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MALIGNANT ENDOVASCULAR PAPILLARY HEMANGIOENDOTHELIOMA OF THE SKIN

The Nosological Situation

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Abstract. Description of a case of malignant endovascular papillary hemangioendothelioma of the skin, in accordance with present-day criteria for its individualization. The problems involved in its nosological description are analysed, and the necessity to proceed to a complete surgical excision and possibly even to a regional lymph node dissection, is argued.

"Among the prerequisites for comparative studies of cancer are international agreement on histological criteria for the classification of cancer types and a standardized nomenclature." With these words, the WHO begins the preface to Volume 3 of its recent International Histological Classification of the Tumours; a little later in the same volume, Enzinger (6) adds: "Although knowledge of soft tissue tumours has increased greatly in recent years, there is still much confusion concerning their incidence and behaviour and the best mode of therapy".

Both statements are worthy of meticulous consideration, above all when one is concerned with malignant vascular tumours rarely met with, involving a terminology of the most varied kind and considerable difficulties in histological interpretation and classification.

Among the most recent classifications, are Wilson Jones' (13) (Table I); and Mascaro's (10) (Table II).

If it is the case, as we have indicated, that malignant vascular tumours are out of the ordinary, much more so is angioendothelioma, considered in isolation (3, 8). The following data are in this sense very significant: at the Bellevue Hospital in New York, between 1925 and 1946, there were only 8 cases (5); at the Mayo Clinic, among cancers of the pelvis operated on between 1935 and 1954, 2 hemangioendotheliomas are recorded (7); and, up to 1965, 155 observations were made at the Laboratory of Surgical Pathology of Columbia University, of which the greater part were visceral or osseous in their localization (12).

It is necessary to draw attention to the great variety of descriptive nomenclature used in reference to malignant tumours, localised or regional, of the vascular endothelium: viz. angioendothelioma, angiofibrosarcoma, angiosarcoma, endotheliosarcoma, hemangioendotheliosarcoma, hemangioblastoma, hemangioendothelioblastoma, malignant endothelioma, angioblastoma, and malignant hemangioendothelioma, the latter denomination being the one accepted by the Committee on Tumour Nomenclature of the International Union against Cancer.

In addition, there are certain histological types in connexion with which the question arises as to whether or not they are in fact malignant tumours, and even whether or not they are really genuine vascular tumours. This is the case with regard to endovascular papillary (vegetating) hemangioendothelioma, a case of which we had occasion to study.

CASE REPORT

A 54-year-old white man, born in the province of Granada (Spain), was examined on 23 April 1971. He had a hard infiltration on the upper edge of the left exterior ear, some pain, a central ulcer with clearly-delineated borders,

Table I. *Malignant vascular tumours*[a]

1. *Malignant angioendothelioma*
1.1. Idiopathic
1.2. Secondary to ...
1.2.1. Lymphangiectasis in chronic lymphoedemas (the commonest example being the Stewart and Treves tumour)
1.2.2. Radiotherapy
1.2.3. Vascular naevi
2. *Angiosarcoma*
3. *Kaposi's sarcoma*
4. *Malignant hemangiopericytoma*

[a] According to Wilson Jones, E. (13).

violet-coloured erythema, bleeding easily, and necrotic base (Fig. 1). The process had started in December 1970, with edema and bluish-red colouring, ulcerating after 2 months. There were no palpable lymph nodes. Clinico-analytic examination and radiology were normal.

A "punch" biopsy, diameter 5 mm, of the borders of the ulcer was carried out. It was fixed in formalin, embedded in paraffin and sectioned at 5 μm. Staining: Hematoxylin-eosin, and Bielschowsky-Maresch method.

Histopathological examination provided the following information: Atrophy of the epidermis, with almost rectilinear dermo-epidermal juncture. At some points, hyperkeratosis, predominantly follicular; at others, exulceration and ulceration partly covered by serchaematic crust.

The whole of the dermis, with evident thickening, was occupied by very swollen, intercommunicating and poorly defined capillaries, the endothelium of which was atypical, varying in size and shape, and in many places projecting inwards in interwoven papilliform digitations with the appearance of giant renal glomeruli (Figs. 2, 3). In the lumen of the vascular channels there were many red and white blood cells (Fig. 4). The endothelial cells, within varying grades of differentiation, were pleomorphic, and showed anisocytosis and nucleocytoplasmic anomalies. In some, the protoplasm was scarce and basophilic, and in others, clearly acidophilic or in plain vacuolization. Nuclear hyperchromatism was constant. Infrequent mitosis, but atypical. Cell aggregates floated free in the lumina of vessels. Some cells formed syncytiae. There were also cellular groups reminiscent of adipose tissue, having excentric nuclei, which lay between the capillaries in the stroma; some were anaplastic and surrounded by reticulum fibres, like the vascular channels, as shown by the specific stainings. Fibrosis and edema of the strema round the tumoral and capillary masses were evident.

Table II. *Malignant vascular tumours*[a]

1. *Perithelial proliferation*
1.1. Fibroblastic:
1.1.1. Systematized (Kaposi's disease)
1.1.2. Localised or regional (Angiopoietic Fibrosarcoma)
1.2. *Pericytary:* Malignant hemangiopericytoma
2. *Endothelial proliferation*
2.1. Systematized. Systematized proliferating Angioendotheliomatosis
2.2. Localised or regional. Malignant Endotheliomas

[a] According to Mascaro Ballester, J. M. (10).

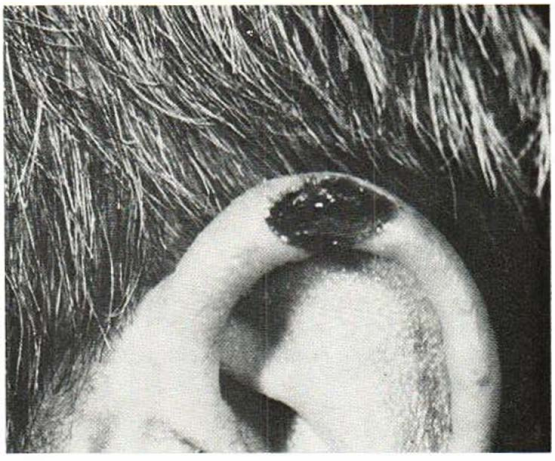

Fig. 1. Hard infiltration on the upper edge of the helix, left exterior ear, with central ulcer, clearly outlined borders which bleed easily, and necrotic base.

Diagnosis: Malignant endovascular papillary hemangioendothelioma.

We therefore performed a complete surgical extirpation of the tumour. On gross examination, the piece removed, when cut in half, showed an ulcerated zone below which was a brittle, spongy, greyish tissue with some haemorrhaging spots.

It was impossible to determine the limits of the upper edge of the ear cartilage. Histological examination confirmed the data given above, revealing and corroborating the partial destruction of the underlying cartilage. The frozen sections which were stained with Sudan IV showed an absence of fat in cells similar to that in adipose tissue. One year after the operation, the clinical cure continues, without adenopathy. The patient remains under observation.

DISCUSSION

Masson (11) described an endothelial, papilliferous proliferation in a hemorrhoidal mass, calling it "hémangioendothéliome végétante intravasculaire"; and later Henschen (9) demonstrated histologically similar lesions at other sites (ureteral caruncle, nasal polyps, larynges, in the digestive tract, the uterus . . .).

More recently, Civatte & Harter (2) have shown a case localised in the facial skin, and Dabska (4) of the Institute of Oncology in Warsaw has studied in detail 6 cases in children observed over a period of 14 years (1953–1967) (Table III).

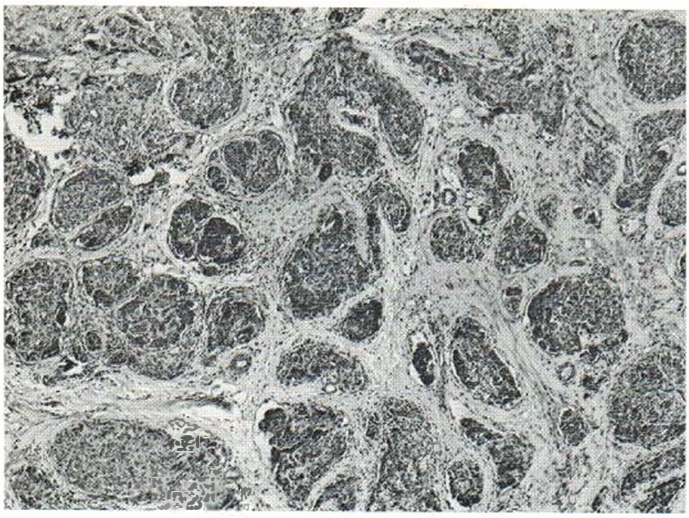

Fig. 2. The whole of the dermis is occupied by swollen, intercommunicating and poorly defined capillaries, the endothelium of which projects inwards, forming interwoven papilliform digitations (H.E., × 35).

According to her observations, she distinguishes two clinical varieties, the one where there exists a diffuse swelling of the skin, and the other where there is a real intradermal tumoration, implying a local invasive capacity and possible metastatic lymph nodes. Histologically, the manifestation is similar to a marked endovascular papilliferous proliferation of the atypical endothelium, which, when projected in the vascular lumen, gives to the affected vessels the appearance of giant renal glomeruli. Taking into account the relative malignancy of the tumour, Dabska proposes the name "malignant endovascular papillary angioendothelioma" and advises complete surgical excision, followed by dissection of the regional lymph nodes. Radiotherapy is not effective.

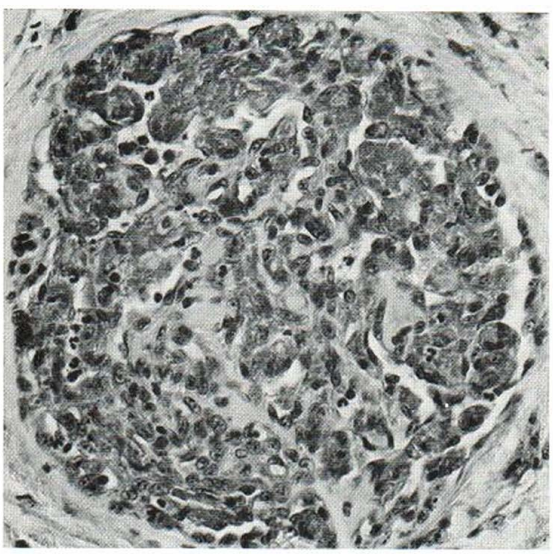

Fig. 3. Interwoven papilliform digitations with the appearance of "giant renal glomeruli" (H.E., × 90).

The clinical-histological situation described evidences all the characteristics necessary for a precise diagnosis (4, 11). We draw attention to the topography (the exterior ear) and the destruction of cartilage (which we have not found in any published description), the age of the patient (frequent nevertheless in "classic" malignant hemangioendothelioma), and the clinical appearance of the tumour with central ulceration (only one of Dabska's cases, number 2, was ulcerated, and it was localised to the heel).

Two fundamental problems arise:

1. Is it a vascular tumour, or is it a question of a histological manifestation, caused by the thrombosis and partial recanalization of a vessel? The latter is hardly probable since in no case are we dealing with a single capillary (1), but always with many—all the dermal capillaries are affected. Furthermore, in cases of cutaneous affections in which thrombosis and partial recanalization are common, as happens for example with late chronic radiodermatitis, we have not found a histological example of the kind we are now dealing with, and there are no bibliographical references in support.

2. Are we concerned with a benign vascular tumour, or an authentic malignant one? In agreement with Dabska (4), the existence of mitosis

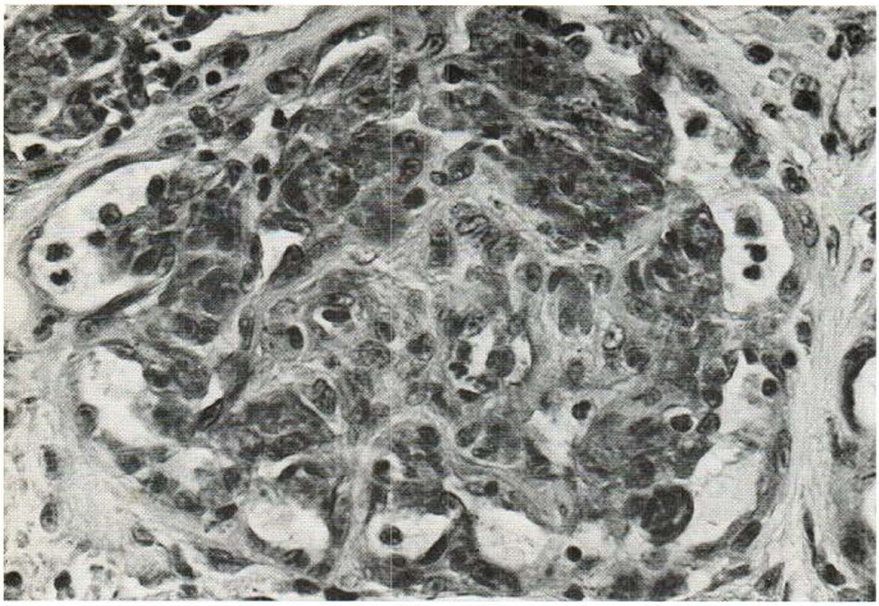

Fig. 4. The lumen of the vascular channels is filled by many red and white blood cells. Pleomorphic endothelium (H.E., ×250).

and nucleocytoplasmic anomalies, as well as of identical situations in the regional lymph nodes which are interpreted as metastatic, lead one to judge the case as one of a tumour, at the very least potentially malignant, and in consequence radical treatment (surgical excision) is unavoidable. Furthermore, it would be difficult to find an explanation for the destruction of cartilage or bone (4) if the tumour were benign.

To sum up, we believe that the endovascular papillary hemangioendothelioma corresponds to a neoplastic proliferation of the endothelium and must be considered as a variant of "malignant hemangioendothelioma". Its clinical analysis is not very precise and is variable; its histology, on the other hand, allows of its identification. Taking into account its evolutionary characteristics and the possibility of metastasis, radical and de-

Table III. *Malignant endovascular papillary hemangioendothelioma*[a]

Marie Sklodowska Curie Institute of Oncology, Warsaw

Sex	Location	Diam. size (cm)	Metastases	Treatment	Follow-up (years)
♀	Knee	9	No	1949: X-ray 1953: excision	14
♂	Heel	4	No	1956: excision	11
♀	Cheek	4	No	1951: X-ray 1956: electroexcision 1957: electrodesiccation	16
♀	Temple	8	No	1957: excision, then X-ray	10
♀	Neck	5	Reg. lymph node	1961: excision	6
♂	Palm	5	Axillary lymph nodes	1963: Amputation of 3 fingers and metacarpals	4

[a] Modified from Dabska, M. (4).

finitive treatment is inescapable, and is achieved by the necessary surgical excision since it is radio-resistant.

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