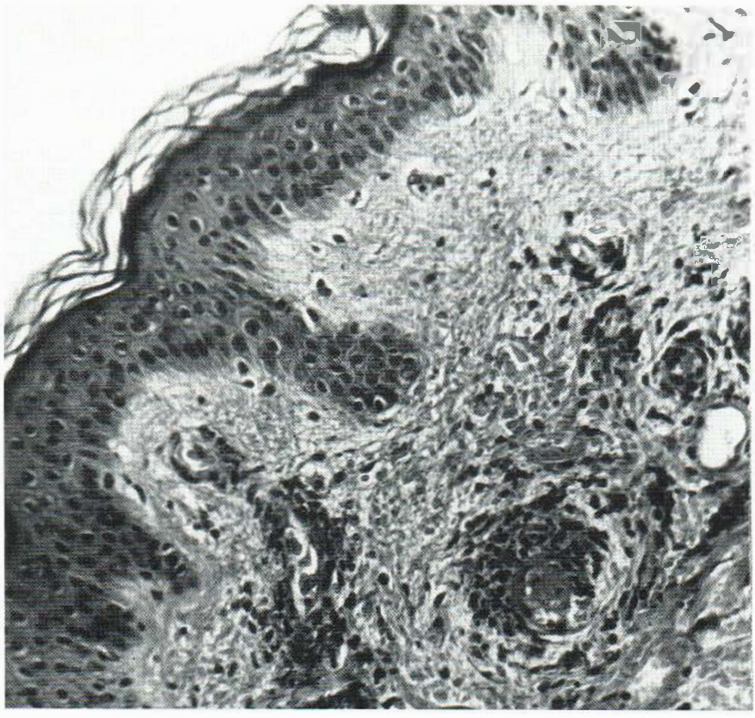

Fig. 2. Skin histopathology in case No. 2. For description, see text.

Rechallenge with the drug was not done as this procedure caused a fatal periarteritis in one case previously described (6). In all other cases, PTU was only indirectly proved to be the cause of the vasculitis, as the skin symptoms subsided when the medication was discontinued. The polyarthralgias seem to be a more persistent problem, as it may continue for several months after discontinuation of therapy (8). Karbimazol can replace PTU without further occurrence of cutaneous vasculitis (8) despite the basic structural similarity of the two drugs. However, it may be safer to discontinue thionamide drug therapy entirely (7).

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Malignant Clear Cell Hidradenoma: A Case Report

D. B. Czarnecki,[1] Ian Aarons,[2] J. P. Dowling,[3] B. Lauritz,[1] P. Wallis,[2] and E. H. Taft[1]

[1]*Dermatology Unit and* [2]*Pathology Department, Prince Henry's Hospital, South Melbourne, and* [3]*Pathology Department, Royal Melbourne Hospital, Melbourne, Australia*

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Abstract. A 73-year-old man developed a malignant clear cell hidradenoma on the nose. Although it was treated

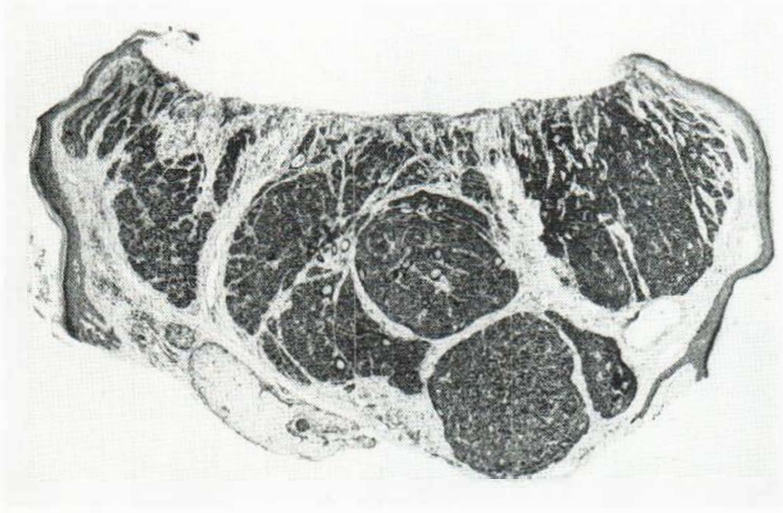

Fig. 1. Lobulated epithelial tumour masses extensively infiltrating the dermis. The overlying epidermis is normal. H & E, ×20.

solely with diathermy and curettage it did not recur. These rare tumours are histologically malignant but do not always behave aggressively. In some patients local growth and recurrence may occur, while in others the tumour metastasizes widely. There is no way of predicting how these tumours will behave.

Key words: Malignant clear cell hidradenoma; Local disease; Metastases

Clear cell hidradenomas arise from the eccrine sweat duct (1, 2). These tumours are composed of tubular lumina and sheets of epithelial cells, a varying portion of which is glycogen filled and has a clear cytoplasm when stained with hematoxylin and eosin. Because these tumours have no distinctive clinical features, the diagnosis can only be made histologically. Most such tumours are benign, but local recurrence often follows incomplete excition. Malignant tumours are rare. They can remain strictly localized or disseminate widely.

The following is a report of a locally aggressive malignant clear cell hidradenoma. The literature on the subject is reviewed.

CASE REPORT

A 73-year-old man was first seen in early 1980 with a slowly growing, asymptomatic papule on the left ala nasi which had been present for about 8 months. There was no previous history of skin disease but a bladder carcinoma had been resected 13 years previously.

The lesion was dome shaped, flesh coloured and measured 0.6 centimetres in diameter. It was not ulcerated, but telangiectasia was evident on the surface. As the exact nature of the lesion was not known it was treated by shaving flush with the skin surface followed by diathermy and curettage to the base.

After histological examination and diagnosis the patient was referred for plastic surgery, but his poor physical health, due to severe mitral valve disease and chronic obstructive airway disease, prevented surgery.

A careful follow-up of the patient was carried out until he died of heart failure 10 months later. There was then no evidence of recurrence of the tumour.

Histology

The epidermis was intact although attenuated by the tumour which extended well into the reticular dermis. It was composed of irregular nodular masses of closely-packed neoplastic cells arranged mostly in sheets. There were cord-like structures and poorly formed acini as well as duct-like structures lined by highly irregular epithelial cells. The cell cytoplasm was characteristically clear and contained glycogen (PAS-positive, diastase-sensitive). The cell boundaries were distinct and well demarcated. The nuclei were generally small and situated mostly centrally. They were hyperchromatic and showed appreciable pleomorphism. Mitoses were evident in modest numbers, some being quite bizarre. In addition there were numbers of multinucleated giant cells. Foci of lymphocytes and plasma cells were scattered throughout the supporting stroma. See Figs. 1, 2 and 3.

DISCUSSION

Malignant clear cell hidradenomas can only be diagnosed by histological examination as there are no distinctive clinical features. The tumours are slowly growing, asymptomatic, flesh coloured papules or nodules. Most patients have been elder-

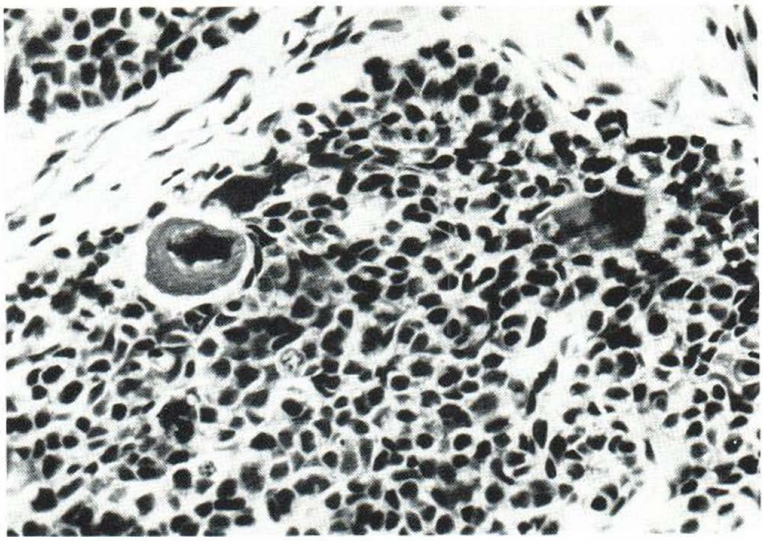

Fig. 2. There are two tumour-cell types, one with small dark-staining nuclei, the other having large, central, paler nuclei. Bizarre forms are present. H & E. ×280.

ly and the head is the site most frequently involved. Trauma from incomplete excision, diathermy, or radiotherapy causes accelerated growth. However, the tumours may remain localized (3, 4), or they may metastasize widely (5–8). There is no way of predicting how an individual lesion will behave.

Wide local excision is the treatment of choice. If the lesion is large, prophylactic lymph node dissection is indicated (Headington, personal communication, April 1980). Radiotherapy is contraindicated because of its effect on growth rate. Chemotherapy has not been tried.

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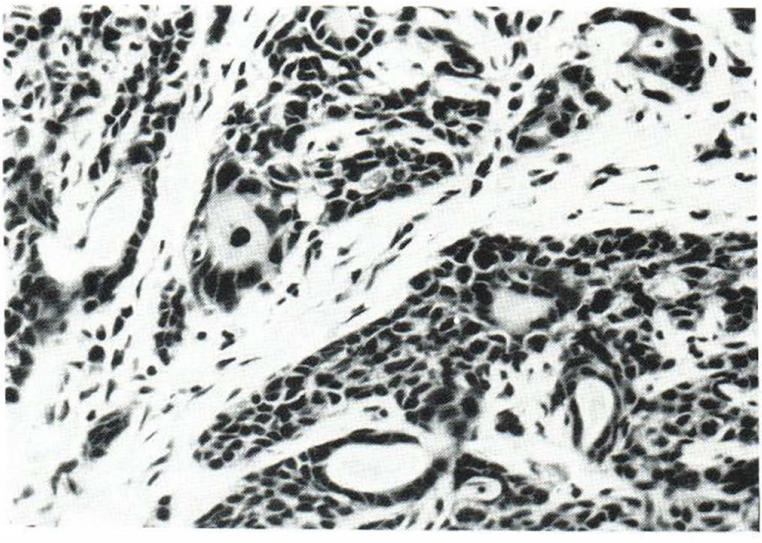

Fig. 3. An area of tumour presenting number of irregular ductal structures lined by a single layer of neoplastic epithelial cells. Some lumina contain faintly acidophilic granular material. H & E. ×280.

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The Klippel Trenaunay Weber Syndrome Presenting with Cutaneous Bleeding

S. J. Adams and W. J. Cunliffe

Department of Dermatology, St. Jame's University Hospital, Leeds, England

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Abstract. A patient with the Klippel Trenaunay Weber syndrome presented with prolonged bleeding from the finger tips during strenuous exercise. Tissue fibrinolysis studies showed increased fibrinolytic activity suggesting that the blood vessels in the affected limb may be functionally as well as anatomically abnormal.

Key words: Klippel Trenaunay Weber syndrome; Bleeding; Fibrinolysis

The Klippel Trenaunay Weber syndrome as described by Klippel & Trenaunay in 1900 (6, 7) is characterized by vascular naevoid abnormalities confined to a limb. It is associated with hypertrophy which may be gross, and varicose veins.

CASE REPORT

The patient is a 43-year-old building labourer whose presenting complaint is that when using his arms energetically (when digging, for example) he may bleed profusely from the tip of his right forefinger. The bleeding usually stops after sustained pressure has been applied to the finger, but on three occasions he has attended the casualty department because of persistent bleeding. On each visit the bleeding was such that cautery was required to quench it.

On examination the patient was healthy except for the physical signs in his right arm which was enlarged, and showed marked varicosities throughout the whole limb.

In addition he had multiple teleangiectases extending from the neck throughout the limb but which were very heavily grouped on his fingertips (Fig. 1). Blood pressure was 130/85 mmHg in both arms. No arterial or cardiac bruits were present but a venous hum was audible over the right arm.

Investigations

Normal hematopoietic, renal and liver functions were revealed. Chest X-ray was normal and soft tissue radiograph failed to demonstrate any abnormal vascular channels in the bones of the right arm. Twenty-four hour urinary oestriol excretion was within our normal range.

Blood was drawn from right and left antecubital veins; the euglobin lysis time was estimated and found not to differ significantly (right arm 345 min; left arm 360 min). However, skin biopsies taken from corresponding sites in right and left arms were assayed for tissue fibrinolytic activity (2) and this was found to be increased in tissue from the right arm (47% vs. 24%).

Because of the increased skin fibrinolytic activity observed in the right arm, we prescribed oral epsilon aminocapnoic acid and the patient has had no further bleeding in the past 5 months.

DISCUSSION

Cases of unilateral hypertrophy and hemangiomatous malformations were described in the mid-nineteenth century (1), but Klippel & Trenaunay in 1900 (6, 7) grouped together the triad of signs now called eponymously after them and F. Parkes Weber.

The condition has been associated with paraplegia (5) and even malignant change (3). It appears to be more common in boys, with the arm affected twice as frequently as the leg; the head and trunk are affected in 10% of cases (4, 10).

We were surprised to find no difference in venous fibrinolytic activity between right and left arms. As venous endothelium produces fibrinolytic activators, we might expect increased fibrinolytic activity in the affected arm with more numerous venous channels. However, if the veins per unit area were to produce fewer activators than normal, the amount of fibrinolytic activity seen in the abnormal limb would be similar to that in the normal limb, thus explaining our finding and giving support to the concept that the veins in the abnormal limb are functionally as well as anatomically abnormal.

Bleeding from telangiectatic vessels is not usually a problem with these patients. The mechanism of action of the bleeding could be a failure of contraction in an abnormal part of the vessel wall.

Fibrinolytic activators are produced by vascular—particularly venous—endothelium and the