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Case report

Intravascular glomus tumour: a previously undescribed phenomenon

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Summary. We report the first case of an intravascular glomus tumour, which was located in the right forearm of a 40-year-old male. Microscopically the lesion originated from the wall of a vein and protruded into the lumen of the affected blood vessel. The tumour cells were characterized immunohistochemically by the presence of vimentin, actin and myosin. Within the tumour, small nerves, immunopositive for S-100 protein and neurofilaments, could be identified. Histogenetically, the tumour is thought to derive from intramural epithelioid cells of the venous part of an arteriovenous anastomosis.

Key words: Intravascular glomus tumour – Blood vessel – Immunohistochemistry

Introduction

Although large series of glomus tumours have been reported in the literature (Mullis et al. 1972; Anagnostou et al. 1973; Tsuneyoshi and Enjoji 1982; Enzinger and Weiss 1988), an intravascular lesion appears never to have been described. Therefore, we present the case of a 40-year-old male patient with an intravascular glomus tumour of the right forearm.

Materials and methods

The case was collected retrospectively from the files of the Department of Histopathology, St. Thomas's Hospital, London and picked out of a total of 80 glomus tumours. The lesion was excised from the right forearm of a 40-year-old African male who had presented clinically with a painless "cyst".

After surgical removal, tissues were fixed in 10% unbuffered formaldehyde solution, embedded in paraffin wax and processed conventionally. Consecutive sections (4 μm) were stained with haematoxylin and eosin.

For immunohistochemical demonstration of the respective antigens the APAAP method (Cordell et al. 1984) was performed using the following primary antibodies after protease digestion: (i) anti-desmin (monoclonal, dilution 1:50, DAKOPATTS, Glostrup, Denmark); (ii) anti-panneurofilament protein (monoclonal, 1:100, Dakopatts); (iii) anti-vimentin (polyclonal, 1:10, EPIGNOST, Linz, Austria); (iv) anti-myosin (polyclonal, 1:100, BIOGENEX, San Ramon, CA, USA); (v) anti-myoglobin (polyclonal, 1:500, Dakopatts); (vi) anti-human muscle actin (HHF 35, monoclonal, 1:20, ORTHO, Neckargemünd, FRG); (vii) anti-alpha smooth muscle actin (monoclonal, 1:2000, SIGMA, Deisenhofen, FRG); (viii) anti-F VIII-RAG (monoclonal, 1:50, Dakopatts); (ix) anti-S-100 protein (polyclonal, 1:500, Dakopatts).

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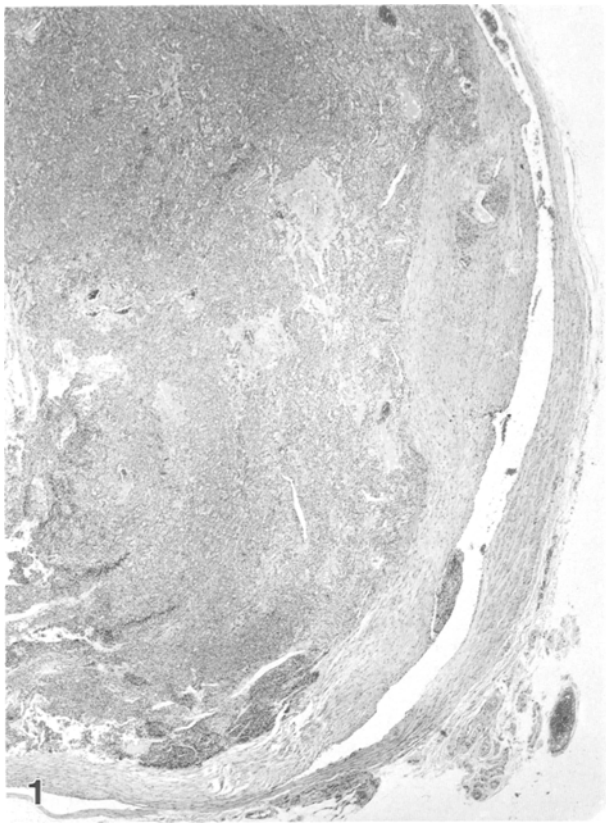
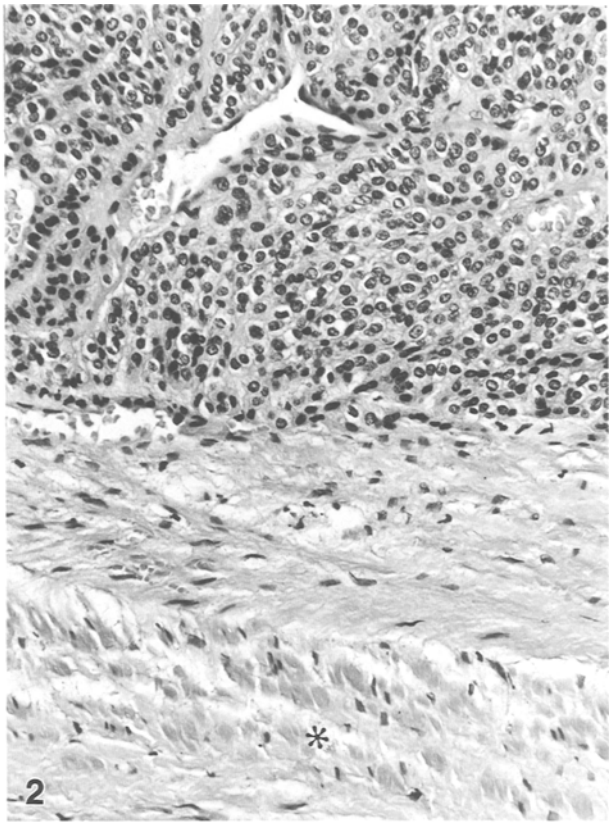
Results

Macroscopically the lesion corresponded to a nodule of 0.7 cm in diameter. The cut surface was brownish red.

Histologically the tumour obviously originated from the wall of a vein by a broad stalk and protruded into the lumen of the affected blood vessel. The lumen was nearly completely obstructed and only occasional blood filled; endothelial lined slit-like spaces could be seen (Fig. 1). Examination of the perivascular tissue revealed no tumour cells. Peripherally the tumour was covered by the intact smooth muscle layer of the affected blood vessel and centrally by vascular endothelium which was pushed into the lumen by tumour growth. At high power the tumour characteristically consisted of rounded, monomorphic cells arranged around capillaries (Fig. 2). However, in some areas the cells became smaller, exhibiting a hyperchromatic ovoid to spindled nucleus. Mitoses and necroses were absent. Within the tumour, blood vessels of arterial or venous subtype without relationship to the tumour cells could be detected.

Immunohistochemistry revealed the tumour cells to be positive for vimentin, smooth muscle-type actin, broad-range muscle type actin and myosin. With antibodies to S-100 protein and panneurofilament protein small nerves within the tumour stroma could be detected, whereas the tumour cells were consistently negative. Applying antibodies to desmin, the muscular wall of the vein was strongly positive and was thus clearly discernible from the desmin-negative tumour (Fig. 3).





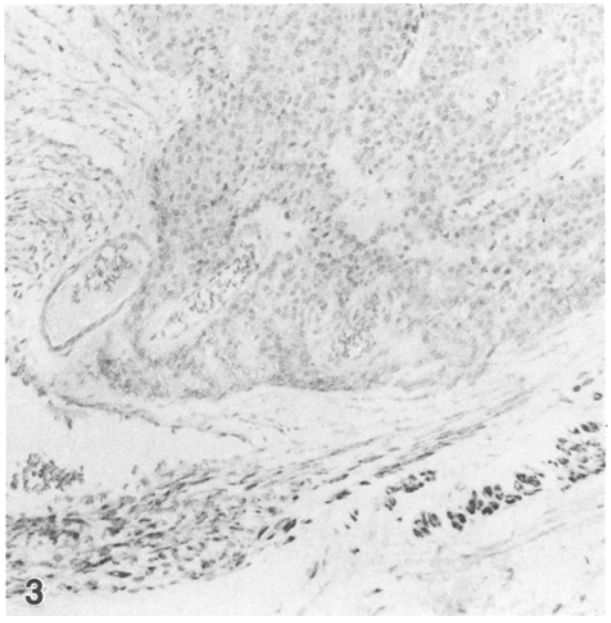


Fig. 1. Dilated blood vessel, the lumen of which is nearly totally filled out by a tumour. Thus between the vessel wall and the tumour only slit-like spaces remain. H&E, ×25

Fig. 2. Tumour origin from the vessel wall reveals peripherally the vascular smooth muscle layer (*asterisk*), then fibrous tissue and centrally a typical glomus tumour. H&E, ×250

Fig. 3. Smooth muscle cells of the vascular wall are immunopositive for desmin, whereas the tumour cells (*upper half*) are not reactive. APAAP method, ×180

The tumour cells were immunonegative for F VIII-RAG and myoglobin. However, the cell layer over the luminal surface of the tumour was immunopositive for F VIII-RAG, corresponding to its endothelial nature.

Discussion

In general glomus tumours are located in the extremities and display a characteristic pattern composed of monomorphic rounded cells arranged around a varying number of small capillaries (Enzinger and Weiss 1988). Deviations from the rule concern unusual localizations including bone (for review, see Sunderraj et al. 1989) and stomach (for review, see Kanwar and Manaligod 1975), oncocytic cytological features (Slater et al. 1987) and transition to malignancy (Aiba et al. 1988; Enzinger and Weiss 1988). However, an intravascular glomus tumour as in our case appears never to have been described in the literature.

The histological diagnosis of the case presented was based on the characteristic morphology (Enzinger and Weiss 1988) as well as on immunohistochemical investigations revealing the tumour cells to be immunopositive for myosin (Miettinen et al. 1983; Aiba et al. 1988) and actin (Aiba et al. 1988; Miettinen 1988; Dervan et al. 1989). These findings confirm the myogenous nature of the tumour cells (Miettinen et al. 1983; Dervan et al. 1989) and correspond to the presence of microfilaments detected by electron microscopy (Tsuneyoshi and Enjoji 1982; Miettinen et al. 1983).

The intravascular localization of the tumour may be explained in different ways. Firstly, the tumour could have grown into the lumen from outside. This can be ruled out by the fact that the vessel wall was intact and no tumour cells were detectable in perivascular areas. Secondly, special mesenchymal cells of the vessels wall could have differentiated to smooth muscle-like glomus tumour cells. However, such cells have never been described in normal veins. Thirdly, and most convincing, the affected blood vessel may represent the venous part of an arteriovenous anastomosis where the involved vessels, in particular the directly anastomosing vessels, are known to contain "epithelioid (glomus) cells" within the wall (Popoff 1934). These cells may be regarded as the site of tumour origin. This would explain satifactorily that the given tumour was covered by an intact vascular muscle wall and has pushed forward the endothelial layer by protruding into the lumen.

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