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Cutaneous angiosarcoma presenting as an unusual facial bruise

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Abstract

Angiosarcoma is a rare vascular tumour accounting for 2% of soft tissue sarcomas, which together represent less than 1% of all cancers. Although well described in specialist literature, it is unusual in general medical practice. We describe a case in which the initial appearance as a bruise gave rise to diagnostic uncertainty and delay.

Keywords: *angiosarcoma, sarcoma, facial bruising, rash, elderly*

Case report

An 83-year-old woman presented with symptoms and signs consistent with a posterior circulation cerebrovascular event. Computed tomography (CT) imaging confirmed a recent infarct in the left cerebellar hemisphere, with generalised, small vessel, ischaemic change. She was found to have a raised cholesterol, and diabetes was confirmed on formal testing. Clopidogrel 75 mg/day and atorvastatin 10 mg/day were started as well as dietary adaptations to address her diabetes. With rehabilitation in a multidisciplinary stroke unit, there was excellent progress in mobility and functional independence.

One month after admission, she developed apparent bilateral bruising on the bridge of the nose with erythema of the anterior scalp. These were initially attributed to pressure from spectacles and hair curlers, respectively, although there was no actual history of trauma. The platelet count and clotting studies were normal. Over the next 10 days, the lesions extended, and those on the bridge of the nose darkened, becoming violaceous. She developed peri-orbital oedema and progressive skin thickening in the maxillary regions with sub-mandibular and cervical lymphadenopathy. She remained systemically well and had no other skin lesions nor generalised lymphadenopathy.

ENT examination revealed no abnormality of the anterior nares, mouth or nasopharynx and chest radiograph revealed no mediastinal lymphadenopathy. The LDH was normal, but the ESR had risen from 22 to 40. There was no leukocytosis and no evidence of a serum paraprotein. Open biopsy of the glabellar lesion and punch biopsy from the edge of the spreading scalp lesion revealed diffusely infiltrating, malignant cutaneous angiosarcoma, invading the subcutis. CT of the neck and thorax confirmed extensive cervical lymphadenopathy with compression of both internal jugular veins. There was no mediastinal or axillary lymphadenopathy and no evidence of pulmonary metastases.

Over the next 2 weeks, the skin lesions and lymphadenopathy progressed rapidly (see Figure 1). The tumour was considered unsuitable for surgery or radiotherapy and the patient unsuitable for chemotherapy. Discharge home was achieved with the support of the palliative medicine team, but she required re-admission to a hospice because of bleeding, despite daily dressings and topical adrenaline, and visual impairment due to peri-orbital oedema. She died shortly afterwards, 10 weeks after the first appearance of the skin lesions.

Discussion

Cutaneous angiosarcomas are highly malignant tumours, occurring most commonly in elderly men. Fifty per cent occur in the skin of the head or neck, particularly the scalp or upper face, as in our patient. There is frequently multifocal disease. The aetiology is uncertain but some cases are associated with prior irradiation and chronic lymphoedema.

Most patients present with an enlarging bruise, a blue-black nodule or non-healing ulceration, but unexplained facial oedema may be a presenting sign. In the early stages it is commonly confused with traumatic bruising, as in our

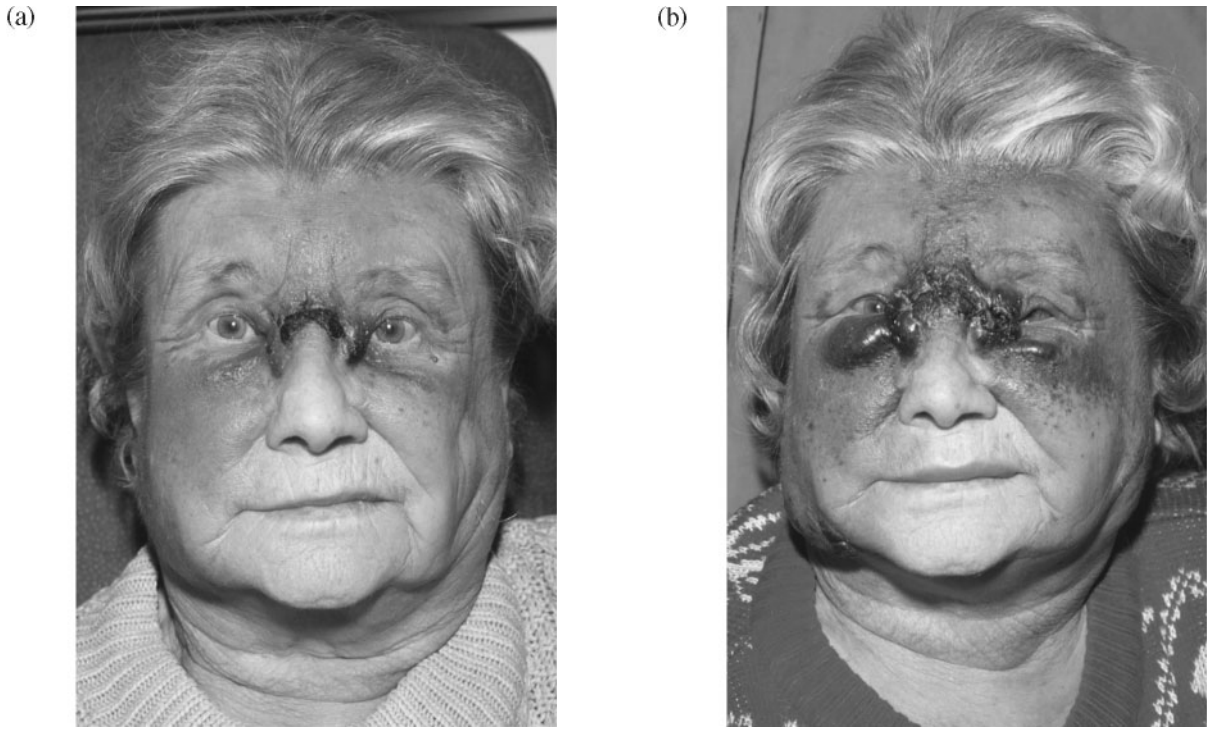

Figure 1. Photographic illustration of the lesions demonstrates the rapid progression (a) within 4 weeks of presentation and (b) 2 weeks later.

case, but can also be mistaken for allergic reactions, cellulitis or rosacea [1]. The differential diagnosis is wide, but includes Kaposi's sarcoma, sarcoidosis with lupus pernio, dermatomyositis, cutaneous lymphoma and fixed drug eruptions. The diagnosis should be confirmed on biopsy, revealing endothelial cells demonstrating cytological atypia, and well-formed vascular spaces. Angiosarcoma cells typically extend far beyond the clinically defined margins, making radical surgery difficult. Their management should be undertaken by a specialist multidisciplinary sarcoma team.

The prognosis is poor, even with aggressive treatment. The median survival is reported to range from 15 to 24 months and the 5-year survival from 12 to 33% [2]. Size is the most important determinant of survival [2, 3]. Small tumours with no spread beyond the subcutaneous tissues have the best prognosis, independent of tumour grade [2, 3]. Wide resection is the treatment of choice for localised tumours and can be supplemented by post-operative external beam radiotherapy or chemotherapy [3, 4]. Routine exploration of the neck is not recommended [2] but some authors recommend regional cervical node dissection at the time of primary excision, for high-risk larger tumours [3]. For locally advanced tumours, the role of radiotherapy is unclear, particularly in the presence of lymph node or distant metastases.

Chemotherapy is of limited benefit. Until recently, different pathological types of soft tissue sarcoma were treated on common protocols. In Europe, single agent doxorubicin is the standard treatment [5, 6]. There are reports of prolonged survival with cutaneous angiosarcoma treated with intra-arterial or systemic doxorubicin [2]. Because liposomal doxorubicin is licensed for the treatment of Kaposi's sarcoma, it has been used in combination with radiotherapy in poor prognosis angiosarcoma [7, 8]. Intralesional interferon alpha 2b and interleukin 2 have also been used [9]. Response to treatment with paclitaxel has been reported by several authors, unfortunately without prolonged survival benefit [10, 11]. Increased understanding of their tumour biology has lead to the development of novel targeted treatments for some sarcomas [12]. Currently there is interest in the use of vascular endothelial growth factor (VEGF) receptor antagonists for angiosarcomas [13].

Key points

- The initial non-specific appearance of this uncommon tumour may lead to diagnostic delay with an adverse effect on prognosis.
- An unexplained progressing bruise of the face or scalp, as in the case described, should alert the clinician to the possibility of angiosarcoma.
- Early biopsy of suspicious lesions, while still limited in size, allows for the possibility of resection and improved outcome.

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Hemihyperhidrosis in cerebral infarction

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Abstract

Increased sweating on the hemiparetic side in cerebral infarcts is not a common clinical finding. The onset, severity and duration of symptoms can vary. The structural lesion responsible for this is a subject of conjecture. We present the case of a 66-year-old man who developed hemihyperhidrosis secondary to a cerebral infarct.

Keywords: *hemihyperhidrosis, cerebral infarct, CVA, stroke, hyperhidrosis, elderly*

Case report

A 66-year-old male smoker on treatment for essential hypertension and dyslipidaemia presented with a sudden onset of left-sided weakness affecting his face, arm and leg associated with difficulty in swallowing. There was no evidence of any autonomic dysfunction. A diagnosis of partial anterior circulation stroke was made. His CT brain scan revealed a large hypodense area in the right fronto-parietal region extending into the temporal lobe, which confirmed the clinical diagnosis of a right middle cerebral artery territory infarct.