Cutaneous Angiosarcoma of the Forehead with Parotid and Lung Metastases

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Introduction
Cutaneous angiosarcoma is a rare subtype of soft tissue sarcoma. Up to 50% of cases occur in the head and neck and the most common subsites are the scalp and upper face.1 It is an aggressive neoplasm with a tendency for early distant metastasis. We present a challenging case of metastatic cutaneous angiosarcoma treated at our institution.

Case History
A 77-year-old gentleman presented with a 4 month history of a “bruise” in the glabellar region, with no report of prior trauma. The patient was initially diagnosed with a hematoma, but sought further opinion when the discoloration did not resolve. The patient denied any pain or numbness over the area or vision changes. The review of systems was negative for fever or weight loss. His past medical history was significant for hypertension and diabetes. He was a non-smoker.

Physical exam demonstrated an ecchymotic skin lesion of the glabella and central forehead. Palpation revealed an irregular texture of the skin surface and a fullness to the deeper subcutaneous tissue (Figure 1). Neck exam revealed no lymphadenopathy.

Punch biopsy in the glabellar area was performed. Pathology was positive for a vessel-forming neoplasm consistent with angiosarcoma.

Discussion
Cutaneous angiosarcoma of the head and neck can present in many forms, including violaceous nodules, macules, ecchymotic areas, or ulcers. The wide variety of clinical presentations can make diagnosis difficult. Histologically, it appears as anastomosing vascular channels lined by large endothelial cells. The majority of cases occur in elderly white men, and most common risk factor is a history of prior irradiation.3,4 Multifocal disease is common at presentation. Cervical metastasis is uncommon at initial presentation; however, reports have shown up to 25% of patients presenting with distant metastasis. The most common site is the lung.5

Although no optimal treatment regimen has been established, multimodal therapy is required for management. Initial surgical treatment is often difficult due to multifocality and poor control of margins. One study of 29 patients achieved negative margins in only 21% of specimens.6 Due to the extensive nature of disease, some authors recommend total scalp irradiation.6,8 Paclitaxel and docetaxel have shown to be effective chemotherapeutic options.7 DeMartelaere et al. found that neoadjuvant chemotherapy may have a role for periorbital angiosarcoma in potentially obviating the need for radical surgical resection.8

Initial size and stage of the lesion correlates with long-term prognosis. Lydiatt et al. found that all patients with tumor size above 3cm or unresectable lesions died of disease.9 A recently published study treating patients with surgical resection and adjuvant therapy reported a 5-year survival rate under 22%.10

Conclusion
Cutaneous angiosarcoma is a rare, aggressive neoplasm occurring in the head and neck. Treatment is complicated by the difficulty making early diagnosis, obtaining negative surgical margins, and by the tendency for early metastatic spread. Early diagnosis and interdisciplinary treatment protocols are crucial for effective management of this disease.

References

Studies
MRI with contrast revealed a 1cm by 3.3cm heterogeneously enhancing mass of the soft tissues of the glabella, forehead, and pre-septal left orbit. An additional lesion was seen in the left temporal region. No underlying changes to the frontal bone or meninges (Figure 2). Initial chest CT revealed small pulmonary calcifications. Whole body PET scan did not reveal any abnormal activity at the primary site or distant areas.

Cutaneous Angiosarcoma of the Forehead

![Figure 1. Forehead lesion.](image1)

![Figure 2. MRI scan showing forehead and left temporal lesion.](image2)

![Figure 3. MRI with left parotid mass.](image3)

![Figure 4. CT chest with multiple ground-glass opacities.](image4)

![Figure 5. Parotidectomy specimen.](image5)