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CASE REPORT

Rapid Progression of Angiosarcoma in a Man with a Left Psoas Muscle Hematoma and a Recent EVAR of Abdominal Aorta

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ABSTRACT

Angiosarcoma is a rare type of soft tissue cancer with several clinical presentations and a poor prognosis. We present a case of a 75-year-old man who was admitted due to anemia and fatigue. The patient had undergone an endovascular repair (EVAR) of a 9 cm infrarenal aneurysm of the abdominal aorta two months ago. A computed tomography (CT) scan of the abdomen on admission indicated a Type-II endoleak and a large hematoma of the left psoas muscle with multiple sites of intramuscular extravasation. Osseous metastases were found at the head of the left femoral head and at the iliac bones. A CT guided biopsy of the femoral head revealed an angiosarcoma of unknown primary site a few days after the patient had died from intra-alveolar hemorrhage caused by lung metastases.

Keywords: angiosarcoma, psoas hematoma, EVAR, osteolytic lesions, intra-alveolar bleeding.

INTRODUCTION

Angiosarcoma is a rare cancer comprising 1% of soft tissue cancers, which in turn consist of less than 1% of adult malignancies. They emerge from lymphatic or vascular endothelial cells, meaning that they can occur anywhere in the body (1). Due to their rarity, existing knowledge mainly originates from accumulation of case reports. They are more common in the older age, particularly between 60-71 years, without gender predilection, except for the cutaneous form, which has a male prevalence. The cutaneous form accounts for about half of cases, with the head and neck being most frequently involved. The rest are located in parenchymal organs such as liver, bone, heart, spleen or deep soft tissues. Less than 10% have no clear primary site and are found when they have already metastasized (2, 3).

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They are classified into cutaneous angiosarcomas, lymphoedema-associated angiosarcomas, radiation-induced angiosarcomas, primary-breast angiosarcomas, and soft-tissue angiosarcomas, each with its own subtypes (1). They are thought to arise spontaneously, although some reports describe angiosarcomas arising on previous benign vascular lesions (1).

The most common risk factors are history of radiation therapy and chronic lymphoedema (Stewart–Treves syndrome). Other risk factors are some familial syndromes (neurofibromatosis, Maffucci syndrome and Klippel-Trenaunay syndrome), exposure to toxins (vinylchloride, thorium dioxide, arsenic, anabolic steroids) and foreign substances (accidentally retained surgical gauzes, vascular and orthopaedic prostheses and gouty tophus). A correlation between vascular grafts and aneurysms on the site of the graft has also been suggested in literature (4-13).

The clinical presentation of angiosarcomas depends on their location. Cutaneous forms present as bruises, discoloured nodules, persistent ulcerations, while deeper soft-tissue forms located in mediastinum, peritoneum and retroperitoneum, present as rapidly growing palpable masses causing pain and discomfort. Cardiac angiosarcomas present with dyspnea and chest pain as well as hypotension, tachycardia and syncope. They form a well-defined mass that extends from the pericardium into the cardiac chambers or a diffusely infiltrating mass with pericardial and sometimes epicardial extension. As hepatic lesions most commonly present with abdominal pain, weakness, weight loss, jaundice, hepatomegaly and ascites. Osseous angiosarcomas merely of long bones such as tibia, femur and humerus cause bone swelling and pain of the affected site and pathological fractures (3). Angiosarcomas deriving directly from the aorta are very rare and they can present as space-occupying and bulky limiting the luminal diameter masses. Finally, there have been reports of aortic angiosarcomas presenting as aneurysms, leading to delayed diagnosis, all of them having a broad range of symptoms (lower/upper limb claudication, abdominal/back pain, weight loss, fever, fatigue, dyspnea and stroke). They can be distinguished from thrombus or atherosclerotic disease on CT angiography because they are hyper-enhanced on arterial phase. Additionally, peri-aortic infiltration of the mass can be observed suggesting tumor rather than thrombus or atherosclerotic disease (14-17).

Angiosarcomas usually spread haematogenously, with the lungs being the most common site of metastasis, and may present as lung hemorrhage, haemorrhagic pleural effusion or pneumothorax. Other common sites include liver, bone, soft-tissue structures and lymph nodes (2). The prognosis is very poor, especially if metastatic disease is found upon diagnosis. Even with localized disease, the five-year survival is 35%, with a median survival of seven months (1, 2).

The treatment requires a multidisciplinary approach. Localised disease is usually treated with surgical removal with additional adjuvant radiotherapy and/or chemotherapy. For metastatic angiosarcoma, cytotoxic chemotherapy is the primary treatment option (anthracyclines, ifosfamide, and taxanes) (1, 2). ❑

CASE PRESENTATION

A 75-year-old man in good functional status presented to the ER due to progressive fatigue/weakness and an episode of presyncope three days ago. No additional symptoms were reported. The patient had a history of arterial hypertension, dyslipidemia and gastroesophageal reflux disease. He was an ex-smoker and had undergone an EVAR with ALTO endograft of a 9 cm infrarenal abdominal aortic aneurysm, which was found incidentally two months ago on a CT scan due to severe back pain. Upon recent presentation, he was febrile (38°C), hemodynamically stable without oxygen supplementation needs. The clinical examination revealed a sizable hematoma on the left thigh. The electrocardiogram showed sinus rhythm without sings of ischemia and the chest x-ray had no remarkable findings. The laboratory tests showed significant anemia (Hb 6 g/dL), that was normocytic, with remarkably high inflammation markers (WBC 22000 k/μL (neutrophilia) and an elevated CRP. No platelet, coagulation or hemolytic disorders were observed.

An abdominal CT scan was performed on an emergency basis in order to exclude leakage from the EVAR. The CT scan showed a type II endoleak, with the aortic walls around the graft being within the previous measurements, which was not a sufficient cause for the hemoglobin drop. However, a large hematoma of the left psoas and multiple sites of extravasation within the ipsilate-

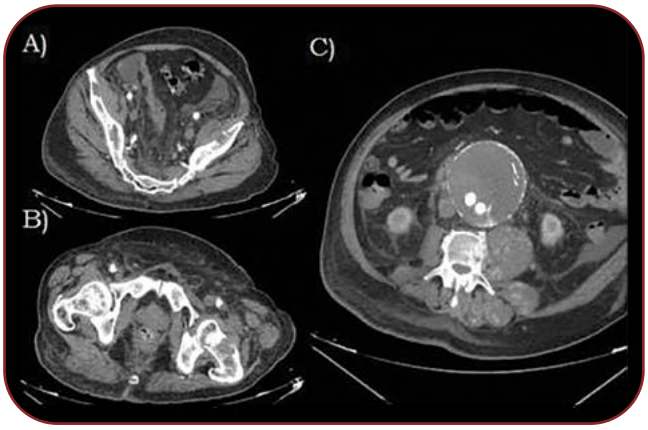

FIGURE1. A) Osteoiytic lesions on both iliac bones
B) Osteoiytic lesions on the head of the left femur
C) Hemorrhagic lesions within the left psoas and ipsilateral paraspinal muscles

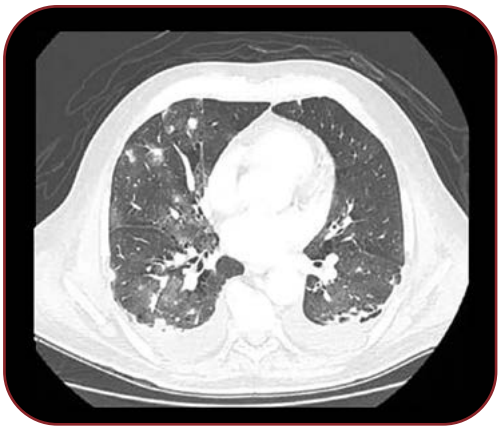

FIGURE 2. Multiple bilateral nodular lesions in both lungs with halo sign (probable hemorrhagic metastases)

ral paraspinal muscles were revealed. Furthermore, osteolytic lesions on both iliac bones on the head of the left femur and on the third lumbar vertebra were described (Figure 1). The aforementioned findings were attributed to secondary metastatic lesions of a malignancy of unknown primary site.

After general surgery, invasive radiology and vascular surgery consultation, conservative treatment was decided as the more suitable, since the bleeding was not associated with the EVAR, embolization was not plausible due to the multiple hemorrhagic foci and surgical intervention would be meaningful only if the lesions were excisable (18). The patient was admitted to the Internal Medicine ward as a tumour of left psoas with osseous metastases. He was transfused with packed red blood cells and fresh frozen plasma and treated with broad spectrum antibiotics, while blood cultures were pending, initially with pipera - cilin/tazobactam and vancomycin and afterwards with meropenem and vancomycin as fever and high inflammation markers persisted. Without an apparent infectious cause, fever was attributed to the hematoma with a possible superinfection or to a paraneoplastic syndrome. In addition, opioid analgesics were used for pain control.

The differential diagnosis included sarcoma, melanoma, gastrointestinal (GI) cancer and prostate cancer. A CT scan of the head, neck and thorax and endoscopies of the GI tract were arranged and a CT-guided biopsy of the osteolytic lesion of the head of the left femur was performed. Fundoscopy was unremarkable for melanoma findings.

On the fifth day of his hospitalization, in anticipation of the biopsy result, the patient presented hemoptysis and CT pulmonary angiography was performed on an emergency basis, which was negative for pulmonary embolism but showed bilateral multiple dense nodular lesions with halo sign (probable hemorrhagic metastases) and intraalveolar bleeding (Figure 2). At that point, the patient started having an altered mental status, with progressively increasing oxygen needs. Pending the pathological diagnosis and with active bleeding from multiple sites, including the lungs that resulted in threatened airway, several discussions were made with the patient's family regarding the treatment plan. Intubation was excluded as an option due to the localization of the bleeding in the small airways and the poor prognosis of a metastatic tumor. The patient gradually developed increasing respiratory distress with increasing demands in oxygen supplementation. He was subsequently put on palliative morphine infusion. He succumbed after. The biopsy result, which came after the patient's death, was positive for a well-differentiated angiosarcoma. ❑

DISCUSSION

Aneurysms have been related to angiosarcomas in two ways according to the literature. On one hand, grafts used for the surgical treatment of aneurysms have been associated with the development of angiosarcoma on the site of the graft 6-120 months (mean 68 months) after graft placement (4). On the other hand, there have been cases of angiosarcomas mimicking ane-

urysms, meaning that aneurysm may have been secondary to angiosarcoma of the dilated vessel (14-17). In our case, no radiological abnormalities of the aorta were described that raise suspicions about the pathology of the aorta and no further exams that involved the aorta were conducted. However, the incidence of angiosarcoma even in the presence of an aneurysm is extremely low and, given the rarity of this entity, it is not advisable to incorporate pathology testing of the aneurysms into daily medical practice.

Retrospective examination of the pre-operative (EVAR) CT scan revealed a small osseous lesion at the left iliac bone and a small asymmetrical thickness of left psoas, while in the one-month follow up postoperative CT scan at least three such lesions could be identified, all having gone unnoticed. In the absence of typical symptoms, alternative pathologies should be sought to explain the clinical condition of patients with incidentally diagnosed abdominal aortic aneurysms. ❑

CONCLUSION

Angiosarcomas are rare but very aggressive malignancies. Due to their positions in body, they may mimic other clinical entities. Here we present a case of angiosarcoma with invasion of psoas, pelvic bones, femur and lung which was firstly manifested with low back pain, was confused by an incidental detection of an infrarenal aneurysm of abdominal aorta, which was treated with ELVAR and later with a hematoma of psoas and lung hemorrhage. ❑

Conflicts of interest: none declared.

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Ethical approval: The publication of the case report was approved by the Ethics Committee of the Hospital.

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