# Story behind a granulomatous facial lesion – angiosarcoma the hidden monster

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## Abstract

Cutaneous angiosarcoma is rare and highly aggressive tumour of skin and soft tissue with the predilection to white skinned elderly. Given its variable clinical presentation it can mimic several clinical entities making the diagnosis difficult. Early diagnosis important for its prognosis and will confirmed by histopathology and immunohistochemistry upon clinical suspicion. Herein we report a case of cutaneous angiosarcoma presenting as a facial granulomatous plaque.


## Introduction

Infiltrative (Granulomatous) facial lesions are a well known entity to presenting to a dermatology clinic. Underlying aetiology needs to be rule out using appropriate investigations excluding the clinical differential diagnoses. Common aetiologies of infiltrative (granulomatous) facial lesion are infectious diseases including leprosy, deep fungal infections, leishmaniasis and inflammatory conditions for example Wagener's granulomatosis and sarcoidosis. Angiosarcoma a highly aggressive tumour and presenting as a perinasal granulomatous plaque is not common. Thus we report a case of angiosarcoma presenting as a perinasal granulomatous plaque.

## Case report

A 75-year-old male with a background history of poorly controlled type 2 diabetes presented to dermatology department with asymptomatic perinasal skin lesion for two months duration. Other than the disfigurement he did not reveal any sinister symptom on systemic inquiry.

On examination there was a poorly defined, erythematous, indurated, firm to hard plaque with irregular borders and irregular surface without any epidermal changes (Figure 1) and the lesion was negative for apple-jelly colour on diascopic examination. There were no similar lesions elsewhere in the body and systemic examination was normal including regional lymph node examination.

His investigations revealed normal haematological parameters including normal ESR, renal function, liver function, CXR and Mantoux (5 mm) were normal, slit skin smears for AFB and Geimsa were negative, TB PCR and leishmanial PCR were negative.

The histopathology revealed only a chronic inflammatory cell infiltrate with possible early granuloma formation. The special stains which were negative include grocott and fite stain. Tissue cultures for tuberculosis/leishmania/fungi were negative.

Since the patient was getting symptomatic with local obstructive features in the half way through investigations (6-8 weeks) and the first biopsy didn't revealed any diagnostic clue a second, deep, incisional biopsy was performed.

The histology of it showed skin with a vascular lesion with complex anastomosing vascular channels lined by atypical endothelial cells suggestive of an angiosarcoma. Then it was confirmed with immunohistochemical studies with strongly positive for CD 31, vimentin and SMA and negative for CD 45, desmin, Myo D 1, Melan A and HMB 45 which confirmed the diagnosis of high grade angiosarcoma. The patient was referred to oncology unit. Currently the patient is on chemotherapy and radiotherapy to which the tumour has responded.

## Discussion

Facial infiltrated or granulomatous lesions are not very rare in a dermatology clinic. Clinical differential diagnoses are vital to plan out subsequent investigations. Histopathological and microbiological studies are pivotal in the process of making a complete diagnosis. Perinasal granulomatous lesion has it's own set of differential diagnoses that a dermatologist tends to consider at a glance including sarcoidosis, leprosy, cutaneous tuberculosis and mucocutaneous leishmaniasis[1]. Being a tropical country, the above differentials should be the first priority. However some rare entities such as angiosarcomas and other malignant neoplasms should be considered in the differential diagnoses as they may mimic granulomatous facial lesions as in our case and also the prognosis depends on the early diagnosis and early treatment.

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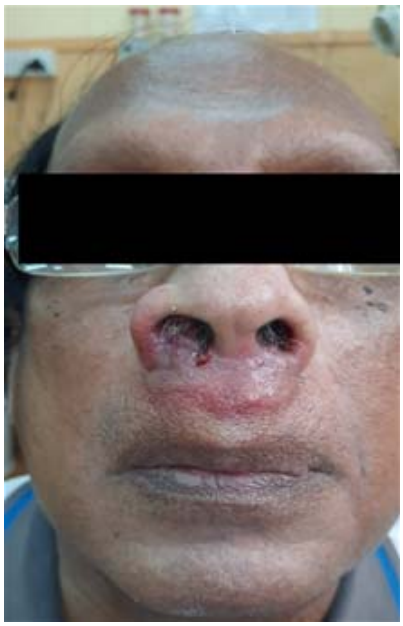

**Figure 1.** *Initial presentation.*

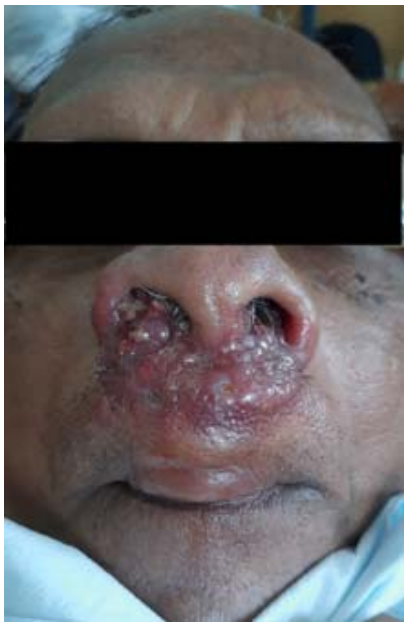

**Figure 2.** *After one month from initial presentation.*

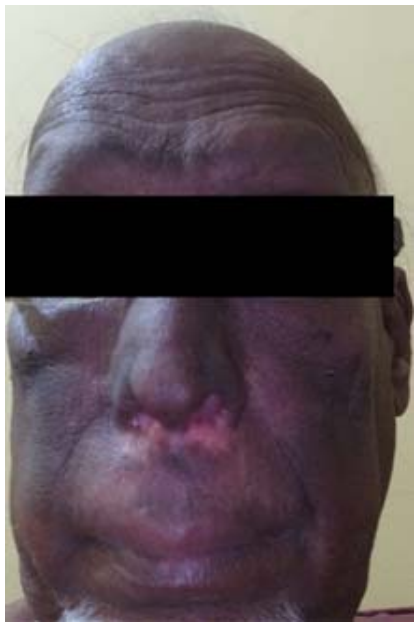

**Figure 3.** *After treatment.*

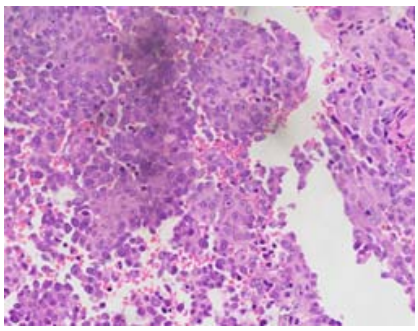

**Figure 4.** *H&E section.*

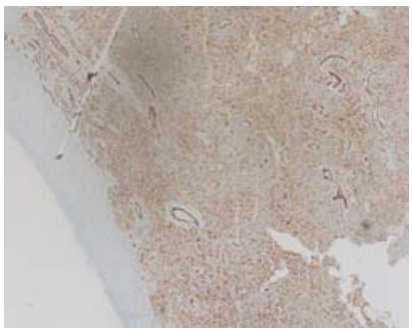

**Figure 5.** *CD 31 positive stain.*

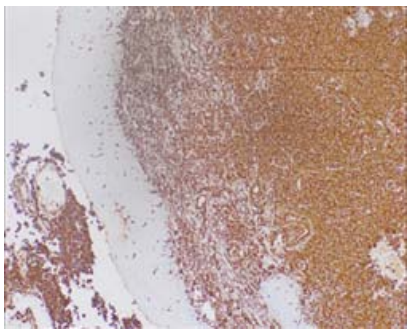

**Figure 6.** *Vimentin positive stain.*

Angiosarcomas are highly aggressive tumour of skin and soft tissue with five year survival of less than 35%[1,2]. Clinical presentation of cutaneous angiosarcoma of head and neck of the elderly include bruise-like lesion, 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. AS on the face and scalp of elderly population – most common type,
2. AS either in association with chronic lymphedema or secondary to a prior surgery (Stewart-Treves syndrome),
3. AS following chronic radio dermatitis or skin trauma and ulceration[3].

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 angiosarcoma 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, immunostaining for markers will help in the diagnosis without any delays.

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

In the context of varying clinical presentations of angiosarcoma it should also include in the clinical

differential diagnoses of infiltrative skin lesions particularly head and neck area in elderly with rapid deterioration of symptoms or signs.

## References

1. Pestoni C, Paredes-Suarez C, Peteiro C, and Toribio J. "Early detection of cutaneous angiosarcoma of the face and scalp and treatment with placitaxel". *Journal of the European Academy of Dermatology and Venereology* 2005; **19**(3): 357-9. View at: Publisher Site | Google Scholar.
2. Aguila LI, Sánchez JL. "Angiosarcoma of the face resembling rhinophyma". *Journal of the American Academy of Dermatology* 2003; **49**(3): 530-1. View at: Publisher Site | Google Scholar.
3. Mobini N. "Cutaneous epithelioid angiosarcoma: a neoplasm with potential pitfalls in diagnosis". *Journal of Cutaneous Pathology* 2009; **36**(3): 362-9. View at: Publisher Site | Google Scholar.
4. Mak KH, Tsang WY. An unusual case of the cutaneous angiosarcoma of the face and scalp. *Hong Kong Dermatol Venerol Bull.* 2003; 209-13. [Google Scholar].