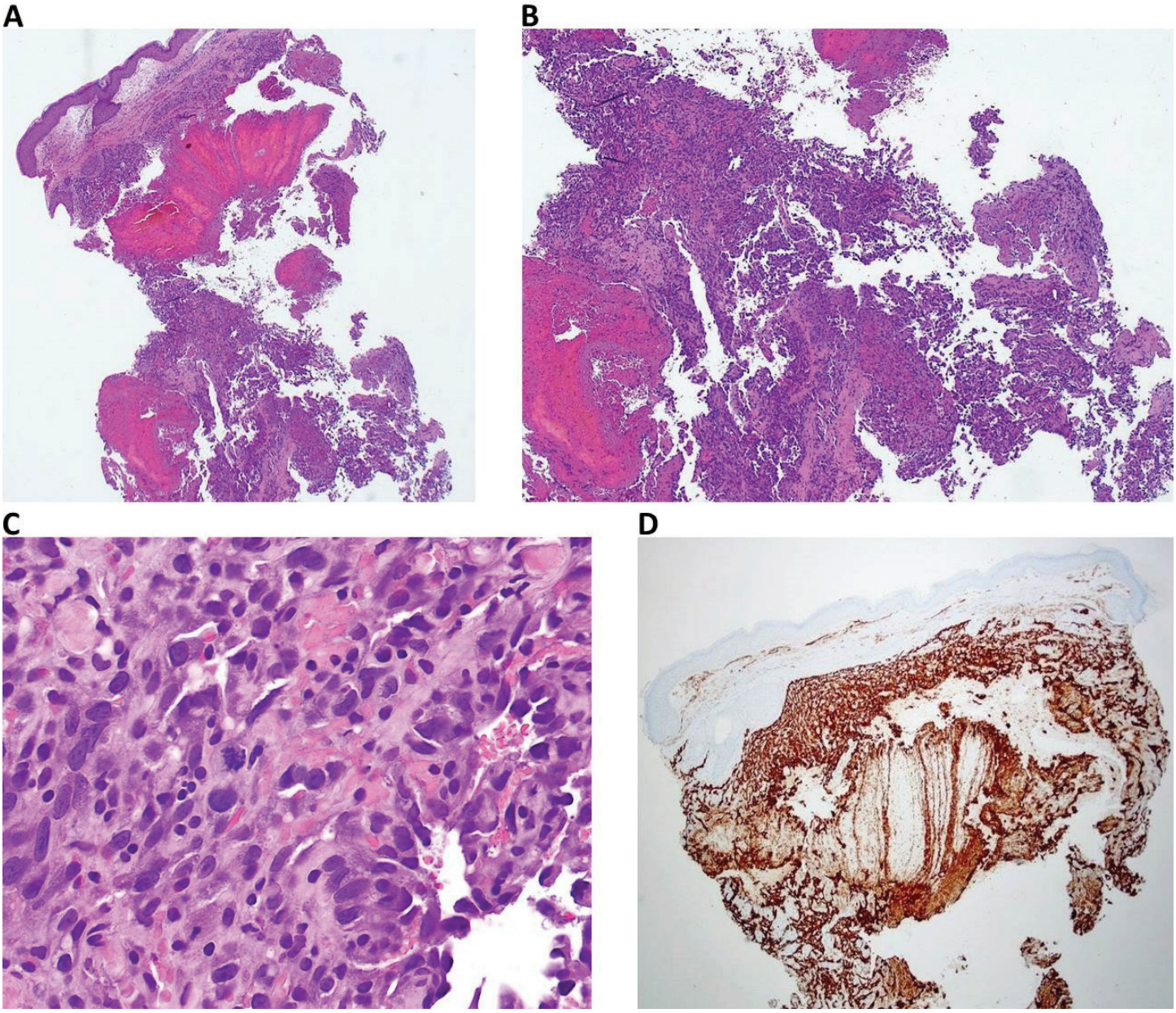

Figure 2. *Histopathologic Findings: A) Diffuse replacement of the dermis by an infiltrative and hemorrhagic vasoformative proliferation, H&E 20%. B) Complex inter-anastomosing jagged channels, H&E, 40%. C) Vascular channels lined by plump pleomorphic hyperchromatic endothelial cells with atypical mitotic figures, H&E 400%. D) CD31 immunohistochemical stain, diffuse positive staining, 40%. The cells of interest failed to label with cytokeratin, Factor XIIIA, and HHV-8 immunohistochemical stains. Cultures and PAS, GMS, and AFB-stained slides all failed to identify an infectious etiology.*

None of the authors have any conflicts of interest to report.

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