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LETTER TO THE EDITOR

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# Primary intimal type leiomyosarcoma with rhabdomyosarcomatous differentiation of the thoracic aorta

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Sirs: We have read the paper by Clelia Miracco et al. [1], reporting a case of a primary intimal type leiomyosarcoma with rhabdomyosarcomatous differentiation of the thoracic aorta, with great interest. We would like to add two additional cases to the still relatively small number of sarcomas of the great vessels (thoracic aorta, and inferior vena cava ([IVC]) described in the English literature in the last century [1]. We have diagnosed these cases in our department during the last 2 years. One patient was female; her main complaint was severe back pain, and a CT scan revealed an aneurysm of the thoracic aorta with imminent rupture. Urgent resection of the aorta had to be performed, since the aneurysm ruptured while the patient was waiting for a scheduled operation. Microscopic examination revealed an epithelioid leiomyosarcoma (LMSC) in the vessel wall, which was almost evenly distributed in all layers, the main part being at the site of the rupture, and in the immediate vicinity (Figs. 1, 2). The diagnosis was confirmed by positive immunohistochemical reactions for smooth muscle actin (SMA; Fig. 3), desmin, and vimentin antibodies. Following the histological diagnosis, a meticulous search for a primary tumour was unsuccessful. Finally, a primary LMSC of the aorta was diagnosed. The patient died 5 months later of widespread metastases (vertebrae, pelvis). The rapid clinical deterioration and the quick development of metastases indicate an intimal-type LMSC of the aorta; however, no autopsy was performed.

The other case was in a male patient with low back pain. The CT scan revealed a huge retroperitoneal tumour, probably of adrenal gland origin. A small piece of the tumour was sent for intraoperative frozen examination. On the basis of the clinical information provided and the histological picture (whorling of bland-looking tumour cells with elongated eosinophilic cytoplasm, no mitoses, no necrosis), a probably benign nerve sheath tumour of the adrenal gland was diagnosed. The surgeon later reported that the tumour had no association with the adrenal gland and had an easily removable retroperitoneal margin, but was attached to the wall of the IVC, so that only a part of the tumour was resected. Macroscopically, a greyish-white firm lobulated tumour with areas of necrosis was seen, with an advancing margin on one side. It was evident that the resection line traversed the mass. No association with the resected adrenal gland was found. On histological examination part of the tumour showed a completely benign picture, consistent with the findings observed on the frozen sections. A few sections showed rather dedifferentiated areas with focal necrosis and numerous typical and atypical mitoses, striking indications of an obviously malignant neoplasm (Fig. 4). Immunohistochemical reactions showed strong positivity for SMA Fig. 5), desmin, and vimentin. Finally, a partly well-differentiated leiomyosarcoma, originating most probably from the wall of the IVC, was diagnosed. The patient remains well 4 months after the operation. The clinical and histological pictures of our reported cases showed the characteristics of leiomyosarcomas of the great vessels described in earlier papers reviewing relatively large series of cases [3–5].

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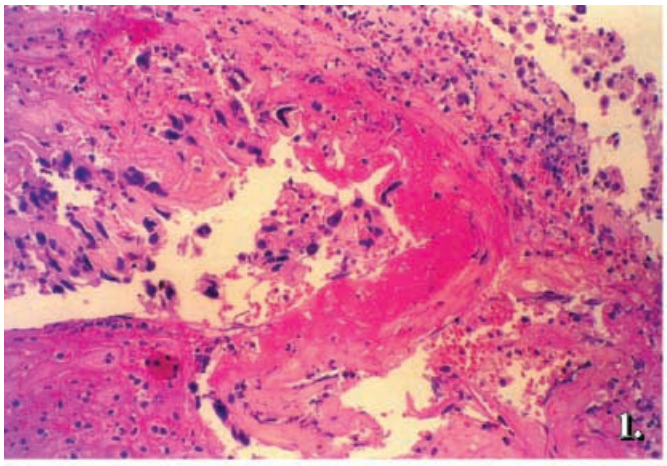


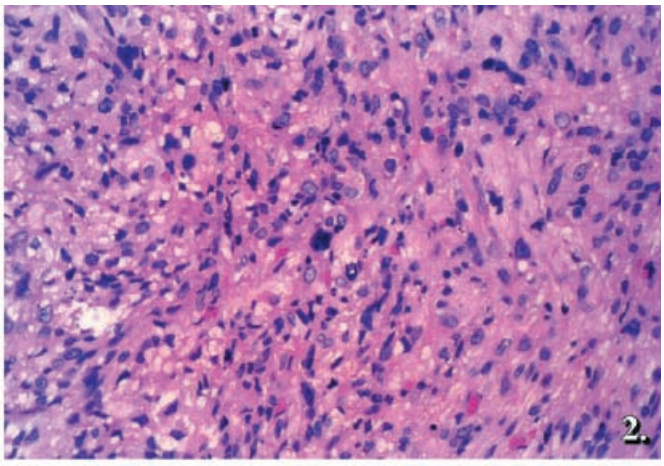


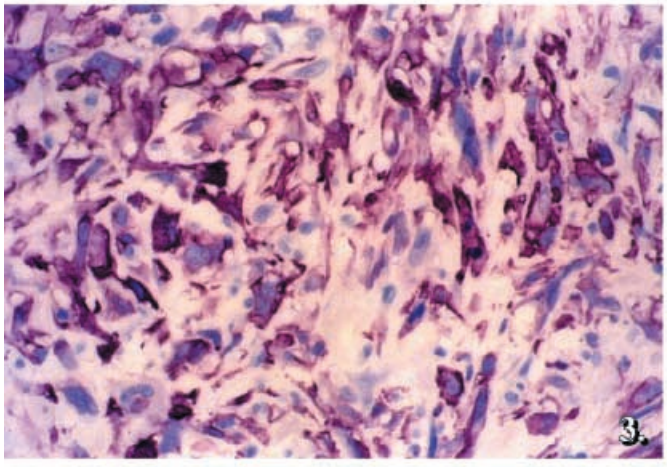


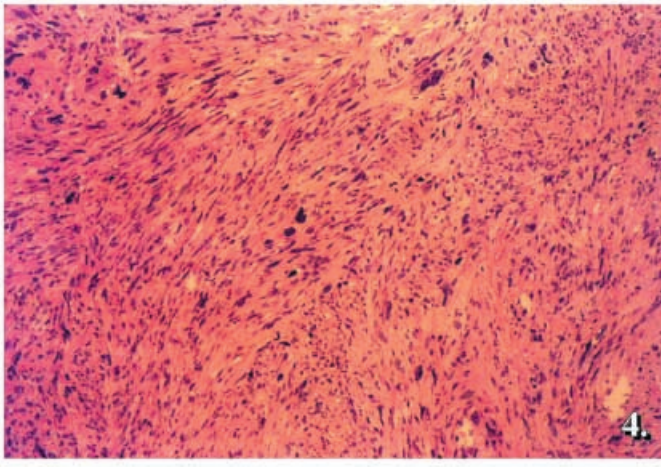


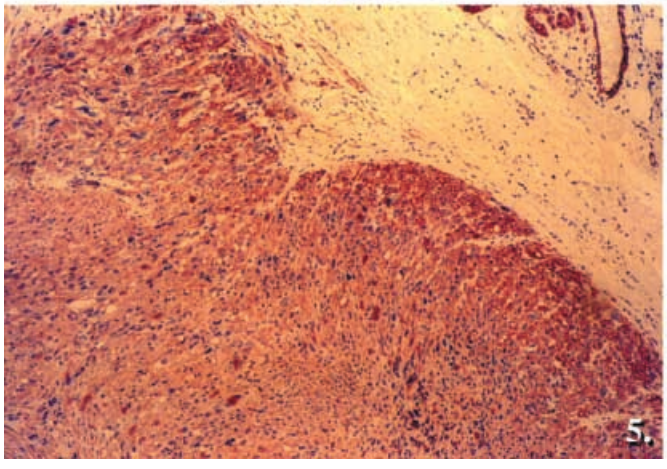


**Fig. 1** Case 1. Site of rupture of the wall is covered by fibrin, but some tumour cells are also visible. H&E, ×100

**Fig. 2** Case 1. Epithelioid smooth muscle cells, with atypical, hyperchromatic nuclei, with mitotic figures. H&E, ×400

**Fig. 3** Case 1. Immunoreactivity for smooth muscle actin. Peroxidase, ×400

**Fig 4** Case 2. View of the obviously malignant part of the tumour. H&E, ×400

**Fig. 5** Case 2. Immunoreactivity for smooth muscle actin. Note pushing border at the retroperitoneal surface. Immunoperoxidase, ×200

## References


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