Case report

ANGIOSARCOMA OF THE FACE AND SCALP

G. Torlone, A. Rossodivita, E. Caracciolo, A. Felli and S. Chimenti

ABSTRACT

The authors describe the case of a 72-year-old man, who presented two infiltrated and ulcerated plaques of the scalp and multiple angiomatous lesions on the face. The histological examination showed typically angulated and irregular spaces lined by cells which formed one or more layers and the typical bulging of the malignant endothelial cells into vascular lumens.

A diagnosis of cutaneous angiosarcoma unassociated with lymphedema of the scalp and face was made. A multi-agent cytotoxic chemotherapy was employed as treatment strategy. The authors discuss the nosologic localization of angiosarcoma and underline the therapeutic modalities of this rare neoplasm.

KEY WORDS

angiosarcoma, scalp, face, skin, a unassociated with lymphedema, therapy

INTRODUCTION

Angiosarcoma (AS) is a rare malignant neoplasm, most commonly arises in the skin and superficial soft tissues, although deep tissue and viscera can be affected too. The aspect and behavior of this tumor depend on location, and in literature as has been reported in various organs (1). Hence, AS is more properly considered as several closely related tumors rather than as a single entity and has been classified by Enzinger and Weiss into five groups: 1) cutaneous angiosarcoma unassociated with lymphedema; 2) cutaneous angiosarcoma associated with lymphedema (Lymph-angiosarcoma); 3) angiosarcoma of the breast; 4) radiation-induced angiosarcoma; 5) angiosarcoma of deep soft tissue. AS of the face and scalp of the elderly forms a distinctive subgroup of cutaneous AS unassociated with lymphedema (1).

AS is usually located on the head and neck particularly the scalp and upper forehead (1). Clinically three types are described: ulcerating, diffuse superficial spreading, and nodular (2,3). Clinically is quite different from classical Kaposi's sarcoma who appears as multiple subcutaneous plaques or nodules that show a typical violaceous hue, usually affecting the lower extremities of elderly men, with a male predominance. Tumor's behaviour is indolent with gradual increase in number of lesions associated with lymphedema, visceral lesions occurs late, often discovered at autopsy (4,7).

Secondary diffusion to regional lymph nodes and blood stream occurs in the late stage of disease and death can result due to local spread of the lesion rather than from metastases. In some cases distant metastases can occur. Sites affected

by metastases are most commonly cervical lymph nodes, followed by lung, liver and spleen (1).

The survival time reported is about 20 months after diagnosis. Only 12% of patients survive 5 years or more (4). Patients with lesions smaller than 5 cm in diameter seem to respond better to the treatment, and this event emphasizes the relevance of the early diagnosis (23). A few cases of spontaneous regression of AS, with the complete resolution without any therapy, have been reported (5). Other factors such as sex, location, and histological grading could not be correlated with.

CASE REPORT

A-72-year-old man had two ulcerated plaques localized on the scalp and purple-reddish angiomatous lesions on the face, of about eight months duration. Clinical examination showed an ulcerated, hemorrhagic and infiltrated lesion on the right side of the scalp extending into the temporo-parietal area; the largest diameter was of 10 cm (Fig. 1). A similar lesion measuring 3 cm at its largest diameter was present in the occipital region. In addition multiple angiomatous, symmetrical lesions were seen in the frontal and temporal areas. The lesions were not painful. The patient did not mention a preceding trauma. The lymphnodes were not palpable, the liver and spleen were not enlarged.

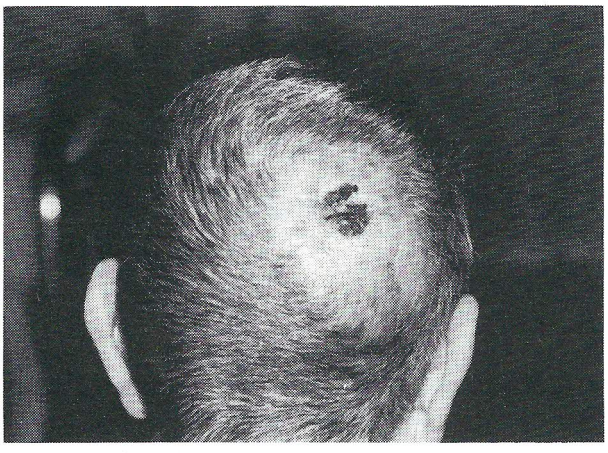

Figure 1. *Clinical aspect of the ulcerated lesion on the scalp*

The x-ray chest examination, liver sonography, skull CT scan as well as laboratory tests (blood chemistry, hematology, immunologic assays, anti-HIV test and urine analysis) were all within the normal range.

One biopsy was taken from the largest nodule on the scalp and a second one from the face. The hematoxylin-eosin staining of the first lesion revealed a proliferation of poorly differentiated spindle cells displaying atypia and increased mitotic activity (Fig. 2) in the dermis. Spindle-shaped tumor cells were arranged in bundles throughout the entire dermis spreading in multiple directions. There was an increased number of vessels with protruding endothelial cells and an abundant extravasation of erythrocytes among the collagen bundles. The second biopsy showed a well expressed angiomatous pattern with a large number of newly formed irregularly shaped vessels. The vascular spaces were devoid of erythrocytes and numerous endothelial cells of tomb-stone appearance were noted (Fig 3).

The immunohistochemical investigations were performed on the paraffin-embedded material using the standard three-step immunoperoxidase technique. Monoclonal and poly-

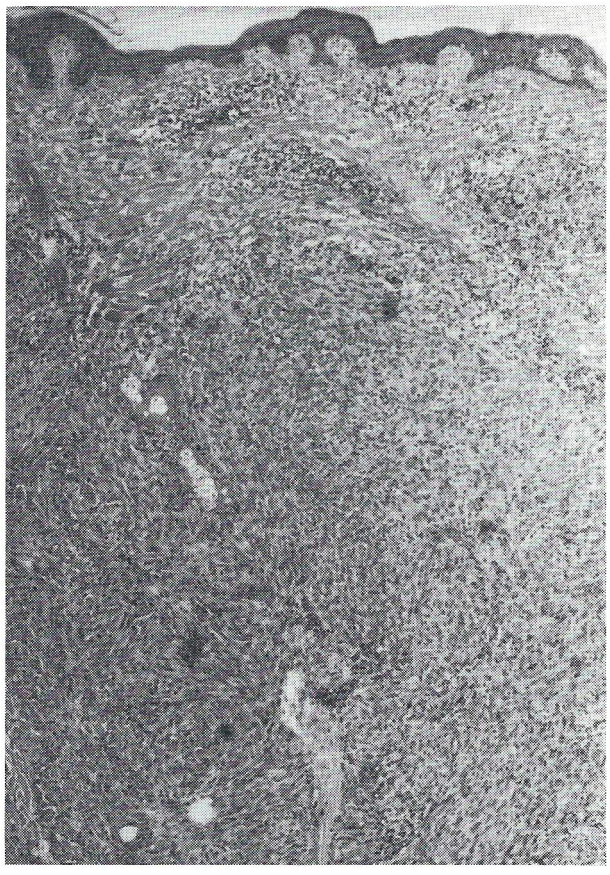

Figure 2. Scanning magnification shows proliferation of poorly differentiated spindle cells in the dermis. (hemato-xylin-eosin x 10)

clonal antibodies were used, the results are summarized in table 1. The CT scan of the chest revealed the presence of two micrometastases in the left side of the lung. Considering all the afore mentioned data the diagnosis of angiosarcoma was made and a systemic chemotherapy with epirubicin ifosfamide was introduced.

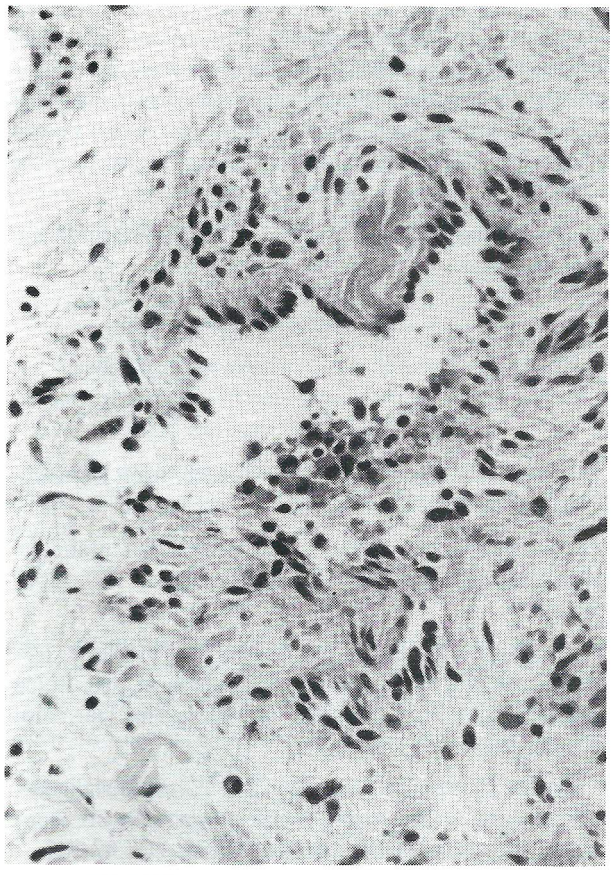

Figure 3. Higher magnification illustrates vascular spaces with erythrocytes and numerous cells with "tomb stone" like appearance. (hematoxylin-eosin x 100)

DISCUSSION

AS of the face and scalp usually occurs in white men, though there have been occasional reports of the disease in black and oriental people (11-14). Some authors underlined the possibility that actinic damage to fair skin might play a role in the pathogenesis of this neoplasm (1,4).

Typical AS is histologically composed of undifferentiated spindle cells and of a network of vascular channels in the dermis, ranging in size from small capillaries to sinusoidal spaces, lined by abnormal endothelium. The endothelium may be expressed as a single layer of cells or formed by either papillary projections or solid masses protruding into the vascular lumen. The endothelial cells show a variable degree of atypia and pleomorphism. Mitoses are usually few. The endothelial origin of the tumor is proved by use of reticulum staining, which allows to differentiate AS from hemangiopericytoma (16).

According to cellular differentiation the grade of malignancy can be determined. Increasing variability in the size of vascular channels associated with more marked pleomorphism, mitotic activity and solid spindle cell areas are believed to be features of progression and greater malignancy. While some authors have attached prognostic significance to histological features, others have been unable to identify such a correlation (1,13).

The proof that spindle cell tumors with a vascular component are of endothelial origin may be confirmed by immunocytochemical staining with antibodies to factor VIII-related antigen (16).

Differential diagnosis includes benign hemangioma, angiolymphoid hyperplasia with eosinophilia, hemangiopericytoma, Kaposi's sarcoma and spindle cell squamous carcinoma (1,7,16).

Histogenetic studies performed in AS of the face and scalp are yet inconclusive. We confirmed the observation of Burgdorf et al. (17) that AS for the most part lacked the stainable factor VIII RAG, but occasionally cells of malignant appearance were positive (6,17,18). Despite numerous electron microscopic (18,19) and histochemical (20,22) studies, there does not exist an accepted method by which malignant tumors of blood vascular endothelium could be distinguished from tumors of lymphatic endothelial origin (18,23). Maddox and Evans looked at prognostic factors and suggested that smaller tumors and those with a marked cellular infiltrate did rather better, but that the tumor site was the most telling feature (23).

Due to the rarity of AS and the lack of significant experience a clear consensus for treatment has not been reached.

Table 1. Immunohistochemical investigations

ANTIBODY	RESULT	SPECIFICITY
Anti-S 100 protein	-	Melanocytes, nerves
Anti-factor VIII A	+/-	Endothelial cells
BMA-120	+	Endothelial cells
LCA	-	Leukocytes common antigen
PCK	-	Pan-cytokeratines

Some authors advocate surgery if the lesions can easily be removed, with or without postoperative radiation therapy (10,24).

Others support the use of primary radiation therapy with surgery held in reserve (4,10). The use of chemotherapy as primary treatment or as an adjuvant to surgery has not been investigated enough (10,25). However, some authors suggest systemic or intra-arterial chemotherapy for patients with non-operable lesions (10,25,26).

In our case the lesions were extensive and coexisted with two pulmonary micrometastases. The authors believe to obtain a better result with systemic chemotherapy. After a follow-up of 9 months the patients seems to respond to the treatment with regressing of skin lesions and a partial reduction of pulmonary micrometastases revealed by chest CT scan.

Further studies using epirubicin and ifosfamide are recommended.

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