Angiosarcoma of the scalp mimicking a sebaceous cyst

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Abstract

A 58-year-old Caucasian woman presented with a cystic lump behind the right ear that was clinically diagnosed as an infected sebaceous cyst. The lesion was treated with incision and drainage followed by antibiotics for 3 months. Because there was no resolution, a biopsy was performed that revealed a high grade angiosarcoma. She expired 2 months later. Cutaneous angiosarcoma is an extremely aggressive tumor. Therefore early diagnosis and management is crucial in providing better patient care.

Clinical synopsis

A 58-year-old Caucasian female presented to the clinic with a pruritic cystic lump behind her right ear. The nodule had been present for an unknown period of time and she attributed it to an insect bite. An infection was suspected and she was treated with antibiotics. One month later she presented to the hospital again with complaints of enlargement of the lump and pain. Physical examination showed a soft and spongy lesion with purple streaks behind the right ear. A sebaceous cyst with infection was suspected. An incision was performed with drainage of some yellow-pink fluid that was negative for microorganisms on culture. At the follow-up visit 1 month later the purple-blue mass in the right postauricular area appeared to be
much larger, measuring 8x5x3 cm (Fig. 1). The lesion extended from the superior to the inferior edge of the postauricular groove without involvement of the pinna. There were areas of superficial breakdown and drainage of some serosanguineous and purulent fluid. Multiple enlarged, firm, and mobile lymph nodes were palpated in the right mandibular region and in the right anterior and posterior triangle of the neck.

An incisional biopsy of the lesion showed a dermal neoplasm composed of infiltrating irregular, vascular or sinusoidal anastomosing channels (Fig. 2A). Slit-like vessels with dissection of the dermal collagen were prominent. The vessels were lined by a disorganized proliferation of polyhedral atypical endothelial cells with a high nucleus to cytoplasm ratio, scant amphophilic cytoplasm, and dark blue nuclei. Micropapillae of the neoplastic cells with a hobnail appearance were seen projecting into the lumen of the neoplastic vessels (Figs. 2A and 2B). There were also focal clusters of solid tumor cells. By immunohistochemical staining, the neoplastic cells showed strong positivity to CD31, CD34 (Fig. 3A) and Factor VIII (Fig. 3B) with a moderate to high proliferative rate by Ki67 staining (Fig. 3C). A high grade angiosarcoma of the scalp was diagnosed. A subsequent biopsy of a cervical lymph node revealed many atypical cells that were highly suspicious for metastatic malignancy. The patient expired 2 months after her diagnosis was made and 5 months after the presentation to the clinic.

![Figure 2A](image1.png)  ![Figure 2B](image2.png)

**Figure 2A. Microscopic photograph, low power**  
**Figure 2B. Microscopic photograph, high power**

![Figure 3A](image3.png)  ![Figure 3B](image4.png)

**Figure 3A. CD34 Stain**  
**Figure 3B. Factor VIII stain**
Comment

Cutaneous angiosarcoma primarily affects elderly patients and is commonly located on the head and neck, especially the scalp. Currently, angiosarcoma of the scalp in elderly patients is recognized as a unique subtype that is also known as senile angiosarcoma or malignant angioendothelioma [1, 2, 3]. It commonly occurs in elderly white men with a male-to-female ratio of 3:1 [1, 3]. The predisposing factors for angiosarcoma of the scalp is uncertain. Post-radiation cutaneous angiosarcoma of the chest wall after treatment of breast carcinoma is well recognized. Chronically edematous skin, as seen in post-mastectomy arms and chest wall or rarely in the abdominal wall may be the site for the development of angiosarcoma [4]. One association with angiosarcoma of the scalp is prior radiation. Our patient did not have a history of radiation.

Clinically, angiosarcomas of the scalp have a variety of presentations. Most cases start as an ill-defined bruise-like lesion with an indurated border. The tumors may be single or multifocal, blue or violaceous, and raised or flat. In the advanced stage, the tumor is typically large, spongy, and poorly demarcated as well as elevated, multinodular and blue-red. Hemorrhage and ulceration are also frequently seen. It is difficult to determine the extent of the tumor clinically. In our case, the patient presented with as an unusual cystic lump mimicking an infected sebaceous cyst.

Microscopically, the classic angiosarcoma is composed of a disorganized vascular or sinusoidal anastomosing network with infiltration into the dermis; the neoplastic slit-like vessels dissect the dermal collagen. The tumor cells are polyhedral, atypical endothelial cells with a high nucleus to cytoplasm ratio and scant amphophilic cytoplasm. Angiosarcoma may also have other histological variants that include the spindle type, solid epithelioid type, hemangioma-like type, granular cell variant, and pleomorphic type [5].

Several benign and malignant tumors are among the differential diagnoses. Hobnail hemangioma, also named targetoid hemosiderotic hemangioma or atypical hemangioma, may show arrays of angulated vascular channels in the dermis with dissection of the collagen bundles. These tumors can also show papillary projections into the vascular lumen lined by neoplastic endothelial cells in a hobnail pattern. All these features are similar to those in classic angiosarcoma. However, hobnail hemangioma usually develops on the skin of the extremities in young adults and microscopically shows a distinct biphasic appearance composed of a superficial portion with dilated vessels and a deeper portion with attenuated slit-like capillaries. The tumor cells in hobnail hemangioma show bland nuclear features, a low proliferative rate, and often abundant hemosiderin-laden macrophages. Papillary
endovascular angioendothelioma may be confused with angiosarcoma, but it is an intravascular tumor and angiosarcoma is almost never confined solely to a vascular lumen. Histologically, spindle cell angiosarcoma often resembles Kaposi’s sarcoma. However, these two entities usually differ substantially in clinical features. Kaposi’s sarcoma is usually positive for HHV8 and occurs in patients with HIV infection or immunodeficiency [4]. Epithelioid angiosarcoma may mimic other epithelioid tumors, including angiomatoid squamous cell carcinoma, melanoma, epithelioid leiomyosarcoma and clear cell sarcoma. Pleomorphic angiosarcoma can be confused with atypical fibroxanthoma, undifferentiated carcinoma, melanoma, and malignant fibrous histiocytoma. Immuno stains as well as electron microscopy and cytogenetic analyses are often helpful in the differential diagnosis of these tumors.

Angiosarcoma of the scalp is difficult to treat; a combination of surgical resection with a wide and deep margin and radiation has been recommended [6]. Recently, chemotherapy using docetaxel has shown promising results in angiosarcomas of the scalp [7]. Surgery alone is often not feasible because of the multifocal nature and local spread pattern of these tumors. Frequently during surgery, the extent of the tumors is very difficult to determine due to the extensive microscopic spread.

The prognosis for patients with angiosarcomas of the scalp is poor because these tumors have a tendency for local recurrence and metastasis by lymphatic or hematogenous routes. In our case, the patient died soon after diagnosis with metastasis. Clinical studies have shown that the most important factor in determining prognosis is the size of tumor. Tumors less than 5 cm are associated with a significantly better prognosis than those larger than 5 cm. An early diagnosis offers the only chance of possible control of this highly lethal tumor. Differential diagnosis of a cystic lesion in the scalp of an elderly patient should include the possibility of malignant neoplasm.

References


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