Abstract  Angiosarcomas are rare malignant mesenchymal tumours, characterized morphologically by anastomosing vascular channels lined by atypical and proliferative active endothelial cells. An epithelioid cytome- 

phology of tumour cells is often seen focally in angiosarcoma, whereas purely epithelioid angiosarcomas are rare. Although angiosarcomas show a vascular differentiation they are almost never confined to pre-existing blood vessels. We describe three cases of intravascular epithelioid angiosarcoma arising in the carotid artery of a 60-year-old man, in the infrarenal part of the abdomi- 

nal aorta and both renal arteries of a 69-year-old woman, and in the abdominal aorta of a 68-year-old man. In all cases malignant tumour tissue was found incidentally after disobliteration of thrombosed vessels. Histologically, purely epithelioid angiosarcoma composed of solid sheets of epithelioid tumour cells was seen; immunohistochem- 

istry confirmed the endothelial differentiation of neo-

plastic cells. The reported cases show that angiosarcoma can occasionally arise within a pre-existing vessel.

Key words  Angiosarcoma · Epithelioid angiosarcoma · Intravascular angiosarcoma · “Intimal” sarcoma · Soft tissue tumours

Introduction

Angiosarcoma is a rare and clinically aggressive mesenchymal neoplasm occurring in different clinical settings. Cutaneous angiosarcomas arise mainly in the head and neck region of elderly patients, in patients with chronic lymphoedema of the limbs (so-called lymphoedematous angiosarcoma), or after radiotherapy. Furthermore, angiosarcomas occur in the skeletal system, in the breast, in parenchymatous organs (liver, spleen, ovary, thyroid gland), and in soft tissues [3]. Histologically, angiosarco-

ma is characterized by a diffuse rather than lobular or nodular growth pattern, as seen in benign vascular lesions. At higher power, the anastomosing neoplastic vessels are lined with atypical and proliferative active tu-

mour cells with endothelial differentiation. In a recent extensive study of angiosarcomas of soft tissue, the morphological spectrum has been delineated and the striking variations of morphological features within a given neo-

plasm emphasised [7]. Frequently, a focal epithelioid cytomorphology characterized by abundant eosinophilic cytoplasm and vesicular nuclei with prominent nucleoli is seen in angiosarcomas of soft tissue [7]. However, purely epithelioid angiosarcomas are rather rare and easily mistaken for metastatic carcinoma [4].

Independently of clinical and morphological features, angiosarcomas are only very rarely confined to a pre-ex-

isting blood vessel, either because they do not arise from the endothelium of major vessels or because they destroy and invade so rapidly that angiocentricity cannot be verified [12].

Recently, we have seen three cases of intravascular epithelioid angiosarcoma.

Materials and methods

The cases were identified in the consultation files of one of the authors (D.K.). The specimens were fixed in 10% buffered formalin, conventionally processed and embedded in paraffin wax. Sections 4 µm thick were stained with haematoxylin and eosin, periodic ac-

id-Schiff reaction (PAS), and Goldner trichrome staining. Immunohistochemical studies were performed on paraffin sections with the alkaline phosphatase-anti-alkaline phosphatase method (AP-

AAP) using appropriate positive and negative controls throughout. The antibodies used are listed in Table 1 with their dilutions and sources. Mitotic rate was expressed as the average mitotic count seen in 10 high-power fields (HPFs: 1 HPF=0.159 mm² for the microscope used); 30 HPFs were counted in each case. Clinical
Details and follow-up information were obtained from the hospital records, the laboratory request forms, and the referring pathologists if possible (see Acknowledgements).

Results

Case 1

A 61-year-old man had a 5-week history of initially transient, later progressive and permanent neurological deficit with right-sided hemiparesis and dysarthria. Magnetic resonance imaging showed an infarction in the parietal lobe. Duplex sonography of the left internal carotid artery revealed a poorly echogenic area, which was considered to be thrombotic material. Two weeks later a follow-up sonography demonstrated a dramatic increase of the “thrombus” formation described before. An operation was considered, and a preoperative angiography was performed (Fig. 1). At exploration after arteriotomy a very soft, red-brown material was found incompletely filling the lumen of the common and internal carotid artery below a stenosis at the probable upper end of a former endarterectomy (17 months ago). The internal carotid artery above and the common carotid artery below the bifurcation were resected over a length of 8 cm.

Histological examinations showed blood vessel structures with prominent fibrosis in the intima and media, foci of intramural haemorrhage and scattered inflammatory cells. Extensive atherosclerotic plaques with focal calcification and cholesterol clefts were seen. In addition, malignant tumour tissue, which was limited mainly to the intima and showed infiltration of the media only focally, was identified. The neoplastic tissue was composed of large polygonal cells with abundant eosinophilic cytoplasm and vesicular nuclei with prominent nucleoli. These pleomorphic tumour cells were arranged in solid sheets or in a diffuse pattern with small holes and slits containing occasional erythrocytes (Fig. 2). The mitotic rate was increased to up to 5–6 mitoses/10 HPF, including atypical mitoses; foci of coagulative tumour necrosis were noted.

Immunohistochemical stains demonstrated vimentin positivity and a clear membranous positivity of tumour cells for CD31 (Fig. 3). Focally, tumour cells stained weakly positive for pancytokeratin. Stains with antibodies against CD34, factor VIII-related antigen, muscle-specific actin, and α-smooth-muscle actin were negative.

The patient was checked carefully, but no other primary neoplasm was found by extensive clinical and radiological investigation. Up to 7 months after tumour excision no signs of recurrence or systemic metastases were noted.

Case 2

A 69-year-old female patient presented with high-grade stenosis of the infrarenal aorta and both renal arteries. Disobliteration was performed and histological investigation showed high-grade atherosclerosis with attached thrombotic material. Mainly on the surface of the thrombotic material and focally attached to the intima poorly differentiated malignant tumour tissue was identified. The polygonal epithelioid tumour cells showed a solid arrangement and contained enlarged round to oval vesic-