

Primary Cutaneous Angiosarcoma on the Nose in a Patient with Multiple Nonmelanoma Skin Cancers

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ABSTRACT

Cutaneous angiosarcoma is an uncommon, potentially metastatic and highly aggressive vascular tumor that may arise as de novo or be associated with previous radiotherapy. A 70-year-old female with a solitary lesion on the nose was initially diagnosed as actinic keratosis. However, when the recurring lesion at the same region within 6 months was reexcised, the histological diagnosis was definitively established as well-differentiated angiosarcoma. This case was presented in order to increase awareness of this rare malignancy in an uncommon localisation, especially in the light of pathological findings. Moreover, occurrence of cutaneous angiosarcoma within a short period following a previous excision in a patient with a history of multiple nonmelanoma skin cancers was interestingly pointed out.

Keywords: *angiosarcoma; face; nonmelanoma skin cancers.*

INTRODUCTION

Cutaneous angiosarcoma of the head and neck is mostly localised at the scalp of elderly men with poor prognosis.^{1,2} Although a definite diagnosis can be delayed because of the rarity doubled with complex and difficult pathological identification of this highly malignant soft-tissue cancer, early detection and immediate treatment are essential.²

CASE REPORT

A 70-year-old Caucasian female was presented with a plaque on the nose. She was learnt to be operated numerous times for different individual lesions on the face in the last 30 years. Pathological diagnoses of right infraorbital epidermoid carcinoma 10 years ago, left infraorbital basal cell carcinoma nine years ago, and

left infraorbital keratoacantoma and basosquamous cell carcinoma on the forehead eight years ago were noted. She gave no history of previous radiotherapy.

Dermatological examination revealed an ill-defined, erythematous plaque on the right nasal dorsum. Focal thinning of the rete ridges in hyperkeratotic epidermis, enlargement, hyperchromasia, atypia and loss of polarity of the keratinocytes neighboring basal layer focally were detected histopathologically. Solar elastosis and superficial perivascular mononuclear lymphocytic infiltration in dermis were additional findings consistent

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with actinic keratosis.

The patient was represented with an enlarging purple nodule at the same region six months later. On physical examination, a 1.5 cm painless, violaceous nodule was scattered (Figure 1). She had Fitzpatrick type-1 skin with multiple lentigines on the face. Histological examination of the totally excised lesion revealed anastomosis of bizarre vascular spaces lined with atypical pleomorphic endothel-like cells forming a tumoral structure in an irregularly bordered area in dermis (Figure 2a,b). Low mitotic counts (0-5 mitoses per 10 high-power fields) and intense lymphocytic infiltrate were observed. Immunohistochemically, tumor cells stained strongly by CD31, CD34 and factor VIII, and high index of Ki67 were detected. The final diagnosis was considered to be well-differentiated primary cutaneous angiosarcoma. The initial pathologic specimens were reexamined which lacked nuclear pleomorphism and thus, was not found to be related to angiosarcoma.



Figure 1. Close up view of the violaceous fleshy nodular lesion on the nose.

Results of the basic metabolic profile, chest X-ray, computed tomography of the neck and chest revealed no abnormal findings. Following re-excision and reconstruction, the patient was given adjuvant radiotherapy, and she has been surviving for 15 months without any signs of metastasis.

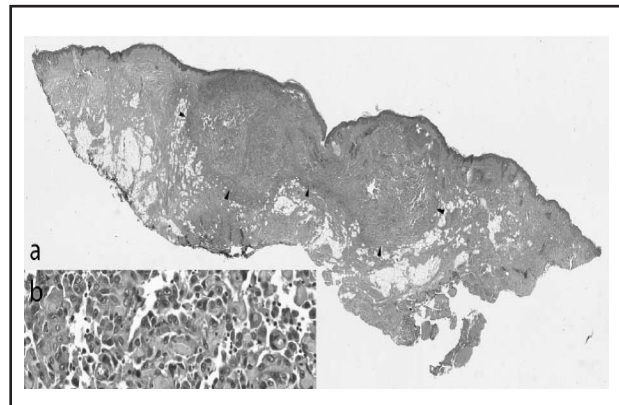


Figure 2. a) Bilobulated nodular tumour mass in dermis signed between black arrowhead, invaded to subcutaneous adipous tissue (whole section, original magnification H&E, X 40). b) Irregular vascular channels lined by atypical endothelial cells (H&E, X 400).

DISCUSSION

Angiosarcoma of the head and neck is a rare tumor of vascular origin originating from endothelial cells primarily localised to scalp, face and neck^{1,3,4} and uncommonly reported at periorbital region and sino-nasal cavities.^{5,6} In women, angiosarcoma was mostly located to the scalp as ulcerated lesions.⁴ The nose is a rare site of presentation and a few cases of angiosarcoma reported on the nose were male patients.^{4,7,8} In a series of 14 patients reported by Morrison¹ and of three cases by Hanke et al⁸ only one female in each was presented with angiosarcoma on the nose. Therefore, the nodular angiosarcoma in a female on the nose was a seldom site reported to date.

Possible relevance of predisposing factors including history of irradiation, local trauma, insecticides, thorium dioxide contrast and vascular malformations were reported in etiology of angiosarcoma.^{1,4,9} Because these tumors are more frequent in Caucasians, sun exposure has also been implicated in its etiology.^{1,3,4} In contrast, some authors reported low rate of skin carcinomas in patients with angiosarcoma.⁹ However, to the best of our knowledge, any case of cutaneous angiosarcoma on the face with multiple nonmelanoma skin cancers and actinic keratosis was not reported. The short period between the two excised lesions alerted us about the possible coincidental association of angiosarcoma and actinic keratosis in the presented case. Nevertheless, lack of histopathological findings of angiosarcoma in the initial specimen and subsequent occurrence of the nodular lesion diagnosed as angiosarcoma after the

previous excision made us consider the effect of local trauma due to the operation.

Variable clinical manifestations of cutaneous angiosarcoma include asymptomatic bruise-like lesions, purple nodules, chronic edema or ulcers.⁴ Enlarging nodular lesion of 5 cm was mostly reported³ while ulceration was found to be the most common presentation in another series.⁹ Several benign conditions such as rosacea, pyogenic granuloma, erysipelas, contact dermatitis and angioneurotic oedema might be mistaken.^{4,5,7} Arteriovenous malformations, Kaposi's sarcoma and nodular melanoma should also be considered in differential diagnosis.⁷ Therefore, histological examination is of crucial importance and difficulty in pathological assessment may lead to delay in diagnosis.⁴ Angiosarcoma contrasts pyogenic granuloma with nuclear pleomorphism by dissecting through connective tissue to create irregular vascular spaces. Kaposi's sarcoma contains cellular zones of spindle cells forming slit-like spaces while anastomosis of vascular spaces lined with atypical cells was diagnostic of angiosarcoma as was the case.² Sharing certain histological components, angiosarcoma may be differentiated with positivity of CD31 and factor VIII from nodular melanoma and hemangiopericytoma.

Five year survival was reported to be 10-30% in patients with angiosarcoma.^{3,4,10} Although the

only significant prognostic factor was supposed as histological grade of tumor in a series,³ the results of most series have indicated the only expressive correlation for tumor size with the survival. Mostly worse prognosis was detected for bleeding and painful lesions larger than 5 cm.^{4,9} In contrast to the finding of predominance of poor differentiation in nodular lesions,⁴ our case presented with a nodular lesion was a well-differentiated angiosarcoma. Prominent lymphocytic infiltrate was suggested to have a more encouraging outcome⁷ while only mitotic counts were found to be significant for prognosis by Naka et al.¹⁰ Mild degree of mitotic counts, intense lymphocytic infiltrate in this asymptomatic, small tumor were the factors promising a better survival of the presented patient. Moreover, improved survival in patients with central facial localisation was suggested. Three patients presenting with nasal angiosarcoma survived 4 to 7 years.⁸ The female case of angiosarcoma on the nose reported in the series of Morrison et al¹ was alive 10 years after diagnosis.

In conclusion, early suspicion and prompt histological study of an enlarging nodular lesion on the nose in elderly should be crucial keeping the diagnosis of angiosarcoma in mind. The presented case of angiosarcoma in a patient with multiple nonmelanoma skin cancers and actinic keratosis raised the possibility of actinic damage to fair skin might be suggested in etiology.

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