A case of facial angiosarcoma mimicking deep fungal infection

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Abstract

Cutaneous angiosarcoma is a rare, yet a highly aggressive skin and soft tissue tumor with a predilection to the head and neck region in white-skinned elderly males[1].

Cutaneous angiosarcomas have varied clinical presentations and could mimic several clinical entities making the diagnosis a challenge. Upon clinical suspicion, diagnosis is confirmed with histopathology examination and immunohistochemistry. Early diagnosis improves prognosis. We report a case of cutaneous angiosarcoma mimicking facial deep fungal infection.

Introduction

Infiltrative facial lesions are a well-known entity in dermatology practice; diagnosis is confirmed with appropriate investigations excluding the clinical differential diagnoses. Common aetiologies for infiltrative facial lesions include infectious diseases including deep fungal infections, and cutaneous leishmaniasis.

Angiosarcoma, a highly aggressive tumour, yet presenting as rapidly progressive facial infiltration extending almost the entire face and scalp is uncommon. Thus we report a case of angiosarcoma presenting aggressively as an infiltrated plaque mimicking facial zygomycosis.

Case presentation

77-year-old previously healthy female presented with rapidly progressing asymptomatic right-sided facial plaque for 4 months, which initially involved the right cheek and subsequently extended to the scalp and right retro-auricular regions within the next 6 weeks. She had no constitutional symptoms.

Examination revealed a well-defined erythematous, firm plaque extending from the right side of the face to the scalp (Figure 1-2). Nodular growths were noted over the plaque. She had no oral mucosal involvement. There were posterior auricular, sub-mental and cervical lymphadenopathy with hard in consistency.

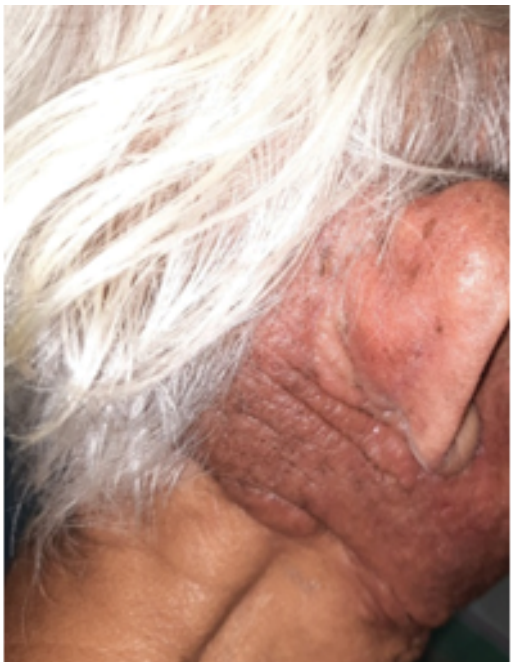

Figure 1.

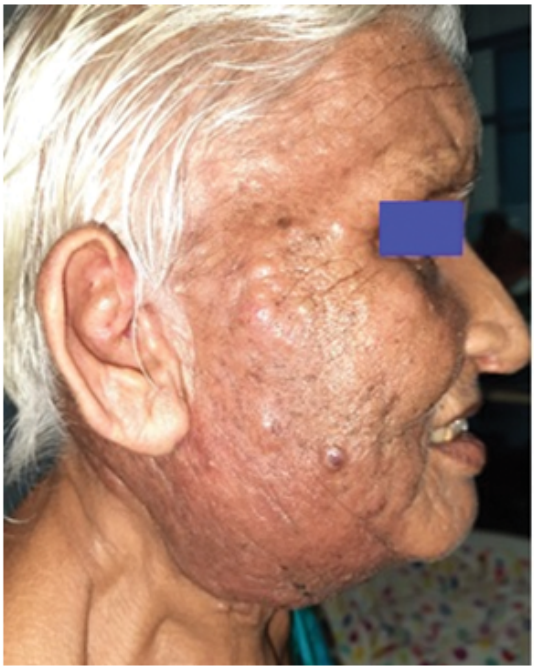

Figure 2.

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Her full blood count was normal. Erythrocyte sedimentation rate 40mm/1st hour. Skin biopsy for fungal culture and the slit skin smear for leishmaniasis were negative. Contrast-enhanced computer tomography (CECT) of head neck, and chest revealed posterior auricular, sub-mental and cervical lymph node infiltrations. CECT chest, abdomen and pelvis did not reveal lymphadenopathy elsewhere.

Histology revealed plum endothelial cell lining, slit-like spaces and insinuating between collagen bundles. Cells displayed nuclear pleormorphism, vesicular nuclei and prominent nucleoli with increased mitotic activity. Immunohistochemistry was positive for CD 34.

Discussion

Angiosarcomas are highly aggressive tumours of the skin and soft tissue with a five-year survival of less than 35%[1,2]. Clinical presentation of cutaneous angiosarcoma of the head and neck of the elderly includes bruise-like lesions, violaceous nodules and plaques, and flat infiltrating haemorrhagic areas. Often these presentations can mimic several other conditions such as vascular malformations, nodular melanoma, lymphoma, sarcoidosis, or facial granuloma[2].

Clinical classification of angiosarcoma includes;

1. Angiosarcoma on the face and scalp of elderly population – most common type.
2. Angiosarcoma either in association with chronic lymphedema or secondary to a prior surgery (Stewart-Treves syndrome).
3. Angiosarcoma following chronic radio dermatitis or skin trauma and ulceration[3].

Facial infiltrated or granulomatous lesions are common in a dermatology clinic. Clinical differential diagnoses are vital to plan out subsequent investigations. Histopathological and microbiological studies are pivotal in arriving at a complete diagnosis.

Common differential diagnoses for facial infiltrated lesions include deep fungal infections, leprosy, cutaneous tuberculosis, cutaneous leishmaniasis and sarcoidosis. However, some rare entities such as angiosarcomas and other malignant neoplasms should be considered in the differential diagnoses as they may mimic infiltrated facial lesions. Early diagnosis and prompt treatment are essential for good prognosis and survival[1,2].

Fast spreading nature with the highest rate of lymph-node metastasis among all soft tissue sarcomas of the head and neck will account for the poor prognosis. Histopathological features of angio-sarcoma are also variable as clinical features. The well-differentiated lesions may mimic hemangioma or lymphangioma, whereas the poorly differentiated lesions mimic melanoma[4]. In such situations, immuno-staining for markers will help in the diagnosis without any delay.

Management options include surgical resection, radiotherapy and chemotherapy which have to be decided on the grounds of tumour bulk, lymph node involvement and patient's co-morbidities.

Due to varying clinical presentations of angio-sarcoma, it should also be included in the clinical differential diagnoses of infiltrative skin lesions particularly head and neck area in elderly with rapid deterioration of symptoms or signs.

References

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