10.1053.paor.2000.0245 available online at http://www.idealibrary.com on IDEAL

CASE REPORT

Leiomyosarcomas of Great Vessels

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Sarcomas of the great vessels are rare. Altogether 400 such cases have been described in the aorta, the pulmonary artery, and inferior vena cava. The clinical symptoms are generally related to embolic phenomena, aneurysm formation, and widespread metastases, especially to bones. With improved diagnostic modalities more cases are diagnosed and treated surgically. Resection of the tumor may prolong the patient's life. In this paper authors present two cases of such rare sarcomas. In our first case a tumor has developed in the thoracic aorta with symptoms of imminent aortic dissection. The tumorous nature of the lesion was revealed only histologically, since neither the operation, nor macroscopic picture gave any clue to its tumorous nature. The second case was a male patient with a huge retroperitoneal tumor arising from the inferior vena cava, which was clinically suspected to be a carcinomaarising in the adrenal gland. (Pathology Oncology Research Vol 6, No 3, 233–236, 2000)

Keywords: leiomyosarcoma, thoracic aorta, aortic dissection, inferior vena cava, retroperitoneal tumor

Introduction

Tumors of the great vessels are rare. About 400 cases have been described so far, the inferior vena cava (IVC) being the most common site of origin. This is followed by the pulmonary artery, and the thoracic aorta as the least frequent location.[5,9] The clinical symptoms and metastatic potential of these tumors are related to their position in the vessel wall. Luminal (intimal) sarcomas metastasize early, frequently, and have a poor prognosis. Patients with mural sarcomas do relatively better, with metastases occurring later, and a longer survival. In our report we describe two new cases of leiomyosarcoma with vascular origin. Patient one was an untreated hypertensive female with symptoms of imminent rupture of an aortic dissection. Even during the operation there was no suspicion of the tumorous nature of the lesion. Macroscopically there was a slight thickening of the vessel wall, but even after knowing the microscopic picture, macroscopic review still did not show a clearly evident tumor. Patient two was a 70 year old male whose tumor of the IVC presented as a retroperitoneal tumor of adrenal gland origin. We think that although these tumors are rare, both clinicians and pathologists should keep in mind the possibility of their presence in the listed clinical situations, since preoperative suspicion could improve the surgical intervention and postoperative results.

Recived: Dec 22. 1999; *accepted:* July 20, 2000

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Clinical history and pathologic findings

Patient 1

A fifty year old female patient was admitted to the Cardiovascular Surgical Department of the Semmelweis University. She had been suffering from hypertension for several years. The symptoms were consistent with a threatening aortic dissection. The diagnosis was confirmed by CT scan of the chest (*Figure 1*). Urgent operation was performed, for the sudden rupture of the thoracic aorta, while waiting for a scheduled graft implantation. The resection specimen was a 10 cm long segment of the thoracic aorta. The site of the rupture was visible. Macroscopically it was covered by a thin necrotic bloodstained material, but no remarkable tumor mass, nor luminal thrombus or marked thickening of the vessel wall was seen. No macroscopically visible tumor could be recognised, even after the review of the resection specimen, following the first microscopic examinations. Microscopically a part of the vessel wall was replaced by tumor tissue, mainly in the vicinity of the site of the dissection (*Figure 2*). The tumor was composed of highly atypical

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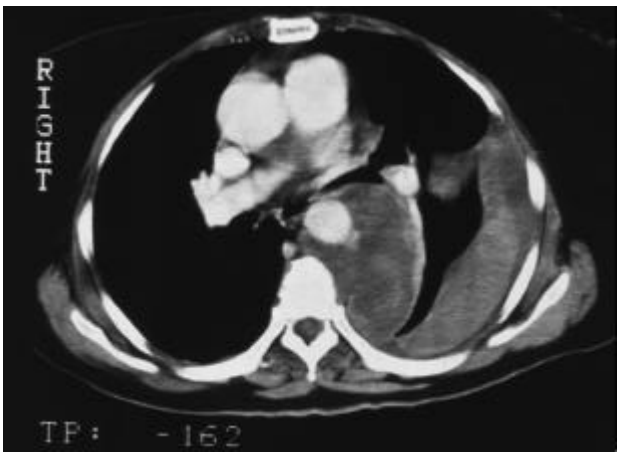

Figure 1. CT scan of patient 1, after rupture has occurred, note widened aortic wall and haematoma along the chest wall.

epithelioid cells with hyperchromatic nuclei, prominent nucleoli and relatively abundant eosinophilic, round cytoplasm. Numerous mitoses, both typical, and atypical were readily visible (*Figure 3*). On low power examination a barely perceptible whorling was seen. Immunohistochemical reactions were negative with cytokeratin, (CK) epithelial membrane antigen (EMA), HMB45, but showed strong positivity with vimentin, desmin and smooth muscle actin (SMA) antibodies. As tumor cells were almost evenly distributed in both the intima, media and the adventitia , at first place a metastatic tumor was suspected. Since the review of the CT images, and the supplementary clinical examinations failed to find a primary tumor, an intimal leiomyosarcoma of the aorta was diagnosed. The patient died five months later with widespread metastases of the vertebrae, the pelvis and the abdominal cavity. Although no autopsy was performed, the lack of primary tumor, and the rapid clinical deterioration makes the histological diagnosis evident.

Patient 2

The 70 year old male patient was admitted to the 1st Department of Surgery. He had progressively intensifying abdominal complaints and low back pain of two years duration. On CT scan, a huge retroperitoneal mass was found with dislocation of the IVC. The tumor was suspected to arise from the right adrenal gland. In the course of the operation a small piece of the tumor was sent for frozen examination, with the clinical data reporting an adrenal gland tumor. Based on the histological picture (whorling of elongated mesenchymal cells, no atypia, no necrosis) and the clinical data, a benign mesenchymal tumor, probably of nerve sheath origin, was diagnosed. Postoperative clinical information revealed that the retroperitoneal margin of the tumor was easily removable, but the main part was attached to the wall of the IVC, therefor only a part of the mass could be resected. Upon macroscopic examination a grayish-white, lobulated, firm mass was found, which showed no connection with the adrenal gland. The latter was also removed during the course of the operation, and it showed a completely normal gross morphology. Microscopically the tumor was composed of whorling fascicles of smooth muscle cells, possessing long, eosinophilic cytoplasm, with empty looking, cigar shaped nuclei, and prominent nucleoli (*Figure 4*). Numerous mitoses, both typical and atypical were readily visible, (35-40/10 HPF) (*Figure 5*). Areas showing completely benign morphological picture could also be detected, but, we do not think that the tumor arose in a benign leiomyoma. The tumor was located in the close vicinity of the adrenal gland, but no association with either the cortex or the medulla was recognised. Immunohistochemical reactions were positive for vimentin and SMA in the tumor cells, while S-100 reaction showed positivity only in the cells of the adrenal medulla. The Ki-67 proliferation marker showed strong positivity in many cells: 80

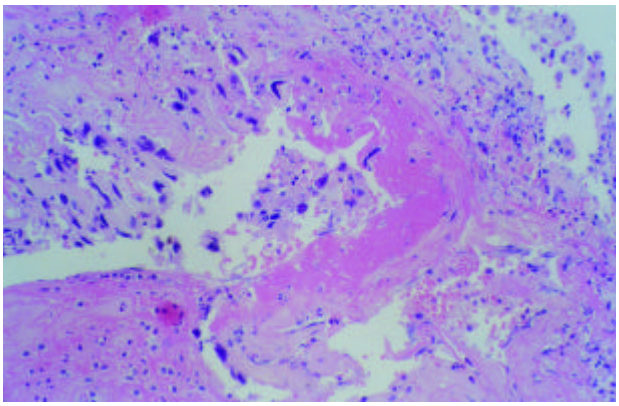

Figure 2. Case 1. Site of rupture is covered by fibrin, the wall is disrupted, note partly necrotic tumor mass at the bottom of the picture.

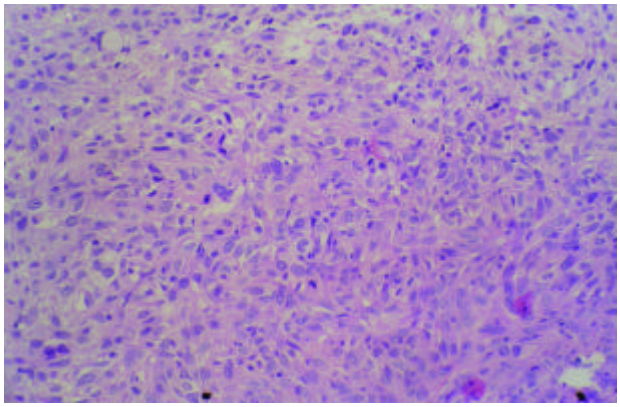

Figure 3. Case 1. High power picture of the epithelioid cells. Numerous mitoses, both typical and atypical can be readily recognised.

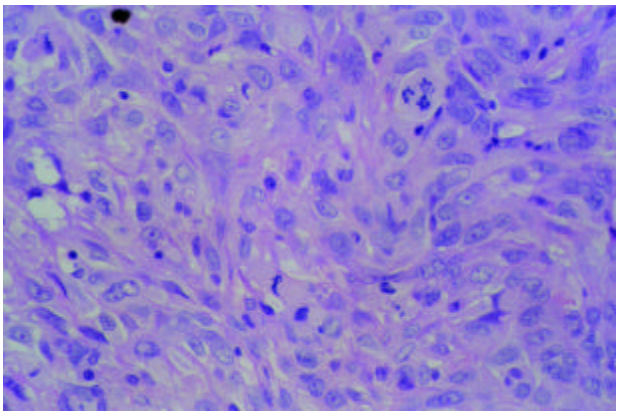

Figure 4. Case 2. Low power view of the tumor. Compressed adrenal gland at the left upper corner of the field.

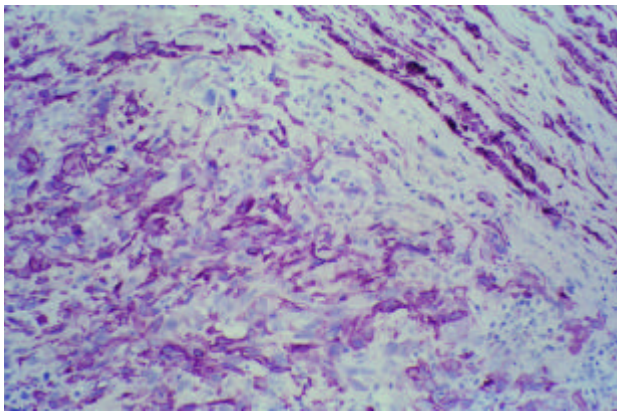

Figure 5. On higher power, whorling, numerous mitoses, hyperchromatic nuclei can be easily recognised.

positive nuclear staining/10 high power fields. Based on the localisation of the tumor, the morphological, and immunohistochemical and clinical findings, a leiomyosarcoma, most probably arising in the wall of the IVC was diagnosed.

Discussion

Tumors of the great vessels occur rarely.[1-6] Those arising in the intima, have a dismal prognosis, since intimal origin is a source of widespread metastases, and seeding of the tumor to distant sites, occurs early. (There are exceptional cases reported in the literature, with long term survival.[11] These tumors are usually entirely intraluminal. They appear histologically as poorly differentiated mesenchymal tumors, showing immunohistochemical characteristics but of myofibroblastic origin.[3-5] Sporadic intimal sarcomas have also been reported showing areas of rhabdomyosarcomatous,[1,2] and angiosarcomatous[4] differentiation. Tumors arising in the pulmonary artery may show areas of osteosarcoma, and myxoid mesenchymal tumors.[4] Usually at the time of diagnosis, numerous metastases have already been occurred. Preoperative diagnosis is almost impossible, since the tumor mass is usually relatively small, and remains hidden by imaging technics, (it may have a deceptively similar picture to a mural thrombus). However, there might be a need for intraoperative histological examination in suspicious cases presenting in the form of aortic aneurysms. Of course surgeons have to be aware of the possibility of the presence of such a tumor, since there are usually very slight alterations giving a clue to the suspicion, like in our first case.[1,8] The cases arising in the media have far better prognosis, though inevitably leading to the death of the patient.[4,5] They may remain hidden for a long time, they have much lower metastatic potential since until tumorous protrusion into the lumen has not occurred, and direct arterial seeding is not a feature. These tumors prove to be better differentiated histologically than their intimal counterparts.[4,5] The LMSCs that arise in the IVC, have better survival rates than those cases arising in the aorta.[4,5] These tumors are more frequently well differentiated leiomyosarcomas, and thorough examination of many histological slides is warranted to confirm the malignant nature of the neoplasm. Small areas composed of less well differentiated tumor cells are usually visible, as well as the relatively high mitotic rate, and areas of necrosis. The above listed features are the clues for the diagnosis of a malignant neoplasm.[4] Radiologists should be aware of the existence of malignant tumors arising in the great vessels: they should keep in mind, that behind a threatening aortic dissection there might exist a primary malignant tumor of the aorta, and particularly behind a huge retroperitoneal mass, there might be a hidden tumor of the IVC. When diagnosed preoperatively, they might be widely resected followed by the prompt implantation of grafts, which may secure a relatively long tumor free survival for the patiens[14] as could have been the fact in our second case. Since there was no radiologic and surgical suspicion of an IVC origin of the mass, only a part of the tumor had been resected, with no graft implantation, and the clinical data given to the pathologist were misleading.

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