MALIGNANT HAEMANGIOENDOTHELIOMA OF SKIN
(A case report)

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Abstract

A rare case of haemangioendothelioma of the skin in an old lady, involving the commonest known location viz neck, face and scalp is reported along with a brief review of literature.

Malignant haemangioendotheliomas may arise from endothelium of vessels of almost any internal organ. These have been most frequently reported in mammary glands, liver, bones and striated muscles[1]. Primary angiosarcoma of the skin is rare. Landell[2] reviewed the world literature and found 177 acceptable cases of angiosarcoma; only 18 of which originated in the skin and subcutaneous tissue. Stewart and Treves[3] reported six cases of lymphangiosarcomas arising in chronic oedematous upper extremities following radical mastectomy. Jones et al[4] reported nine patients with malignant angioendothelioma of the face and scalp, all of which developed in elderly individuals. Reed[5] reported six cases. The majority of the reports have been of one case or a small number of cases. The authors came across only one case in the last ten years and hence it is reported.

Case Report

The patient was an eighty year old female. Her illness began in April 1980. It started as dusky black spots on the right side of neck behind the pinna of the ear and began spreading over the face and neck. Within two months, the lesions became raised, dusky blue in colour and involved the face, scalp and both sides of neck. To start with it was painless but within 15 days it became painful. Patient did not give any past history of trauma or radiotherapy to this region.

On examination, skin of the face, both sides of neck, front and sides of scalp was warm, diffusely dusky red in colour and raised (Fig. 1). Firm tender nodules were palpable on both sides of neck in the raised area. Similar black spots were seen on the upper chest. There was no significant lymphadenopathy.

ESR (Wintrobe) was raised to 60 mm in the first hour. Haemoglobin was 10 gms percent at the time of biopsy; other haematological investigations being normal. After the biopsy, patient was investigated for diabetes, pulmonary tuberculosis and ischaemic heart disease and all investigations gave results within normal limits. LE cell preparations were negative. Serological tests for syphilis were negative. Clinical diagnosis of scleroderma was made and biopsy specimen was taken

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Received for publication on 24—11—80.

Fig. Showing dusky colouration of the skin of face, side of neck and scalp.

from left side of neck, behind the pinna of the ear.

Biopsy examination, under low power revealed an atrophic epidermis, a diffuse cellular infiltrate and clefts between the proliferating cells in the deep dermis (Fig. 2). Higher magnification revealed the collagen disrupted by wide arborising channels of poorly formed blood vessels lined by pleomorphic endothelial cells. The atypical endothelial cells were large, hyperchromatic and showed prominent variation in size, shape and staining characteristics and some showing numerous abnormal mitotic figures. Some of these cells were seen budding into the lumen (Fig. 3). Reticulin stain confirmed the vasogenic nature of the tumour endowed with poorly constructed walls. A diagnosis of malignant haemangioendothelioma was made. Radiotherapy and chemotherapy treatment were recommended. Surgical excision could not be undertaken because of wide extent of the lesion. Patient is now being followed up at frequent intervals.

Discussion

Primary angiosarcoma of the skin has been reported to occur mainly in elderly individuals[5,6,7]. Lesions arising on the scalp or face, especially in the older age group, tend to show histological features of undifferentiated angiosarcoma and to behave clinically in an aggressive manner[8]. Nine out of 13 patients over the age of 60 reported by Girard et al[8] had their lesions on the scalp and face as in our patient. The tumours in this series were usually multiple, ulcerative and spread by local infiltration showing no response to treatment. Similar lesions have been reported in significant numbers[5,6]. In our patient,

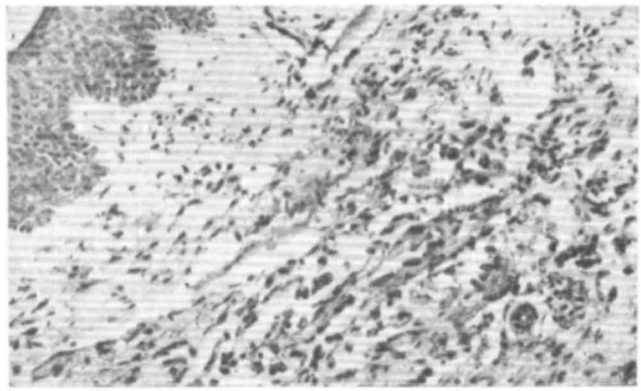

Fig. 2

(Low power) Showing atrophy of epidermis shows cellular infiltration and clefts between the prolifevating cells in the deep dermis. (Magnification 150)

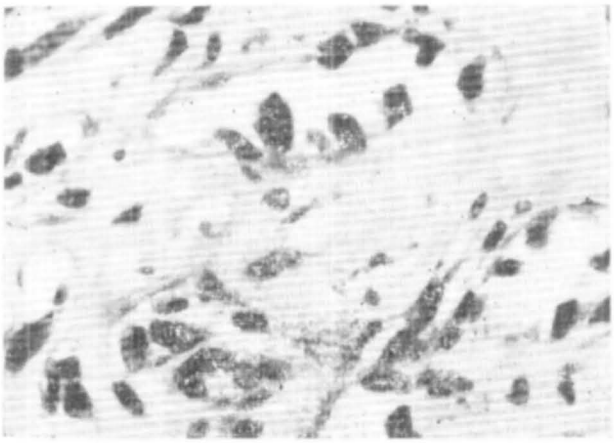

Fig 3
(High power) Shows collagen disrupted by wide arborising channels of poorly formed blood vessels lined by pleomorphic endothelial cells. Some of these cells are budding into the lumen. (Magnifiction 600)

though the tumours were multiple, they had not ulcerated or spread.

The etiology of cutaneous angiosarcomas is unknown. Trauma has been mentioned as a preceding event in the onset of tumours in almost all reported cases. In Girard et al's[8] series only three patients had a known injury at the site of lesions. Our patient also did not give any history of injury or treatment by radiation to the involved area, prior to the onset of the disease. Review of the literature gives no significant information regarding relationship to hereditary factors, geographical location or other possible causative factors. In the same series[8], one patient had active luetic infection at the time of the onset of the lesion while two others had diabetes, glaucoma and anginapectoris. One patient had past history of pulmonary tuberculosis. Our patient did not have diabetes, pulmonary tuberculosis or ischaemic heart disease. Most of the cases reported literature showed extensive local spread with tissue destruction and mutilation. The sites of metastasis in order of frequency were the regional lymph nodes, lungs and heart.

The histologic differentiation between low grade angiosarcoma and pseudo-pyogenic granuloma is difficult [9,10]. The presence of fronds of endothelial cells giving rise to a papillary configuration is an essential feature in making a diagnosis of low grade angiosarcoma. Along with pseudopyogenic granuloma, papular angioplasia was also considered in differential diagnosis. Papular angioplasia may be difficult to differentiate from this condition only on histological examination[11]. The term malignant should be reserved for biological behaviour of abnormal tissue and not for its cytological characteristics. In our patient disease had progressed rapidly so that the biological behaviour was that of a malignant lesion. The "sprouting" or "hiding" of the capillaries is not prominent in benign haemangioma. Livingston and Klemprer[12] and Bardwill et al[13] described these "sinuses with the knob like stroma projections" and referred to this as a sign of malignancy. The presence of hyperchromatic cells in the stroma and other hyperchomatic cells lining the imperfectly formed vascular spaces also favour a diagnosis of angiosarcoma. Usually pleomorphism is minimal and mitotic figures are rare. In this case all the evidences of malignancy were present. Reticulin stain demonstrated reticular pattern

of an intensely vascular lesion. Atypical endothelial cells were seen near or within poorly formed vascular spaces but always within the reticular framework.

References

1. Stout AP : Haemangioendothelioma, a tumour of blood vessels featuring vascular endothelioma cells, Am J Surg, 1943; 118 : 445-464.
2. Landells JW : Malignant tumours of heart and blood vessels, In Cancer Vol 12, London, Butterworths, 1958; pp 512-524.
3. Stewart FW & Treves N : Lymphangiosarcoma in postmastectomy lymphedema, Cancer 1948 ; 1 : 64-81.
4. Jones WA, Helwing EB & Harman LW : Oxidative enzyme activity in the skin of patients with psoriasis, J Invest Derm, 1965; 44 : 189-195.
5. William-Jones E : Malignant angioendothelioma of the skin, Brit J Derm, 1964; 76 : 21-39.
6. Reed RJ, Palomeque FE, Hariston MA, et al : Lymphangiosarcoma of the scalp, Arch Derm, 1966; 94 : 396-402.
7. Walter F, Lever & Gandula Schaumburg Lever : Histopathology of skin, JB Lippincott Company, Philadelphia, Tronto, Vth Edition 1979; pp 617.
8. Girard C, Johnson WC and Graham JH : Cutaneous angiosarcoma, Cancer, 1970; 26 : 868 - 883.
9. Peterson WC, Fusaro RM & Goltz RW : Atypical pyogenic granuloma, Arch Derm, 1964; 90 : 197-201.
10. Wilson-Jones E, Bleehan SS : Inflammatory angiomatous nodules with abnormal blood vessels occurring about the ears and scalp (pseudo or atypical pyogenic granuloma), Brit J Derm, 1969 ; 81 : 804 - 816.
11. Wilson-Jones E and Marks R : Papular Angioplasia, Arch Derm, 1970; 102 : 422-427.
12. Livingston SF and Klemperer P (1926) Quoted by Girard C, Johnson WC and Graham JH (1970).
13. Bardwill JM, Mocega EE, Butler JJ, et al: Angiosarcomas of the head and neck region, Amer J Surg, 1968; 116 : 548-553.

TRUE or FALSE

Immunological responses to infection with Acarus scabeii is possible as evidenced by immediate type reactions with intracutaneous tests.

(See Page No. 242)