

Simultaneous pulmonary and inferior vena cava tumor thromboembolism secondary to retroperitoneal pleomorphic liposarcoma

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Abstract

Retroperitoneal pleomorphic liposarcoma (RPLS) is the least common but the most aggressive subtype of liposarcoma. We herein report a case of tumor embolism (TE) to the inferior vena cava (IVC) and pulmonary arteries in a 54 years old woman with RPLS. The case suggests that fusion fluorine-18-fluorodeoxyglucose positron emission tomography/computed tomography (^{18}F -FDG PET/CT) can accurately detect occult TE. It also illustrates the usefulness of computed tomography (CT) and high ^{18}F -FDG uptake by PET/CT imaging for diagnosing such findings as filling defect in the veins and beaded pulmonary arteries as TE. To our knowledge, this is the first reported case of simultaneous pulmonary and inferior vena cava thromboembolism secondary to RPLS. And occult TE from RPLS has not been reported previously by ^{18}F -FDG PET/CT.

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Introduction

Retroperitoneal pleomorphic liposarcoma (RPLS) is rare, with a high incidence of recurrence and metastases. Thromboembolism is a common and severe complication in oncological patients. Here, we describe a 54 years old woman with simultaneous pulmonary and inferior vena cava (IVC) thromboembolism secondary to RPLS. According to our knowledge, this is the first reported case of simultaneous pulmonary and IVC tumor embolism (TE) secondary to RPLS. Occult TE from RPLS has not been reported previously by fusion fluorine-18-fluorodeoxyglucose positron emission tomography/computed tomography (^{18}F -FDG PET/CT) in medical literature.

Case Report

A 54 years old woman presenting with right flank pain, nausea and vomiting. There was no associated anal cessation of exhaust and defecation, hematuria, waist movement restriction, or weight loss. No symptoms or signs associated with thrombosis appeared in her past and/or her family history. Physical examination showed normal breath and heart sounds, no palpable mass or lymph nodes. Magnetic resonance imaging (MRI) identified a large right retroperitoneal soft-tissue mass. Laparoscopic retroperitoneal biopsy revealed pleomorphic liposarcoma (PLS).

In our hospital, laboratory examinations including hemoglobin, leukocyte count, platelet count, prothrombin time-international normalized ratio-, lactate dehydrogenase, serum glucose, blood urea nitrogen, creatinine, bilirubin, total protein, albumin, and electrolyte levels were all within normal limits. Chest X-rays were negative (Figure 1A). The initial staging ^{18}F -FDG PET/CT images (Figure 2A) revealed intense right retroperitoneal uptake and hypermetabolic thoracic nodular foci. Fusion PET/CT (Figure 2B) showed retroperitoneal tumor uptake as well as right ureteral and inferior vena cava (IVC) invasion (SUVmax 16.5). There was also fork-shaped significant ^{18}F -FDG uptake (SUVmax 5.2) in the right common basal artery and its branches (Figure 2C). These PET/CT findings suggested tumor thromboembolism to the IVC and the pulmonary arteries. The

patient then experienced abrupt onset of dyspnea. Subsequent chest X-rays revealed a nodule (arrow) in the right lower lung (Figure 1B). Contrast-enhanced CT of the abdomen (Figure 3A) and chest (Figure 3B) showed the enlarged retroperitoneal mass with TE in the IVC and the common basal pulmonary artery branches further suggesting the diagnosis of tumor thromboembolism.

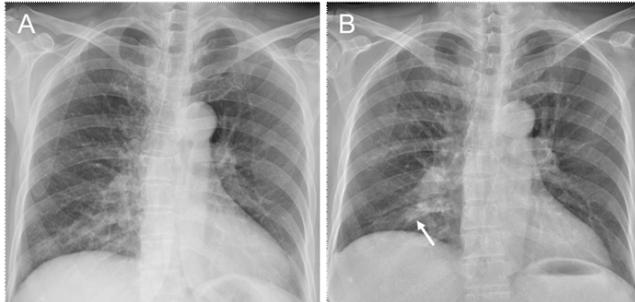


Figure 1. Chest X-rays. (A) negative; (B) pulmonary tumor thromboembolism (arrow).

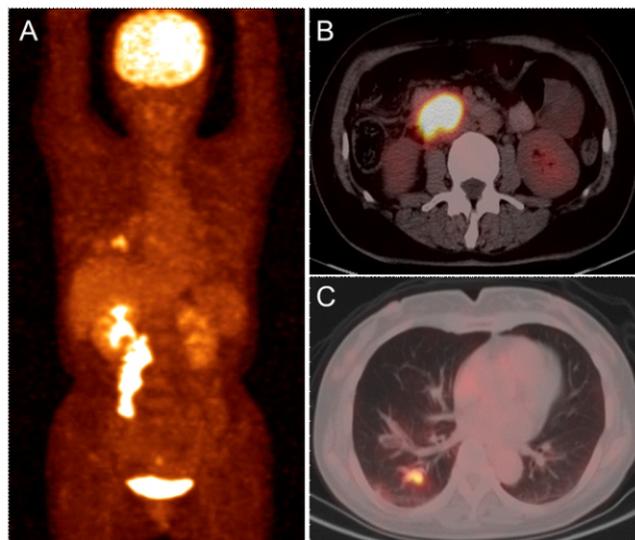


Figure 2. Fluorine-18-FDG PET/CT imaging of primary tumor and tumor embolism. (A) MIP image with RPLS and IVC tumor thromboembolism (arrowhead), pulmonary tumor thromboembolism (short arrow) and hydronephrosis of the right kidney and right ureter (long arrow); (B) RPLS and IVC tumor thromboembolism (arrowhead); (C) pulmonary tumor thromboembolism (short arrow).

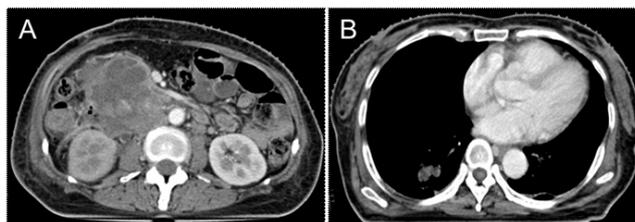


Figure 3. Contrast-enhanced computed tomography of the abdomen and chest. (A) retroperitoneal tumor with TE in the IVC (arrowhead); (B) pulmonary TE (arrow).

She started treatment of low-molecular-weight heparin due to pulmonary and IVC thromboembolism. Owing to the tumor being so large and unresectable, we administered intensive chemotherapy: high dose of epirubicin (70mg), and

dacarbazine (500mg). Palliative local radiation was also administered. Follow-up by abdominal CT showed mild shrinkage of the tumor. At present, she survives 8 months after diagnosis.

Discussion

Pleomorphic liposarcoma confirmed in this case is the least common subtype of liposarcoma and only accounts for 5% of liposarcoma cases [1]. It is often found in the deep soft tissues of the extremities in late adulthood [2] and only rarely has a primary retroperitoneal location [3]. In a series of 57 patients with PLS, most of the tumors (65%) originated in the extremities, with only 7% originating in the retroperitoneum. These are aggressive high-grade sarcomas with a high propensity of recurrence and metastases. Pleomorphic liposarcoma appears as a well-circumscribed mass containing little or no fat on CT and MRI, often with areas of necrosis and hemorrhage [4]. Imaging by PET/CT with ^{18}F -FDG shows intense uptake in the lesions [5], as was the case in our patient (Figure 2B).

Thromboembolism is a common and severe complication in adult cancer patients [6]. Sarcoma is one of the most common sources of TE that invade the IVC and lungs [7]. In most of histologic types, a previously published study has shown that soft tissue sarcomas have a tendency to metastasise haematogenously to the lungs whereas metastases to lymph nodes are not common [8]. Sarcoma tumor cells can be carried in the blood stream and embolise pulmonary circulation [7]. Few cases of RPLS have been reported [5, 9]. To our knowledge, this is the first reported case in medical literature of simultaneous pulmonary and IVC TE secondary to RPLS demonstrated by PET/CT.

Many factors can contribute to thromboembolism in patients with cancer, including intravascular tumor extension, stasis of blood flow, vascular endothelial damage and hypercoagulation [10]. Our case had no preexisted risk factors or disease associated with clotting disorders. Because biopsy on the site of thromboembolism is dangerous, the pathogenesis of thromboembolism in our patient could only be inferred by the radiographic findings. Based on these findings, thromboembolism in our case was considered to be direct intravascular extension of the primary tumor into the IVC and metastases into the pulmonary arteries.

The diagnosis of thromboembolism plays an important role in staging, treatment decisions and prognosis of many malignant tumors. Clinical manifestations of tumor and of bland of emboli are often difficult to distinguish. Imaging modalities are of great significance in the diagnosis of tumor emboli. Usually, thromboembolism is difficult to diagnose by X-rays radiography. Recently, enhanced CT has been recommended for assessment of thromboembolism. At CT, the specific signs of TE include mass-like enhancement of filling defect in the vein and beading of peripheral pulmonary arteries [11]. This finding is very rare and the sensitivity of CT needs to improve. Additionally, in some cases of allergy and/

or chronic renal failure, which is very common in cancer patients, we should use lung perfusion scintigraphy (LPS) instead of contrast enhancement as LPS had been confirmed to be a simple, quick and inexpensive examination with no preparation/contraindication [12]. "Typical" ventilation-perfusion findings include segmental perfusion deficits with normal ventilation [13, 14]. Anyhow, the ventilation-perfusion scan is also not helpful in distinguishing TE from thromboembolic disease.

Since ^{18}F -FDG PET/CT imaging can identify and characterize malignant lesions on a functional level, PET/CT can effectively distinguish TE from thromboembolism and easily determine the cranial extension of TE. The uptake of ^{18}F -FDG by TE has been reported to be considerably higher than that of thromboembolism [15]. The pattern of uptake based on most studies published to date can be either focal or linear along the course of the vessel [16]. Additionally, in previous study, occult TE was found incidentally in 0.12% of oncologic patients [17]. Imaging by PET/CT has the ability to detect occult tumor embolism in different parts of vasculature and in heterogeneous types of tumors as was in our case [17]. With the detection of occult tumors, PET/CT can be helpful in evaluating the correct stage, the treatment response and provides additional information on the survival rate of cancer patients [17]. In our case, the right common basal pulmonary artery tumor emboli were the only sites of distant metastases. So the patient was treated with intensive chemotherapy instead of surgery. There are many reports of TE in solid cancers such as lymphomas, tumors of the colon, pancreas, lung, breast, and thyroid, sarcomas and many others [17-22]. To the best of our knowledge, unsuspected TE from RPLS has not been reported previously by ^{18}F -FDG PET/CT in medical literature.

In conclusion, RPLS poses the possibility of thromboembolism to vena cava system and pulmonary vessels. Meticulous imaging studies such as CT or ^{18}F -FDG PET/CT can diagnose the primary tumor, the metastatic tumors or tumors embolism even occult. A filling defect in the veins, beaded pulmonary arteries on CT scans and significant ^{18}F -FDG uptake by ^{18}F -FDG PET/CT are valuable findings to diagnose tumor embolism.

The authors declare that they have no conflicts of interest.

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