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Thrombectomy Discloses Intravascular Growth of Chondroid Liposarcoma Mimicking a Long Distance Vena Cava Thrombosis

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Abstract. *A 22-year-old woman with a newly detected chondroid liposarcoma located in the iliac muscle was diagnosed as having bilateral pulmonary embolism. Gadolinium-enhanced MRI further revealed a long distance thrombus reaching from the iliac vein to the right atrium. The thrombus was attributed to a hypercoagulability state which has been described for chondroid liposarcoma. High-dose chemotherapy with autologous stem cell support reduced the tumor burden and led to a symptom-free interval of 6 months. Despite therapeutic anticoagulation, repeated imaging showed no reduction or remodeling of the thrombus. However, when the thrombus progressed again, the patient underwent cardiac surgery and histology revealed the intravascular growth of the known chondroid liposarcoma. We conclude that in sarcoma patients intravascular tumor growth must be kept in mind when imaging is suggestive for thrombosis.*

Liposarcoma is the second most common malignant tumor of the soft tissues (16-18%) and accounts for approximately 20% of all mesenchymal malignancies (1, 2). Most commonly liposarcomas are diagnosed in the lower extremities and the retroperitoneum (3, 4). Intravascular tumor growth has been reported for different sarcoma and lymphoma subtypes. We here report a patient with chondroid liposarcoma that was diagnosed to have a long distance thrombosis, possibly due to paraneoplastic hypercoagulability, and was later diagnosed to have intravascular sarcoma growth. Intravascular liposarcoma as an atypical manifestation must be considered as a differential diagnosis in patients with a suggestive clinical course.

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Case Report

A 22-year-old woman presented with lower backache radiating in the left flank without any history of trauma. During the previous 2 weeks she had experienced progressive fatigue, a weight loss of about 3 kg and dyspnoe for 1 week. On presentation, the pertinent findings of physical examination were an increased sensitivity of the lower back to percussion. Laboratory findings were normal, except for elevated D-dimers. Chest CT showed bilateral pulmonary embolism. Gadolinium-enhanced MRI with 5mm contiguous axial and sagittal T_1-weighted spin echo and sagittal T_2-weighted turbo inversion recovery magnitude (TIRM) disclosed a tumor in the left iliac muscle. The tumor and its metastases in the lumbar spine and the ilium bone had high signal intensity in T_2-weighted TIRM images with patches of low signal in between. Furthermore a thrombus along the inferior vena cava into the right atrium was detected by cardiac MRI (Figure 1A and 1B). CT-guided biopsy of the intramuscular tumor was performed and histopathological assessment revealed a liposarcoma with myxoid and round cell parts.

Despite therapeutic anticoagulation no significant remodeling of the intravascular thrombus was observed. Chemotherapy, consisting of 6 cycles VIP-E (Etoposid-Phosphate, Ifosfamid, Cisplatin, Epirubicin) as well as high-dose chemotherapy with Melphalan and Busulfan with autologous stem cell transplantation, reduced the iliac tumor volume about 50%. This stable condition was conserved for 7 months when the patient was readmitted with the clinical signs of cardiac insufficiency. Echocardiography revealed a progression of the thrombus through the right atrium into the right ventricle. Cardiac surgery with thrombectomy was performed and histology disclosed intravascular growth of the known chondroid liposarcoma. The macroscopic findings of the intravascular tumor are depicted in Figure 1 C and 1D. Histomorphology disclosed the chondroid liposarcoma detected previously in the iliac muscle with myxoid (E) and chondroid (F) parts.

Discussion

Liposarcomas are currently classified into five subgroups: 1, well-differentiated type; 2, myxoid type; 3, round cell type; 4, pleomorphic type; 5, dedifferentiated type (1, 5). In our case liposarcoma had the histopathological criteria of the myxoid and round cell subtype and thus belongs to the low-grade group with a

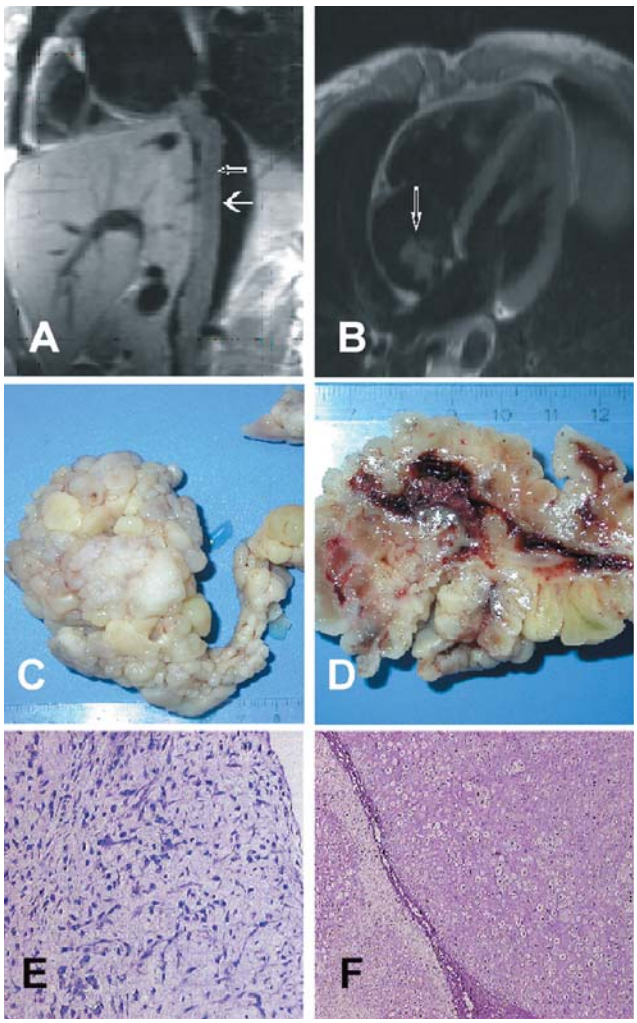

Figure 1. *A: Sagittal magnetic resonance imaging (MRI) of the thorax reveals the long distance thrombus.*
B: Four chamber coronal view. The thrombus floats freely within the inferior vena cava extending into the right atrium. Imaging was performed on initial diagnosis of the thrombus.
C and D: The resected tumor displays a chondroid picture without any evidence of thrombotic material.
E and F: Histomorphology discloses the chondroid liposarcoma detected previously in the iliac muscle. (E: myxoid part of the tumor [Giemsa stain, magnification: x200]; F: chondroid part [Hematoxilin-Eosin stain, magnification x200])

better prognosis than that of other types (5). Myxoid and round cell liposarcoma, even if still classified by the World Health Organisation as two distinct subtypes, share both clinical and morphological features. As in our case, lesions combining both patterns are very frequent and wide agreement exists in considering round cell liposarcoma as the high-grade counterpart of myxoid liposarcoma. Furthermore both entities share the same reciprocal translocation t(12;16)(q13;p11) (6).

The imaging appearances of liposarcomas depend on the histological subtypes and thus, in our case, contain soft tissue and fatty components (7, 8). Evans *et al.* demonstrated that the biological behavior of these tumors is not only dependent on tumor morphology, but also on localisation (9, 10).

Many cancer entities have been described as being associated with paraneoplastic syndromes such as hypercoagulable state, which is most frequently seen in adenocarcinoma or promyelocytic leukaemia, but also for different types of sarcomas, including liposarcoma (11). Thus, the development of the long distance thrombosis of our patient, in atypical localisation and extent, was initially suspected to be due to paraneoplastic hypercoagulability or to tumor invasion into the vessel wall. The fact that the intravascular "thrombus" decreased was attributed to therapeutic anticoagulation and not to the chemotherapy as both therapeutic measures overlapped.

Since the thrombus progressed, despite therapeutic anticoagulation, into the right ventricle, the patient underwent thrombectomy and the intravascular sarcoma growth was detected. The clinical lesson we have learnt is that intravascular growth must be kept in mind when long distance thrombosis is detected in patients with liposarcoma. Although intravascular growth of sarcoma has been described in autopsy studies this has, to our knowledge, never been reported for chondroid liposarcoma *in vivo*.

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