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Case Report

Cutaneous angiosarcoma of the scalp mimicking a keratoacanthoma

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Abstract

Cutaneous angiosarcoma (CA) has a wide range of clinical presentations. In this case report, we discuss a 78-year-old gentleman, who presented with a keratoacanthoma-like scalp lesion that turned out histologically to be a cutaneous angiosarcoma. A brief overview of CA, including its etiology, prognostic factors, clinical manifestations, and treatment options will also be discussed.

Introduction

An angiosarcoma is a rare, highly aggressive and often fatal mesenchymal tumor that originates from vascular endothelial cells. Although it may occur anywhere in the body, it tends to occur in the skin and soft tissue and is especially prevalent in the head ar neck region of elderly males [1]. Cutaneous angiosarcoma of the head and neck region most commonly presents as a single or multifocal enlarging violaceous bruise-like macule or nodule that may bleed intermittently or ulcerate [2]. We report an atypical presentation of a case of CA of the scalp masquerading as a keratoacanthoma.

Case Report

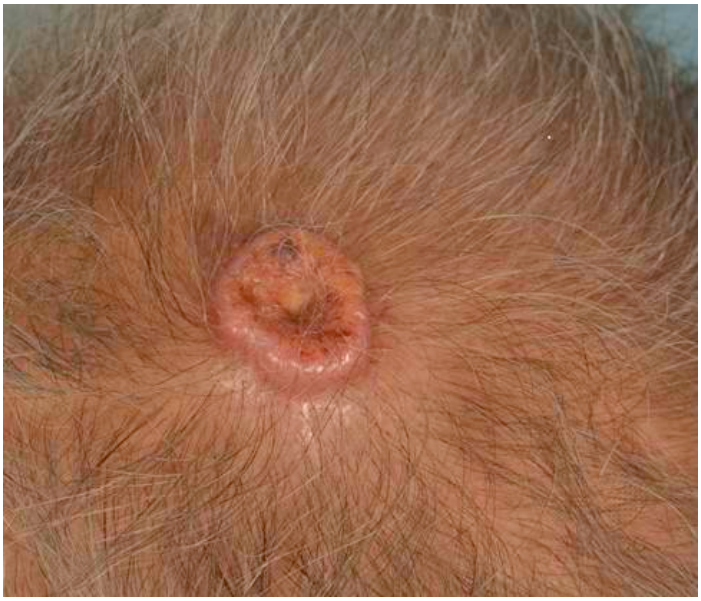
A 78-year-old gentleman presented with a 3 month history of a nodule over the right parietal region of his scalp. It started as a small erythematous papule that enlarged rapidly. It was occasionally itchy, but not associated with any pain, discharge, or bleeding. There were no associated systemic symptoms. There was no history of irradiation to the head and neck region. His past medical history was significant for ischemic heart disease and hyperlipidemia, both under control with medications.

On clinical examination, there was a 1.8cm by 1.8 cm erythematous exophytic nodule on the right parietal scalp with a central crater, bown down to deeper tissues (Figure 1). There was no cervical lymphadenopathy.

Based on the history and clinical appearance, our initial impression was that of a keratoacanthoma of the scalp.

An incisional biopsy of the nodule was subsequently done, which showed an ulcerated tumor composed of inter-anastomosing trabeculae and nests of spindled malignant cells. There was tumor necrosis and readily visible mitoses with focal suggestion of endothelial differentiation. A moderate chronic inflammatory infiltrate was present. The tumor cells were positive for CD31, CD34, and Factor 8, consistent with angiosarcoma (Figures 2-4).

A diagnosis of CA of the scalp was made, and the patient was subsequently referred to the National Cancer Centre, Singapore for further treatment.



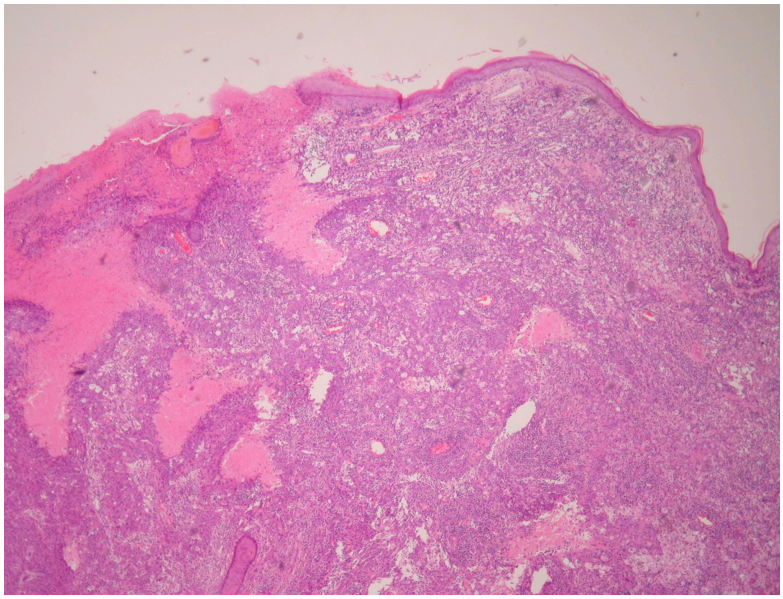
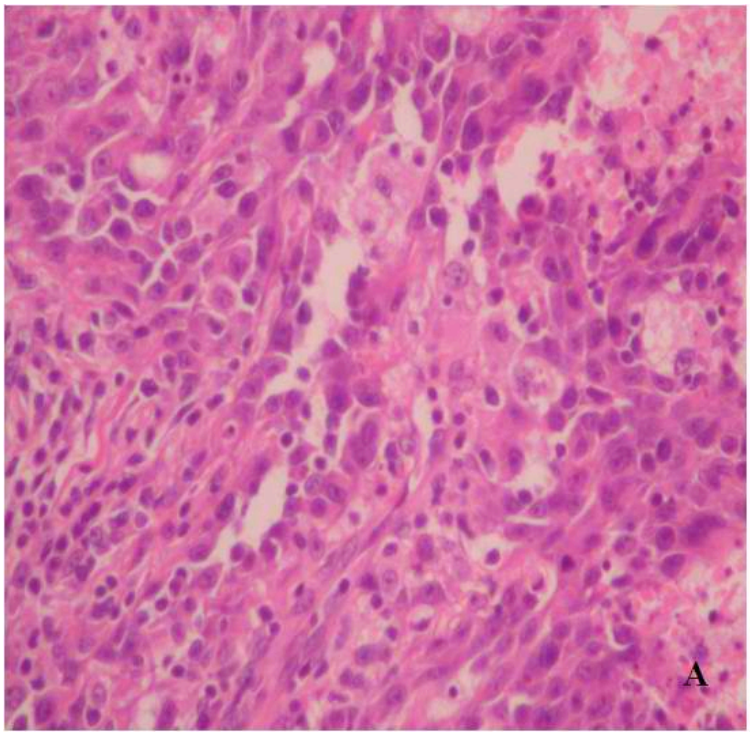


Figure 1: Keratoacanthoma-like nodule over the right parietal scalp.

Figure 2: Low power view, Haemotoxylin & Eosin stain, showing ulceration of the skin with infiltrating sheets and cords of malignant cells and extensive necrosis in the dermis.



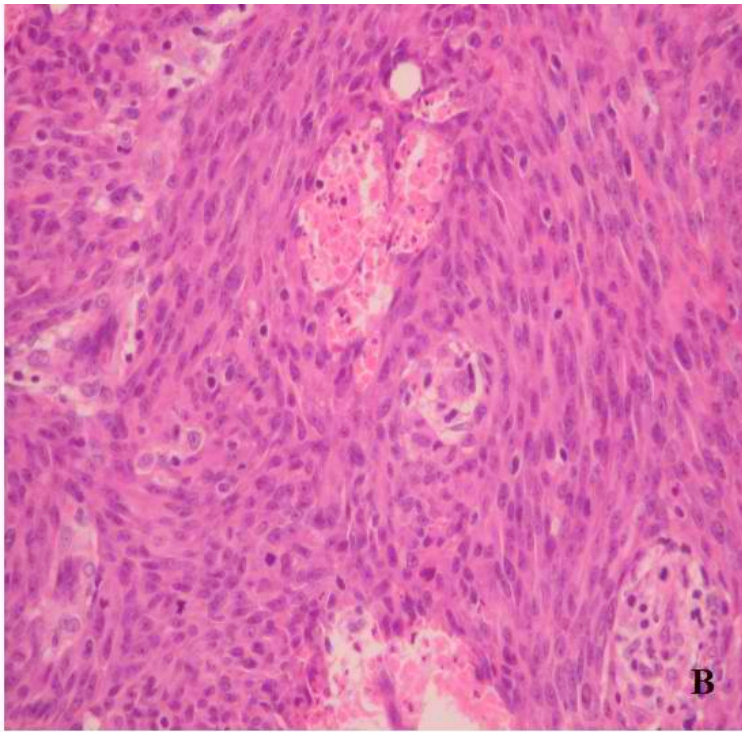


Figure3: High power view, Hemotoxylin & Eosin stain, showing epithelioid area [A] and spindled area [B] comprised of crowded high grad tumor cells forming irregular vascular channels.

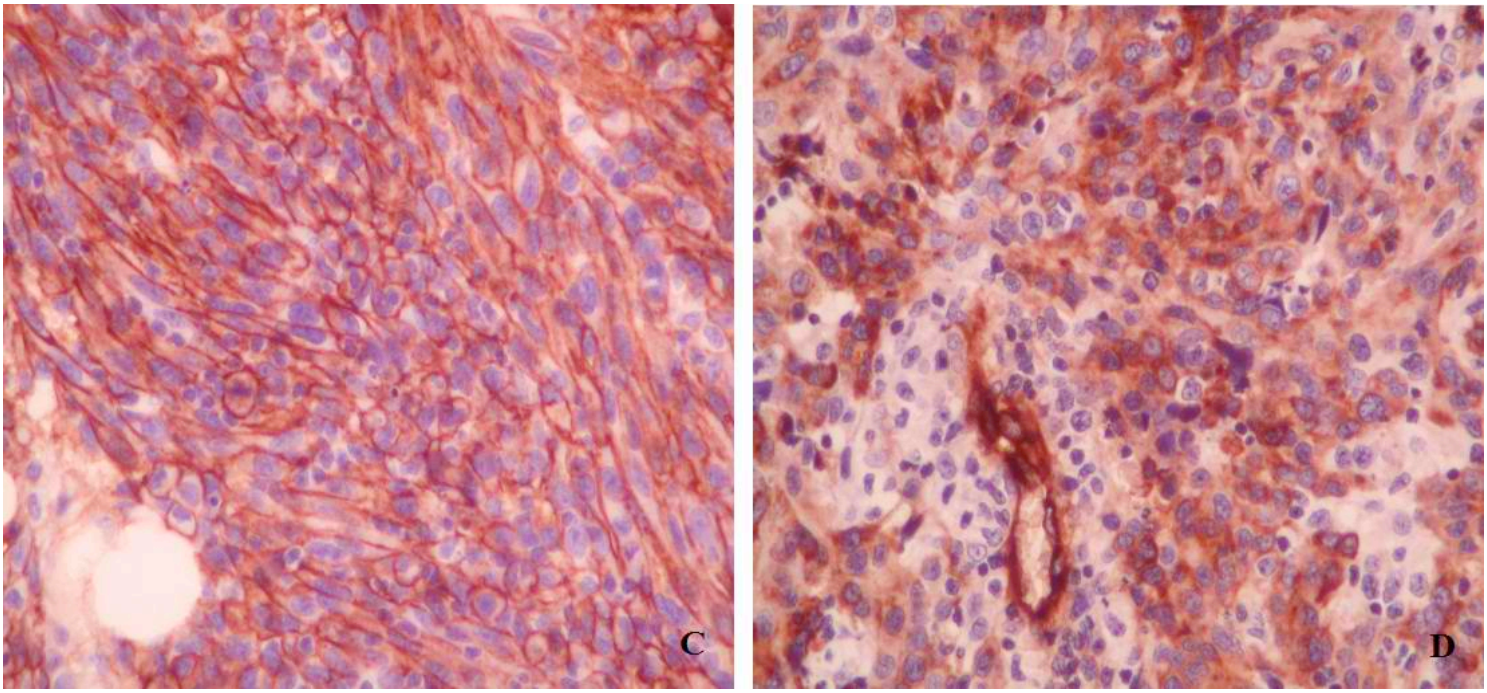


Figure 4: CD 31 [C] and Factor 8 [D] stain the interanastomosing vascular channels formed by the tumor cells.

Discussion

Cutaneous angiosarcoma is a rare neoplasm that accounts for only 1-2% of all sarcomas affecting the soft tissue [3]. A special subtype, with a predilection for the face and scalp of elderly people, was first described by Wilson-Jones in 1964 [4]. Cutaneous angiosarcoma is an insidious tumor with widely variable clinical manifestations, ranging from ill-defined bruise-like patches and plaques in the earlier stages, to elevated non bruise-like nodules in advanced cases, with ulceration and bleeding [2]. Cutaneous angiosarcoma has been misdiagnosed as a pyogenic granuloma [5], a rhinophyma [6], a sebaceous cyst [7] as well as an infective condition like cellulitis [8]. To our knowledge, this is the first report of a CA mimicking a keratoacanthoma.

The etiology for CA remains unknown. UV light exposure, previous irradiation [9], and chronic lymphedema, as in Stewart-Treves syndrome [4], have been cited as possible contributing factors. Angiosarcoma linked to exposure to vinyl chloride, thoriu dioxide (Thorotrast), arsenic and insecticide [3], and preceding trauma [10] have been reported. However, in most cases, as in ou patient, onset is not preceded by any of the aforementioned factors.

Cutaneous angiosarcoma has a grave prognosis. There is often a delay in diagnosis: Patients delay seeking treatment because earl lesions are often benign-looking and asymptomatic. In addition, the myriad of presentations leads to misdiagnosis on the part of the clinician [5]. Disease recurrence is common, ranging from 72.4%-84% [1,2,11], and metastasis is frequent. In a case series reported by Mark et al, distant metastases were detected in 9 out of 28 patients on follow up [11].

The 5 year survival rate ranges from 12-34% [1,10-12]. In a case series of 72 patients, Holden et al reported that only 12% of patients survived 5 years or longer, with half dying within 15 months of diagnosis [12]. The most important prognostic factor appears to be the initial size of the tumor. Poor prognosis is associated with tumors >5cm. Other poor prognostic factors include age >70 years old, multifocal disease, and a lack of a lymphocytic inflammatory response in the tumor [2,12]. The histological grade of the tumor appears not to correlate with prognosis [2,12].

The optimal management of a CA has not been defined. Radical surgery alone has proven to be suboptimal; there is difficulty in achieving negative surgical margins owing to the anatomical site and the multifocality of the tumors [4]. The addition of postoperative radiotherapy has largely produced better outcomes. Pawlik et al demonstrated a statistically significant prolongatio of median survival by 4 times in those who received adjuvant radiotherapy [2]. Similarly, Mark et al. reported an improved survival period with the addition of radiation therapy to surgery [11].

Newer treatment approaches include chemotherapy and biologic agents. Chemotherapy, typically with doxorubicin and taxanes, like paclitaxel and docetaxel, has been used as adjuvant therapy or monotherapy in recent years with success [13]. Intralesional cytokines (IFN-alfa and interleukin 2) and bevacizumab, a recombinant humanized monoclonal IgG1 antibody that inhibits Vascular Endothelial Growth Factor A (VEGF-A) and hence angiogenesis, have also been used to treat CA in conjunction with radiotherapy with success in a few case reports [14,15].

Our patient is currently receiving treatment at the National Cancer Centre of Singapore. Subsequent Computed Tomography imaging done has shown no evidence of metastasis. His tumor is being managed with radiotherapy only because he was deemed t be a poor surgical candidate in view of his comorbidities. We have reported this case because of its unique presentation akin to a keratoacanthoma, making the diagnosis challenging yet interesting.

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