

Case Report

Mesenchymal Chondrosarcoma in Deep Femoral Vein: Case Report and Review of the Literature

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Abstract

The mesenchymal chondrosarcoma is a rare malignant tumor composed of small atypical undifferentiated cells and mature cartilage matrix islands, typically presenting with a bimorph appearance. Its origin in large vessels is extremely rare, with only four cases reported in literature. We report a case of a patient presenting with swelling in the right leg, diagnosed and treated as DVT. After unsuccessful treatment, with worsening of the symptoms, an abdominal CT scan was performed, showing solid formation, well-defined, located in right inguinal region. In the surgical exploration, it was held venotomy of the common femoral vein, followed by tumor thrombectomy and resection of the deep femoral vein. Histopathological analysis and immunohistochemical study were compatible with extraskeletal mesenchymal chondrosarcoma. Staging tests showed pulmonary and bone lesions. Systemic treatment was not performed. The patient is stable, with improvement of the lower limb edema.

ABBREVIATIONS

DVT: Deep Venous Thrombosis; MCS: Mesenchymal Chondrosarcoma

INTRODUCTION

The mesenchymal chondrosarcoma is a rare malignant tumor composed of small atypical undifferentiated cells and mature cartilage matrix islands, typically presenting with a bimorph appearance. It was first described in 1959 by Lichtenstein and Bernstein and occurs mainly in the bones. In 1964, Dowling described the extra skeletal location of the MCS [1]. Today, we know that about one fifth to one third of the cases are located in somatic soft tissues [2,3] and then called extra skeletal mesenchymal chondrosarcoma. Its origin in large vessels is extremely rare, with only four cases reported in the literature [4-7]. Of these, only one is located in femoral vein [4]. We report the second case of femoral vein mesenchymal chondrosarcoma discussing the diagnosis and treatment aspects.

CASE PRESENTATION

Female patient, 74 years old, with multiple comorbidities (hypertension, dyslipidemia, depression, arthrosis, thyroid disorders), with swelling in the right leg for seven months. Physical examination revealed edema in the right lower limb and nodulation in the right inguinal region. The limb's ultrasonography revealed venous thrombosis in femoropopliteal territory and nodule in the inguinal region compatible with lymph node, measuring about 2.5 cm. Anticoagulation was initiated with rivaroxabana. Thorax radiograph and total abdominal ultrasound without alterations. During the surgery it was noticed an expansive lesion, hard inside the right common femoral vein measuring about 3 cm in the largest diameter, being mooted the organized thrombus hypothesis. It was resected iliac lymph node that was increased in size, but with fibro elastic consistence. The lymph node anatopathology study has identified reactive lymphoid hyperplasia. The immunohistochemical analysis defined benign reactive process. The conduct with anticoagulant

treatment was maintained. Patient returned eight months after surgery, still in use of rivaroxaban, with worsening of edema in the right leg. Physical examination revealed right inguinal region without lymph node enlargement. Common femoral vein ultrasonography was performed showing a solid node with a calcification inside measuring 4.0 x 2.5cm. Thrombosis wasn't observed. It was performed abdominal CT scan, which showed solid formation, well-defined, located in right inguinal region, measuring about 4.0 x 2.7 cm. An angioresonance was indicated but the patient refused the procedure.

The surgical exploration of the right inguinal region was performed. It was observed during the surgery an increased volume in right common femoral vein and right deep femoral vein. Held venotomy the common femoral vein, followed by tumor thrombectomy and resection of the deep femoral vein (Figure 1). The injury arose from the deep femoral vein and protruded into the common femoral vein without invasion of the wall.

Histopathological analysis of the specimen revealed biphasic neoplasia, one of the patterns represented by small cells of round to oval hyper chromatic cores and scarce cytoplasm, among vascular channels with hemangiopericytoma pattern (Figure2). This pattern shows an abrupt transition with well differentiated cartilage tissue islands (Figure 3), with foci of calcification. The neoplasia had up to 03 mitosis / 10 high-power fields, areas of necrosis weren't observed. Immunohistochemical study revealed positive for CD99 in small neoplastic cells component (Figure 4) and S-100 protein in cartilaginous component (Figure5) and negative for EMA, CD34 and cytokeratin 7.

Staging tests revealed nonspecific pulmonary micro nodules and sclerotic bone lesions involving the vertebral bodies and the left pedicle of T8, T9, T10 and T11 upper plateau. The patient 8 months after surgery shows stable clinical condition. The lower limb edema disappeared with physical therapy. It was decided not to perform the systemic treatment. Femoral ultrasonography and magnetic resonance of spine and chest showed no alterations. The lung and spine lesions are stable.

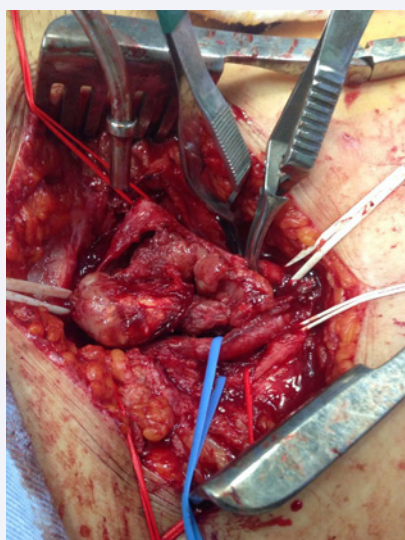


Figure 1 Intraoperative aspect of the tumor resection inside the femoral vein.

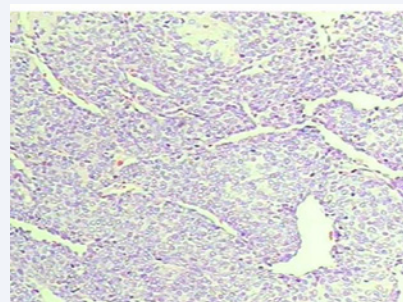


Figure 2 Histopathological analysis - small cells of round to oval hyperchromatic cores and scarce cytoplasm, among vascular channels with hemangiopericytoma pattern.

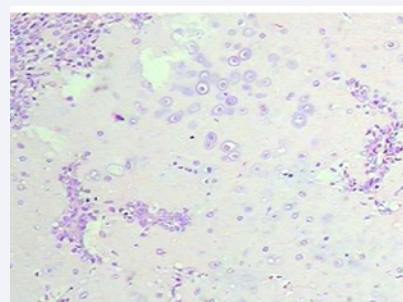


Figure 3 Histopathological analysis - abrupt transition with well differentiated cartilage tissue islands, with foci of calcification.

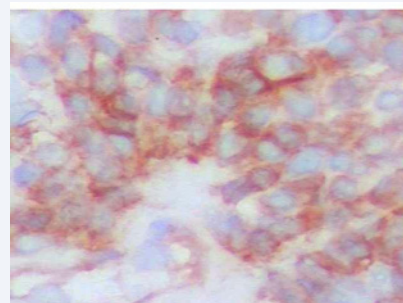


Figure 4 Immunohistochemical study - CD99 (+) in small neoplastic cells components.

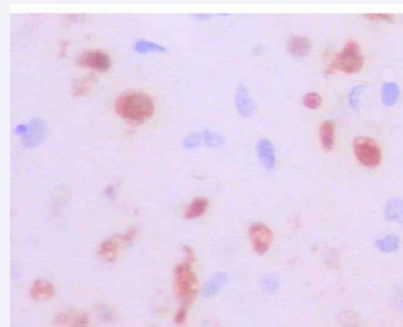


Figure 5 Immunohistochemical study - S-100 protein (+) in cartilaginous component.

DISCUSSION

Sarcomas are malignant tumors relatively rare originated from mesenchymal tissue that includes muscle, fat, bone, blood vessels and fibrous tissue or another supporting tissue. Encompass a wide variety of histological types and often involve members (55% of cases), especially the lower ones [8]. The mesenchymal chondrosarcoma, a chondrosarcoma subtype, is a rare neoplasia of small cells being derived from primitive mesenchymal tissue with potential chondroblasts. It presents most commonly as a primary neoplasia of the bone and might occur in extra-bone sites such as kidney, thyroid, prostate, mediastinum and meninges [2, 9,10]. The extra skeletal mesenchymal chondrosarcoma of intravascular location are even rarer. In PUBMED base, we find only four cases of intravascular mesenchymal chondrosarcoma, one originating from the femoral vein, one in the pulmonary vein, one in the vena cava and the other in the iliac vein [4-6]. Soft tissue sarcomas arising in deep veins of the extremities are unusual. While some cases of synovial sarcoma and leiomyosarcoma in femoral vein have been reported, there are only two reports of chondrosarcoma in this topography, one of them being the mesenchymal type [4,11]. The present case is the second report in this location.

The symptoms are characterized by pain and / or local edema [12,13]. Sarcomas located in the hip or thigh region are associated with a particularly high risk of thromboembolism, being deep vein thrombosis a common event in patients affected by sarcomas of the lower extremities [8].

On imaging studies, the conventional radiography shows calcified tumors in bizarre patterns. Computed tomography shows many intratumoral calcifications, as lobulation in some tumors. The well-defined granular calcification seen in CT and in the conventional radiography is a pattern of mineralization considered diagnostic for this type of tumor [14].

Histologically, the mesenchymal chondrosarcoma consists of solid areas of primitive mesenchymal cells with rounded or fusiform shape, with prominent vascular hemangiopericytoma-like and cartilage islands [9]. Foci of calcification and mineralization can be viewed amid small mesenchymal hyperchromatic cells [15]. On immunohistochemistry, the lesion is positive for vimentin, CD99 and S-100 in the areas of cartilage can be positive for enolase neuro specific or leu-7, but negative for desmin, cytokeratin markers or other epithelial markers [2].

The treatment of choice for mesenchymal chondrosarcoma consists of radical surgery with wide margin resection [4,16,17]. The incidence of local recurrence was significantly higher in patients undergoing surgery with positive margins, suggesting that resection with clear margins is an important objective in the treatment of patients with located disease [17]. The role of chemotherapy and radiation therapy remains controversial [4]. A recent study [17] showed that administration of chemotherapy can be associated with significant reduction in the risk of recurrence and death in patients with localized disease, being recommended in these cases free resection margins followed by adjuvant chemotherapy. However, other authors are skeptics of the claim that only chemotherapy and radical surgery can improve the survival [18].

The prognosis of this cancer in general is poor. The survival rate in 5 and 10 years is 54.6% and 27.3%, respectively [18]. The evolution in patients with metastasis to the initial presentation remains bad and despite the prolonged survival, many will progress to death [17].

In this report the patient presented initially as a thrombosis case in deep vein from the right leg, which delayed the correct diagnosis in 8 months. Initial imaging studies were inconclusive and in the first surgical approach because of lymphadenopathy, the surgeon was pleased with the diagnostic hypothesis of an organized thrombus in the vein, as this is the most frequent diagnosis. However, the patient has not evolved with improvement after anticoagulation and physiotherapy, which raised new evaluation. This time, the tests pointed to an intravascular injury. In the second surgical approach, it was confirmed the diagnosis of extra skeletal mesenchymal chondrosarcoma of intravascular location.

CONCLUSION

The intravascular mesenchymal chondrosarcoma is extremely rare. Nevertheless, it must be considered as a diagnosis in patients with DVT symptoms. Imaging studies play an important role in differential diagnosis of intravascular lesions in these patients and may contribute to an early approach. The treatment is based on resection with clear margins. Adjuvant treatment is still controversial. The prognosis is poor, with high rates of recurrence and metastasis.

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