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Cutaneous Mesenchymal Tumour with Haemangiopericytoma-like Features

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A case of cutaneous mesenchymal haemangiopericytoma-like tumour affecting a 17-year-old male is presented. Clinical features were non-specific. The tumour was studied by light, electron microscopy and immunohistochemistry. The results of these studies suggested the diagnosis of a mesenchymal, poorly-differentiated tumour showing features of haemangiopericytoma. The clinical course after surgery was uneventful after a 9-month follow-up period. ***Key-words: Histology; Immunohistochemistry.***

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Haemangiopericytoma is a rare mesenchymal tumour, first described by Stout & Murray in 1942 (1), which originates from the pericytes of Zimmermann, i.e. modified smooth muscular cells surrounding capillaries and post-capillary venules. This tumour is most frequently found deep-seated in the muscles of the lower limbs and trunk (2). We report the case of a cutaneous mesenchymal tumour with haemangiopericytoma-like features located on the upper arm of a young man; the difficulty in establishing a precise diagnosis is discussed.

CASE REPORT

A 17-year-old student, with an unnoticeable past medical history, consulted us for a firm, elastic, telangiectatic reddish nodule of the upper left arm, bordering the shoulder (Fig. 1). It had appeared 1 year earlier and slowly enlarged to reach a diameter of 15 mm. This asymptomatic cutaneous nodule was attached to the skin but not to the underlying muscle. The patient was otherwise in good health.

A biopsy was taken under local anaesthesia. Histological examination of formalin-fixed, paraffin-embedded tissue revealed a tumour consisting of tightly packed proliferating cells arranged in nodules or cords invading the entire dermis. The tumour cells were spindle-shaped and contained eosinophilic cytoplasm and large, round nuclei (Fig. 2). Within the tumour masses, numerous thin endothelial-lined vascular channels were present. Mitotic figures among tumour cells were numerous (5 per 10 high-power field). No keratinization, pigment, arrangement in nests, haemorrhage or necrosis were present. Histologically, the diagnosis of a mesenchymal, spindle-cell tumour was made, with no further characterization of the precise origin of proliferating cells. Histochemical staining for reticulin revealed a dense meshwork surrounding the vessels and tumour cells. Since the diagnosis remained somehow equivocal, a second biopsy for an ultrastructural and immunohistochemical study was performed.

Electron microscopy revealed oval or polyhedric tumour cells measuring 10 to 30 μm in diameter. Cell nuclei were roundish and contained euchromatin with a thin peripheral rim of heterochromatin (Fig. 3). The cytoplasm was abundant and contained rough endoplasmic reticulum, numerous mitochondria and abundant round myelinoid or multilamellar inclusions (Fig. 4). Tumour cells exhibited interdigitating cytoplasmic processes but no specialized intercellular junctions. Neither melanosomes nor Weibel-Palade bodies or well-formed basal laminae were seen.

Immunohistochemical studies were performed on both frozen and paraffin-embedded sections, with the biotin-streptavidin-peroxidase technique and a large panel of monoclonal and polyclonal antibodies. Tumour cells proved negative for keratin, desmin, neurofilaments, S-100 protein, factor VIII-related antigen (von Willebrand factor), UEA-1 lectin, factor XIIIa, leucocyte common antigen (CD45), CD1a, CD3, CD4, CD8, CD14, CD22, CD30, CD36, CD45RO, HLA-DR, carcinoembryonic antigen, epithelial-membrane antigen, HMB-45, muscle-specific actin (HHF35), neuron-specific enolase and L1 antigen (Mac387). Tumour cells were found positive only for vimentin, strongly suggesting their mesenchymal origin. This immunohistochemical profile excluded the diagnosis of spindle-cell carcinoma, melanoma, lymphoma, endothelioma, myosarcoma, schwannoma, synovial sarcoma, chondrosarcoma and malignant fibrous histiocytoma. Antibodies to basal membrane macromolecules (laminin and type IV-collagen) showed a weak reactivity around tumour cells. Immunostaining of endothelial cells (factor VIII-related antigen, CD36

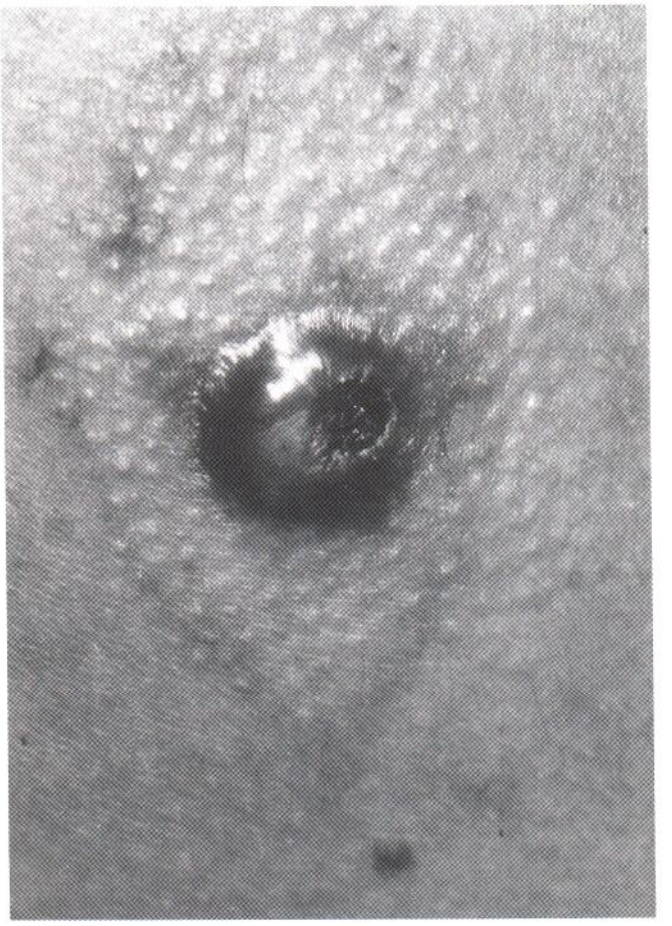

Fig. 1. Cutaneous nodule with a telangiectatic reddish surface. A scar due to the biopsy is seen on the right side.

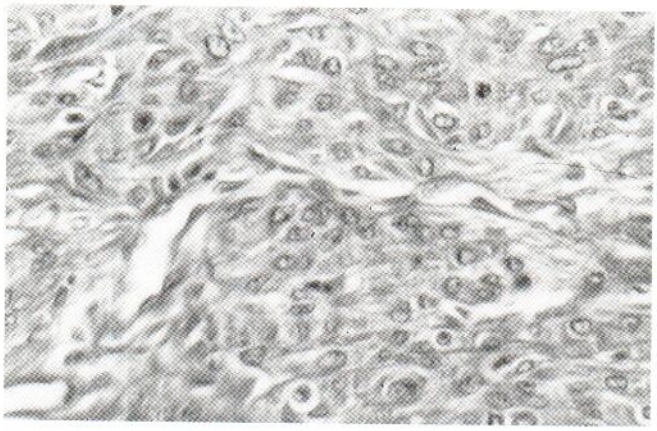

Fig. 2. Low-power histologic aspect of the tumour reveals thin endothelial-lined vascular channels surrounded by monomorphous, large, globular or spindle-shaped cells, occasionally in mitosis.

and UEA-1 lectin) revealed a well-developed vacular network inside the tumour masses (Fig. 5). The presence of vimentin-positive spindle-shaped cells surrounding a prominent vascular network (better decorated thanks to the immunohistochemical staining), excluding all the possible diagnoses mentioned above, suggested the diagnosis of haemangiopericytoma.

A general examination (clinical, chest x-ray, thoraco-abdominal tomodensitometry and bone scintigraphy) did not reveal visceral involvement. Soft tissue echography showed that the tumour was well-defined. Treatment consisted of a large carcinologic local excision, allowing a 3-cm safety margin, deep to the fascia, utilizing total skin grafting for reconstruction. Nine months after treatment, no evidence of relapse or metastatic spread existed.

DISCUSSION

Haemangiopericytomas are usually deep soft tissue and muscle tumours. Rarely, as in our case, the tumour consists of a superficial cutaneous nodule not adhering to the fascia. This particular feature may be related to the age of our patient, since it is more frequently seen early in life (3, 4). The diagnosis is usually difficult to establish clinically, even though the tumour exhibits a firm consistence, purple colour and is covered by telangiectasias. Due to this non-specific presentation, it may be confused with a keloid or a scar, a fibrous histiocytoma or a vascular tumour.

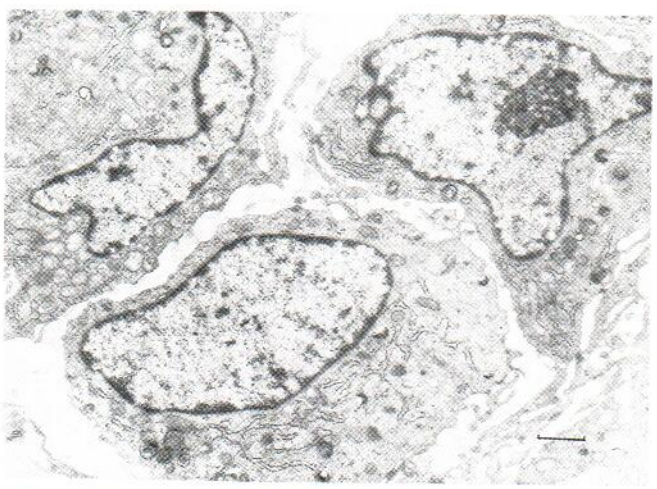

Fig. 3. Three polyhedric tumour cells with round nuclei are seen (electron microscopy – scale bar: 1 μm).

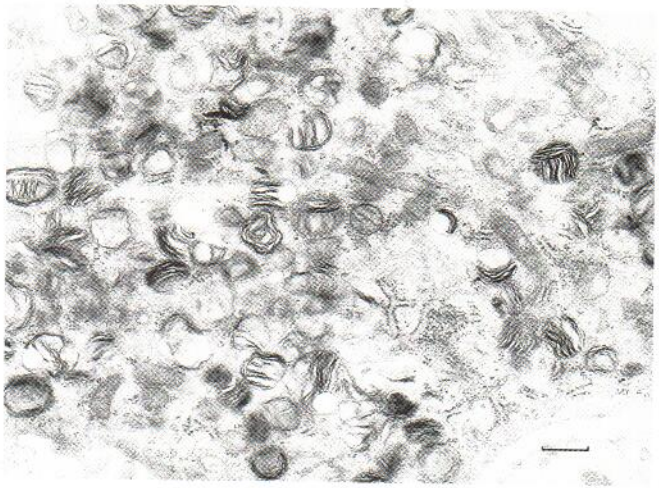

Fig. 4. Cytoplasmic myelinoid or multilamellar cytoplasmic inclusions within tumour cells (electron microscopy – scale bar: 500 nm).

Fibrous histiocytoma, synovial sarcoma, mesenchymal chondrosarcoma and myxoid liposarcoma can mimic haemangiopericytoma (1, 5). Thorough microscopic examination, together with electron microscopic and immunohistochemical studies (2), is usually necessary in order to establish the diagnosis. Our case emphasizes the difficulty in establishing the diagnosis histologically. Pericytes are not readily identifiable by light microscopy since they lack distinctive cytologic features. The perivascular arrangement of tumour cells is not always detectable, since the proportion of tumour cells and vascular channels varies (4, 6). Our case presented some unusual electron microscopic features. Indeed, a remarkable ultrastructural finding was the presence of cytoplasmic multilamellar and myelinoid inclusions, presumably of lysosomal origin. These looked similar to those found within various types of mesenchymal cells (endothelial, fibroblastic, histiocytic, pericytic) in the course of local or generalized metabolic diseases. Therefore, the presence of these inclusions highlighted the mesenchymal origin of the proliferating cells; these pre-

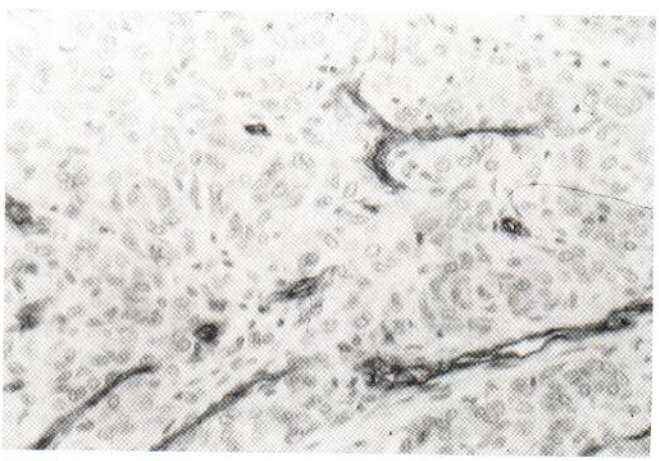

Fig. 5. Immunostaining of endothelial cells by UEA-1 lectin reveals a well-developed vascular network inside the tumour mass.

sented cytoplasmic projections but no distinctive organelles (such as melanosomes or Weibel-Palade bodies) or a well-defined basal lamina or pinocytotic vesicles (3, 7). The immunohistochemical profile of the tumour excluded the diagnosis of lymphoma, melanoma, carcinoma, smooth muscle tumour, endothelial, neural or neuroendocrine tumour (8). The results of the light microscopic, immunohistochemical and electron microscopic studies taken in concert were suggestive of a mesenchymal tumour with features reminiscent of haemangiopericytoma.

The prognosis of haemangiopericytomas is difficult to establish. These tumours may be benign, malignant or borderline. Large-size tumour, dense cellularity, increased mitotic counts (over 4 mitoses/10 high-power fields) and the presence of haemorrhage and necrosis are considered as criteria of malignancy (4). There are no radiographic, tomodensitometric, angiographic or MRI prognostic criteria (9). A recent study suggested that the pattern of reactivity of antibodies to PCNA/cyclin was of value in establishing the predictive prognosis of haemangiopericytomas (10). In our patient the small size of the tumour, the lack of focal haemorrhage and necrosis were favourable prognostic features, as opposed to the increased mitotic rate and the high degree of cellularity. Haemangiopericytomas may metastasize 5 years or more following initial diagnosis, and thus require a long follow-up. Lungs, skeleton and liver are common metastatic sites (4, 11, 12).

The management of this tumour is difficult. Local wide carcinologic excision seems to be the primary treatment. Surgery is often performed alone, since haemangiopericytomas do not respond well to x-ray therapy and palliative x-ray therapy of the surgical site does not improve prognosis (4). Polychemotherapy may be tried in cases with metastatic spread. Adriamycin seems the most effective agent (13, 14). X-ray therapy may be helpful in inextirpable recurrences or metastases, but its efficacy is doubtful (2, 14).

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