Surgical Treatment of Lower Limb Ischemia and Abdominal Aorta Aneurysm in a Patient Suffering from Eosinophilic Fasciitis

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SUMMARY We report on a 65-year-old woman with typical clinicopathologic features of eosinophilic fasciitis (Shulman syndrome) who suffered from lower limb ischemia and abdominal aorta aneurysm. Physical examination, laboratory data and histopathologic findings confirmed the diagnosis of eosinophilic fasciitis. The patient underwent aorto-bifemoral vascular prosthesis implantation. Tissue reactions associated with eosinophilic fasciitis produced serious disturbances of the implanted graft healing.

KEY WORDS: abdominal aorta aneurysm, atherosclerosis, eosinophilic fasciitis, lower limb ischemia, surgical treatment

INTRODUCTION

Eosinophilic fasciitis, first described by Shulman in 1974, is a rare disease more often demonstrated among middle-age male patients (1). It is considered to be a unique clinical syndrome or a variant of scleroderma (1-3). Eosinophilic fasciitis is characterized by acute or subacute development of induration of the skin and subcutaneous tissues of forearms, flank and thighs. The hands, feet and face are usually spared. The Raynaud phenomenon is usually absent. An inflammatory infiltration is present, extending into the fascia and skeletal muscles and mainly consisting of eosinophils (2-4). Autoimmune anemia, eosinophilia, hypergammaglobulinemia and elevated erythrocyte sedimentation rate occur variably (1-4).

This paper presents a very unique case of coexistence of eosinophilic fasciitis with lower limb atherosclerosis and abdominal aorta aneurysm, as well as the therapeutic problems of surgical treatment related to tissue reactions typical to Shulman syndrome.

CASE REPORT

A 65-year-old woman was admitted to Department of Vascular, General and Transplantation Surgery, Wroclaw Medical University, with chronic ischemia of the left lower limb and abdominal aorta aneurysm. The patient noticed first symptoms of ischemia one year before (pain-free walking distance 50 m). Two months before admission,
exacerbation of the disease occurred (10-m pain-free walking distance and presence of ischemic rest pain). Physical examination on admission revealed absence of pulse in the left inguinal region and presence of bruit in the right inguinal region. Duplex Doppler ultrasound disclosed left femoral artery occlusion, abdominal aorta aneurysm (42 mm in diameter) and right iliac artery aneurysm (28 mm in diameter). Abdominal computed tomography (CT) with contrast medium and arteriography (Fig. 1) confirmed the diagnosis. Results of accessory investigations and intensification of ailments indicated the need of vascular surgery.

Because of the diagnosis of eosinophilic fasciitis, oral corticosteroid therapy (Encorton 40 mg/day) was initiated 5 weeks before surgery and was administered for 3 weeks. The initial symptoms of the disease including walking difficulties, joint stiffness (ankle, knee and wrist), myalgia and pruritus appeared one year before and intensified gradually. After 4 months, the patient developed swelling of her feet that eventually involved legs up to the knees, as well as the hand and forearms. Finally, induration of the skin and subcutaneous tissues appeared at the site of edema. Physical examination on admission showed sclerotic and atrophic skin lesions of the extremities and coexistent cutaneous hyperpigmentation. Analysis of a full-thickness biopsy sample of the skin, fascia and muscle from the patient’s left calf revealed an inflammatory process characterized by foci of polymorphic infiltrations composed mainly of eosinophils in the dermis (especially in perivascular location) with mainly eosinophilic inflammatory infiltration of the fascia (Figs. 2 and 3). Laboratory blood tests revealed an elevated erythrocyte sedimentation rate (60 mm/h) as well as total eosinophil count (6128 cells/mm) and hypergamma-globulinemia (325.1%).

The patient underwent aortobifemoral dacron double-velour vascular prosthesis implantation (diameter 16/8 mm). During 2 months after primary surgery, the patient was operated on 4 times for rupture of distal anastomoses in both inguinal regions. Urgent procedures were performed in order to save the patient’s life and the limb. Bacteriological studies and scintigraphy with the use of technetium-labeled leukocytes carried out prior to each procedure revealed no graft infection. As the wall of the right femoral artery was destroyed, it was decided to perform extra-anatomic bypass implantation. The prosthesis made of polytetrafluoroethylene (8 mm in diameter) was implanted to the femoral artery in the adductor canal (via obturator foramen). After the operations no blood flow disturbances in the right lower extremity were
observed. In this situation, critical ischemia of the left lower extremity offered no opportunity to perform successful revascularization and the above the knee amputation was carried out. In the postoperative follow up, serious problems with wound healing were observed, as it was impaired in inguinal regions as well as in the amputation stump. There was no primary intention for healing observed in any locality.

DISCUSSION

Eosinophilic fasciitis can precede or be concomitant to hematologic disorders such as Hodgkin’s disease, peripheral T-cell lymphoma, myelomonocytic leukemia or aplastic anemia in 10% of cases (2). Coexistence with mycosis fungoides or sarcoidosis has also been reported (5). Little is known about the pathogenesis of eosinophilic fasciitis. It has been described in few cases in association with *Borrelia burgdorferi* infection. The role of immune complexes that cause eosinophil migration has been considered. In the literature we found no cases of atherosclerosis and abdominal aorta aneurysm requiring surgical intervention in a patient with eosinophilic fasciitis. However, situations have been described in which the lower limb pain associated with eosinophilic fasciitis was suspected to be an ischemic leg pain (6). In case of our patient the ailments associated with lower extremities were the result of both eosinophilic fasciitis and atherosclerosis. Because of this coexistence the definitive diagnosis was delayed. Vasculitis has been occasionally described in the course of eosinophilic fasciitis (7). In our opinion, the problems of graft healing after primary surgery were associated with arteritis that occurred in the course of disease and was confirmed on histopathology. Such inflammatory reactions are sometimes the reasons of ischemic ulcerations or necrotic lesions of the skin (8,9). We found no connection of the impaired process of graft healing and postoperative wound healing with the administration of steroids because the therapy lasted for less than 3 weeks and was discontinued 2 weeks prior to surgery. This short period of low-dose steroid therapy should not have influenced the tissue healing process. Due to the problems that occurred in the postoperative follow up we did not administer immunosuppressive regimens, systemic glucocorticoids or methotrexate. The use of these drugs usually results in recovery from eosinophilic fasciitis (10,11).

References