

Malignant Change in Composite Haemangioendothelioma Over 13 years

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Abstract:

For descriptive purpose the term "composite hemangioendothelioma (HE)" is used to include the features of more than one subtype of HE. Composite HE rarely undergoes malignant transformation and does not usually metastasise.

A 57 year old man who first presented with a soft swelling on the dorsum of his right hand 13 years ago. It was excised and recurred on multiple occasions regionally on the right hand and forearm. The histological features consistently remained as a composite hemangioendothelioma for 10 years. Thirteen years later it recurred again on forearm and arm which is managed with wide excision and free rectus abdominus muscle flap reconstruction. However, histology showed a features suggestive of frank angiosarcoma.

Key words: Haemangioendothelioma; Malignant vascular tumour; Angiosarcoma; Recurrence.

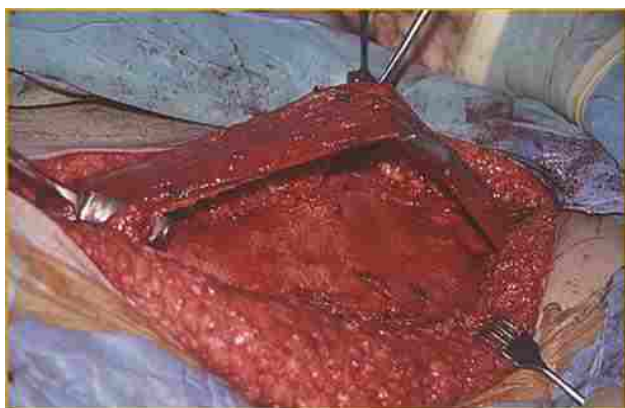


Fig. 1
Angiosarcoma arising in a previously diagnosed composite haemangioendothelioma



Fig. 2
Wide excision of tumour and reconstruction with free rectus abdominus muscle and split thickness skin graft



Fig. 3
Healed flap and skin grafts

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The term "hemangioendothelioma" has been applied in recent years to heterogeneous group of vascular neoplasms, intermediate in both behaviour and histologic appearance between benign tumours (hemangioma) and frankly malignant tumours (angiosarcoma).

The spectrum of HE encompasses spindle cell HE (2), kaposiform HE (2), retiform HE (3), epitheloid HE, and polymorphous HE of the lymph nodes. It is a small group of vascular neoplasms that may show considerable overlap in histological appearances.

We report our experience with a patient characterised as epitheloid HE, retiform HE, and with angiosarcoma-like areas on first excision. Later the lesions recurred locally and was excised with similar histology on multiple occasions. A regional recurrence after 13 years on the forearm and arm showed features of high grade angiosarcoma.

Case Report

A 57 years old man presented with a 3 cm area of ill defined "swelling" on the dorsum of his right hand in 1992 of 3 years duration. The lesion was excised and reconstructed with split thickness graft. Microscopic examination of the specimen showed a poorly circumscribed mass composed of an infiltrate of cells. The tumour was "Composite haemangioendothelioma", being composed of areas with strikingly epitheloid cytology, as well as more spindle cell areas and foci with a retiform appearance. The cells showed a moderate pleomorphism and individual vacuolization. Mitotic figures were scanty. The cells laid in a predominantly hyaline matrix. Immunohistochemistry showed positive staining for factor VIII and vimentin. Staining for Cam 5.2, S100 and desmin were negative. The tumour had a multinodular appearance and showed large foci of tumour in the specimen and extending to all excision margins.

There were multiple local and regional

recurrence over the subsequent 10 years on the right hand, forearm and arm. These were managed by excision and reconstruction either with split thickness skin graft or local fasciocutaneous flap. During this time he was treated by local radiotherapy 50 Gy divided into 25 sessions, to his hand, forearm and arm in 1994 and a trial of chemotherapy in 1997 including 5FU/leucovorin and DTIC with no effect. In 2000 also received thalidomide and interferon

again of no benefit. On number of occasions, he was treated with local radiotherapy approximately 15 Gy to control the pain and bleeding and had two short courses of photodynamic therapy in 2001 and 2002.

Thirteen years after the initial presentation a large area of multiple exophytic and ulcerative disease persisted over the right forearm and lower arm (Fig1) and this necessitated a wide excision of the tumour and reconstruction with a free rectus abdominus muscle flap and split thickness graft in 2002 (Fig 2). The histology revealed a malignant vascular tumour with multiple mitoses and atypical nuclear morphology. This was an extensive and multifocal vascular tumour which focally involves radial and deep margins.

Subsequently, he developed regional recurrence of the tumour in the right upper limb with metastasis in the axillary lymph nodes. Biopsy of specimen from the right arm in Sep, 2003 showed a malignant tumour expanding the dermis and subcutis with predominantly epitheloid morphology with prominent vascular channels. The specimens of lymph nodes from right axilla are extensively replaced by metastatic vascular tumour, showing epitheloid and spindle cell morphology, with extranodal extension. There is no evidence of disease progression since 16 months later, confirmed by staging MRI and CT scans.

Discussion:

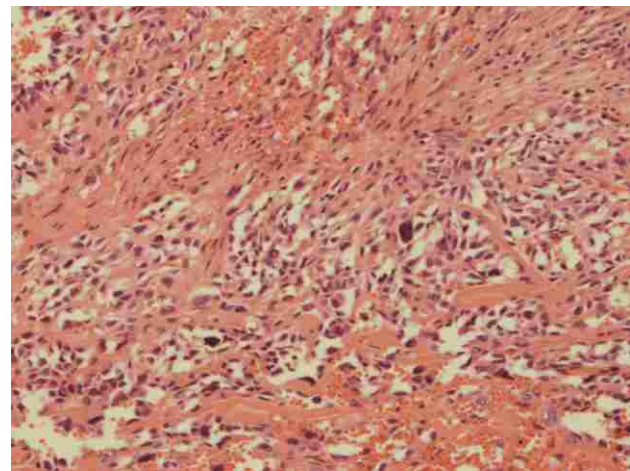
The term "haemangioendothelioma" (HE) has been applied in recent years to heterogenous group of vascular neoplasms, intermediate in both behaviour and histological appearance between benign tumours (haemangioma) and frankly malignant tumours (angiosarcoma).

Haemangioendothelioma is a well differentiated vascular tumour, usually locally aggressive and angiosarcoma is a anaplastic and highly metastasizing counterpart. Therefore, it can be regarded as a rare low grade tumour of vascular origin. All age groups can be afflicted and there is a predilection for men. In order to attempt the prediction of outcome from observance, molecular and biochemical means, haemangioendothelioma may vary in appearance from a small pedunculated nodule to a large invasive mass. They are either skin-coloured, or reddish purple soft nodules, or dusky red infiltrated plaques which have tender raised margins and may be surrounded by a slowly spreading erythema. The lesions may be covered by scales

or haemorrhagic crusts and may ulcerate (4). Angiosarcoma is high grade sarcoma, with an aggressive metastasizing potential. Radiotherapy may induce angiosarcoma in a previously benign lesion. The interval between radiotherapy and malignant change is usually more than ten years which is not in our case. The lymph node metastasis is the dominant pattern in case of angiosarcoma and rarely hematogenous. Pathologically it presents as a haemorrhagic tumour, with histological evidence of neoplastic vascular tissue. The lesion may be multicentric in origin. The most striking feature of composite haemangioendothelioma at low power is the variability in appearance from patient to patient and from area to area. This is due to the relative proportions and arrangement of individual histological components within the tumour.

It is not uncommon for expert sarcoma pathologists to disagree about specific histologic diagnosis. Few pathologists have the opportunity to study many of these rare tumours during their careers, and this lack of experience may contribute to the relatively low concordance rate(5). In our case all specimens were reviewed by the same expert pathologist Christopher D.M Fletcher, over the period of 13 years (5,7).

However, we control the disease by local resections and reconstruction, radiotherapy and chemotherapy. To our knowledge till 2006, he is alive with recurrent composite HE with regional recurrence and conversion into angiosarcoma in the right arm and metastasis in the right axillary lymph nodes. His right upper limb is functional to his daily activities.

**Fig. 4.**

Angiosarcomatoid area shows vascular channel with dissecting fibroconnective tissue and red blood cell extravasation is seen. The vascular channels show a marked degree of cytologic atypia.

Conclusion:

For all Composite HE, the close surveillance for the possible development of recurrence and transformation into malignant disease is recommended. In the long term, the patient can be managed with treatment modalities available without scarifying important structures of the body

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