# **Erythematous Nodular Scalp Swellings Showing Central Ulceration and Hemorrhagic Crusting in an 83-year-old Man**

83-year-old male patient presented with multiple reddish swelling over the scalp since the past 6 months which gradually increased in size. The patient had a history of fall 6 months back, after which he developed dusky red discoloration over the right side of the scalp, which soon progressed to reddish swellings over the same site. After 3 months, he also developed swelling over the left side and the vertex of the scalp, and since the past 1.5 months the swelling over the vertex and the right side developed spontaneous painful raw areas with occasional bleeding. On examination, diffuse ill-defined dusky red firm nodular swellings [Figure 1a] were present on both the left and the right frontal area and the vertex of the scalp. The swellings on the right side [Figure 1b] and the vertex [Figure 1c] showed central ulceration covered with hemorrhagic crusting. Also, all the swellings were present over the hairless area of the scalp and there were no palpable lymph nodes in the neck. Fine-needle aspiration cytology of lymph nodes was normal.

b C

Figure 1: Clinical images of erythematous firm nodular swellings over scalp (a), with the swellings on right frontal area (b) and vertex of the scalp (c) showing central ulceration covered with hemorrhagic crusting

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Baseline investigations such as complete blood count, liver functions tests, and renal function tests were within normal limits. with no atypical cells on peripheral smear. Ultrasonography (USG) of the abdomen and pelvis was normal. USG of the neck showed few sub-centimeter lymph nodes in the bilateral infraparotid, bilateral posterior triangle, and bilateral jugular chain. On histopathology examination, non-nucleated, non-dysplastic squamous epithelium with underlying tumor tissue composed of anastomosing vascular channels lined by markedly atypical polygonal cells containing pink eosinophilic cytoplasm and atypical mitotic figures was seen. Intratumoral red blood cells and focal vasoformative channels were present [Figure 2]. On immunohistochemistry, CD31 and Vimentin markers were positive, whereas CK, Keratin 3 and 5, ERG, Podoplanin D2-40, and CD 34 were negative [Figure 3].

### Question

What is you diagnosis?

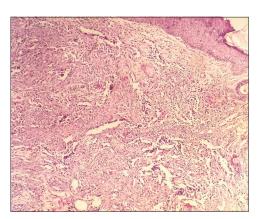


Figure 2: Histopathological image 100x H and E showing tumor tissue composed of anastomosing vascular channels lined by markedly atypical polygonal cells containing pink eosinophilic cytoplasm and atypical mitotic figures. Intra-tumoral red blood cells and focal vasoformative channels are also seen

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Ashish Jagati, Raju G. Chaudhary, Santoshdev P. Rathod, Arwinder Kaur Brar

Department of Dermatology, VS General Hospital, Smt NHL MMC, Ahmedabad, Gujarat, India

Address for correspondence:
Dr. Ashish Jagati,
Department of Dermatology,
VS General Hospital, Smt NHL
MMC, Ellisbridge,
Ahmedabad - 380 006,
Gujarat, India.
E-mail: jagatiashish@gmail.com

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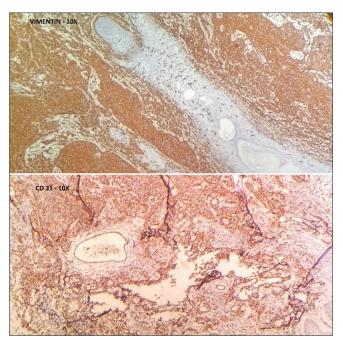


Figure 3: CD31 and Vimentin markers' positivity on immunohistochemistry (10 $\times$ )

### **Answer**

High-grade epitheloid angiosarcoma of the scalp.

### **Discussion**

Cutaneous angiosarcoma of the head and neck is a rare disease. It usually occurs in the dermis of the scalp and less often in the upper face. Typically, the disease manifests clinically with multiple separate foci, especially when it originates in the scalp. According to Morrison *et al.*, the rate of regional nodal involvement, reported to be 20%–30%, is higher than that of most sarcomas.<sup>[1]</sup>

Because most angiosarcoma occurs on the scalp and face of Caucasian population and usually in the non-hair-bearing areas, hence ultraviolet light exposure has been suggested as a contributing cause. [2] In our case also, the lesions were present in the non-hair-bearing areas of scalp. Lydiatt *et al.* investigated the prevalence of metachronous skin cancers as an expression of solar damage and found that 3 of 18 patients had a history of basal or squamous skin cancer, which is same as general population.<sup>[3]</sup>

Low-grade angiosarcoma on histopathological examination is well-differentiated and retains some of the functional and morphologic properties of normal vascular endothelium. They form distinct vascular channels, although these are often irregular in shape and size. Unlike benign hemangiomas, well-differentiated angiosarcomas form vascular channels which creates its own tissue planes as it dissects through the dermal collagen. It is characterized by cells with larger and more chromatic nuclei, with cells often piling up along the lumina, creating papillations. In poorly differentiated (high-grade) tumors, sheets of pleomorphic cells may resemble a carcinoma and vascular channels are lined by atypical endothelium. In highly cellular tumors, the neoplastic process causes such close approximation of cells within vascular spaces that the tumor may appear solid. [3]

Pawlik *et al.* noted that patients with multifocal disease had decreased disease-free survival compared with patients who had only one lesion.<sup>[4]</sup> Death from disease occurs by metastasis. Holden *et al.* reported that only 12% of the patients survived 5 years or longer, with half dying within 15 months of presentation.<sup>[5]</sup>

# Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil

### Conflicts of interest

There are no conflicts of interest.

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