Sarcomatous Degeneration of an Arteriovenous Malformation

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Abstract

Introduction: The prevalence of vascular malformations is estimated at 1.5% in the general population. Sarcomas are a rare group of mesenchymal-origin malignant tumors that accounts for less than 1% of all tumors in adults. We present one case of sarcomatous degeneration in an arteriovenous malformation of 50 years’ evolution.

Material: A 60-year-old male patient was admitted to our department presenting with soft tissue infection and hindfoot bleeding in the context of an arteriovenous malformation of 50 years' evolution and unidentified etiology. The patient had bilateral distal pulses. A 5 x 6 cm ulcerated and over-infected hindfoot lesion, with mamelonated borders that collapsed on digital pressure, was observed at the right lower limb.

Methods: Diagnostic arteriography revealed a vascular formation with angiomatous nidus on the medial surface of the talotibiofibular joint, arising from the posterior tibial artery. The arteriovenous malformation was embolized, with total exclusion thereof at final control. In spite of medical treatment and dressing, bad evolution persisted, with over-infection and appearance of new mamelonated formations, and infracondylar amputation was therefore decided. Results: Anatomic pathology revealed a malignant mesenchymal neoplasm suggestive undifferentiated pleomorphic sarcoma with giant cells (malignant fibrous histiocytoma of the giant cell type). The patient passed away at 3 months of diagnosis due to tumor disease dissemination.

Conclusion: Sarcomatous degeneration is a rare complication of arteriovenous malformations and, since clinical suspicion is low, the diagnosis is delayed, which entails poor results on the short-term.

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INTRODUCTION

The prevalence of vascular malformations is estimated at 1.5% in the general population. Abnormal communication between the arterial and the venous systems are characteristic of arteriovenous malformations (AVM). Sarcomas are a rare group of mesenchymal-origin malignant tumors that account for less than 1% of all tumors in adults. We present a case of sarcomatous degeneration in an arteriovenous malformation of 50 years’ evolution.

MATERIAL

A 69-year-old male patient was admitted to the Department of Angiology and Vascular Surgery with complaints of soft tissue infection and hindfoot bleeding in the context of an AVM of 50 years’ evolution and unknown etiology. The patient was a former smoker and had a personal history of high blood pressure and urothelial bladder cancer treated with transurethral resection.

On examination he had pedal and posterior tibial pulse on both lower limbs. A 5 x 6 cm ulcerated and over-infected hindfoot lesion, with mamelonated borders that collapsed on digital pressure and with sporadic capillary bleeding, was observed at the right lower limb (Fig. 1A).

METHODS

The patient was treated with intravenous antibiotic therapy according to the antibiogram and local dressing. A diagnostic arteriography revealed an increase in blood flow velocity in the right lower limb arterial system with regard to the contralateral limb, and a vascular formation with angiomatous nidus was observed on the medial surface of the talotibiofibular joint with main vascular efferent tributary of the posterior tibial artery.

Under rachianesthesia and 6F right femoral access, the posterior tibial artery was canalized. Intraoperative arteriography revealed the presence of an AVM dependent on the posterior tibial artery (Fig. 2A). The arteriovenous malformation was embolized with Glubran 2 acrylic glue (GEM, Viareggio, Italy), with AVM total exclusion at final control (Fig. 2B). Subsequently, surgical debridement of the ulcer was performed, with a sample of the borders being taken for histopathology examination. After surgical intervention, the lesion was improved and the patient was therefore discharged. At one month of the intervention, the patient was readmitted with the same clinical picture. A second debridement of the wound was performed, with complete excision thereof. In spite of antibiotic treatment and local compressive dressings, the surgical bed showed poor evolution, with infection persistence and appearance of new mamelonated structures, and intracondylar amputation was therefore decided.

RESULTS

Histopathology examination revealed an infiltrating malignant mesenchymal neoplasm ulcerating the epidermis, extensively necrotizing and reaching subcutaneous and striate skeletal muscle planes. Tumor cells expressed vimentin and, locally, soft muscle expressed actin; giant cells expressed CD68, and a proliferative index of 40% (Ki-67) was observed. A histopathological diagnosis of malignant mesenchymal neoplasm suggestive of undifferentiated pleomorphic sarcoma with giant cells (malignant fibrous histiocytoma of the giant cell type) was established.

The patient was referred to the oncology department, and two months after histopathological diagnosis, he was admitted to the internal medicine department presenting with symptoms of progressive dyspnea, which had started two weeks prior. Chest X-ray revealed multiple masses with a “cannonball” pattern. These findings were corroborated by a chest CAT scan showing pulmonary metastases (Fig. 1B). The patient died at 3 months of diagnosis due to tumor disease progression.

DISCUSSION

After gastrointestinal stromal tumor (GIST), most common mesenchymal tumors in adults are undifferentiated sarco-
mas, such as the one we described, followed by liposarcoma and leiomyosarcoma.

AVMs constitute a therapeutic challenge owing to their impact on the cardiovascular system and subsequent hemodynamic alterations. AVM clinical manifestations are dependent on their location and may cause congestive heart failure, venous hypertension and venous or arterial insufficiency and, as a consequence of the latter two, skin ulceration and even gangrene. Development of symptoms is caused by increased shunting that results in arterial steal and venous hypertension, both of which reduce tissue perfusion, which leads to pain, ulceration and bleeding. There are four indications for surgical treatment: bleeding, venous hypertension-derived complications, lesion located in an anatomical region that threatens patient's life (e.g., close to the airway), or that affects vital functions (e.g., hearing, sight). Pain, functional impairment and recurrent infection, among others, are included among relative indications. AVM treatment is intended to produce the closure of all arteriovenous communications by means of conventional surgery or endovascular techniques. Currently, transarterial embolization is the first therapeutic choice.

Mesenchymal tumors occur as painless, slowly growing tumors; when the tumor reaches an important size, pain or compression-associated symptoms, such as edema or paresthesias on the limb can appear. Forty-six percent of sarcomas are anatomically found on the lower limbs (buttocks, groin and lower limb). Although their etiology is unknown, there are predisposing factors for the occurrence of sarcomas, such as genetics: Li Fraumeni, type I neurofibromatosis, treatment with radio- or chemotherapy, chronic lymphedema (post-surgical, filariasis, etc.) and chronic irritation. It is possible that in the presented case the persistence of the arteriovenous fistula for 50 years may have played a role in the development of sarcoma.

CONCLUSION

Sarcomatous degeneration is a rare complication of arteriovenous malformations, and given that clinical suspicion is low, the diagnosis is often delayed, which entails poor results on the short-term.

REFERENCES