

Multiple venous aneurysms in a patient with hypereosinophilic syndrome

WSJ Malskat¹, E Racz¹, S ten Raaij², PJ Lugtenburg³,
V Noordhoek Hegt⁴ and MGR de Maeseneer¹

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Abstract

Hypereosinophilic syndrome (HES) is a diverse group of rare disorders, defined by persistent peripheral blood eosinophilia (>1500 per mm^3), the absence of a primary cause of eosinophilia (such as parasitic or allergic disease), and evidence of eosinophil-mediated end-organ damage. Arterial aneurysms have been previously reported in these patients. This is the first report of a patient with HES and multiple venous aneurysms, causing recurrent pulmonary thromboembolism. Venous aneurysms can represent eosinophil-mediated, potentially fatal end-organ damage in patients with HES.

Keywords

Hypereosinophilic syndrome, venous aneurysms, chronic venous insufficiency

Introduction

Hypereosinophilic syndrome (HES) is a diverse group of rare disorders, defined by persistent peripheral blood eosinophilia (>1500 per mm^3), the absence of a primary cause of eosinophilia (such as parasitic or allergic disease), and evidence of eosinophil-mediated end-organ damage.¹ The symptoms of HES are variable, since it is associated with eosinophilic infiltration in multiple tissues and organs, for instance pulmonary, cardiac, and skin tissue. Different subtypes of HES can be identified based on etiology, such as myeloproliferative HES (chronic eosinophilic leukemia), lymphocytic HES (caused by secretion of eosinophilopoietic cytokines by T-lymphocytes), and idiopathic HES. Cardiovascular complications of HES are a major source of morbidity and mortality, which are reported in 40–50% of patients with HES, and include cardiac necrosis, thrombosis and fibrosis, and thromboembolic complications. A few case reports describe arterial aneurysms in patients with HES.^{2–4} We report the first case of HES with multiple venous aneurysms.

Case report

A 53-year-old male presented at our outpatient clinic with extensive visible varicose veins in both legs for the last 12 years. His medical history revealed HES diagnosed at the age of 19 years (presenting with abdominal cramps, diarrhea, and dyspnea) and two episodes of

pulmonary embolism (PE) at the age of 50 and 52. He was currently treated with prednisone 10 mg daily and long-term anticoagulation (acenocoumarol).

Duplex ultrasound investigation revealed a large sacular aneurysm of the right popliteal vein (Figure 1) and large aneurysms of the right and left great saphenous veins (GSVs) at the saphenofemoral junction (SFJ), respectively measuring 3.4×2.8 cm and 2.8×2.4 cm, with incompetence of both GSVs and tributaries. The diameter of the right GSV was 1.1 cm, with a 15 cm length of the refluxing segment from the groin. The left GSV had a diameter of 0.9 cm with reflux to below knee level. Additional 3D-CT-venography of the lower extremity (with insertion of the catheter in the right common femoral artery and scanning during the venous phase) confirmed the presence of a large popliteal vein aneurysm at the right leg

¹Department of Dermatology, Erasmus MC University Medical Centre, Rotterdam, The Netherlands

²Department of Vascular Surgery, Erasmus MC University Medical Centre, Rotterdam, The Netherlands

³Department of Hematology, Erasmus MC Cancer Institute, Rotterdam, The Netherlands

⁴Department of Pathology, Erasmus MC University Medical Centre, Rotterdam, The Netherlands

Corresponding author:

Marianne GR de Maeseneer, Department of Dermatology, Erasmus MC, Burgemeester's Jacobplein 51, 3015 CA, Rotterdam, The Netherlands.
Email: m.demaeseneer@erasmusmc.nl



Figure 1. Longitudinal ultrasound image of the right popliteal vein with evident blood stasis in the large saccular aneurysm.

and revealed large fusiform aneurysms of the right and left distal external iliac vein as well (Figure 2).

The large saccular aneurysm of the right popliteal vein was assumed to be the cause of recurrent PE in the past and was therefore treated by an aneurysmorrhaphy at the department of Vascular Surgery. Further treatment of superficial venous incompetence was scheduled at our Department of Dermatology. Radiofrequency ablation of the left GSV and phlebectomies were performed first, followed by high ligation with short stripping and phlebectomies of the right GSV and tributaries two weeks later. For both interventions local tumescent anesthesia was applied. Since the aneurysm of the right GSV exceeded 3 cm, we decided to perform a classic operation at the right leg.

Histologic examination of the resected aneurysm of the right GSV showed extensive infiltration by eosinophilic granulocytes next to some lymphocytes and plasma cells in the media of the vein wall (Figure 3).

In view of these histologic findings, the prednisone dosage was increased to 20 mg daily, to reduce the disease activity of the HES. At six-month follow-up the patient did not experience any further problems. It was decided to leave the residual aneurysms at the distal external iliac vein untreated and to continue chronic

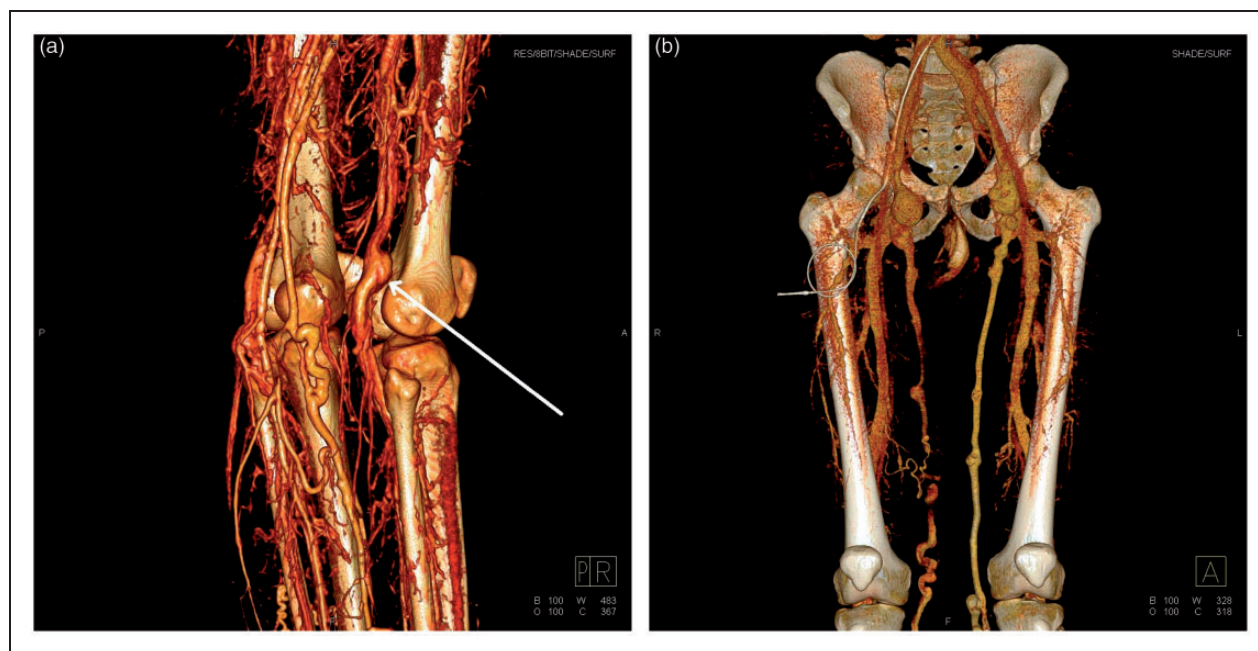


Figure 2. 3D-CT-venography of the lower extremity. (a) Posterolateral view from the right side (right leg is seen at the right side of the figure): presence of a large saccular aneurysm of the right popliteal vein with a diameter of 4 cm. (b) Two large fusiform aneurysms of the right and left distal external iliac veins, with diameter of 4.5 cm.

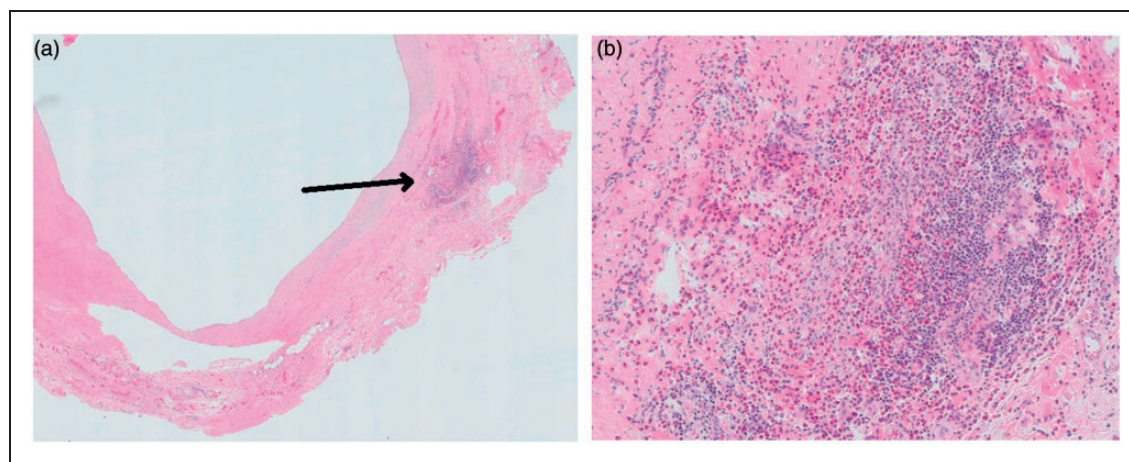


Figure 3. Histologic examination of the resected aneurysm of the right GSV. (a) Inflammatory infiltrate with eosinophil granulocytes of the wall of the resected aneurysm of the right GSV. (b) A close up of the eosinophilic infiltration.

anticoagulant treatment. Further monitoring of the disease activity is scheduled every six months by means of cardiac and pulmonary functional tests and eosinophilic blood count (target value <1500 eosinophils per mm^3).

Discussion

This patient presented with multiple venous aneurysms associated with HES. Vascular involvement of HES is very rare.

In the literature, several cases of coronary artery aneurysms in HES patients have been reported, as well as other arterial aneurysms.^{2–4} The arterial aneurysms revealed extensive infiltration by eosinophils, as described by Sekino et al.,⁴ similar to what we found in the walls of the venous aneurysm and dilated GSV trunk in our patient. This infiltration with eosinophils in the vascular wall causes damage and weakening of the wall as a result of degranulation and release of cytokines and free radicals, similar to the pathophysiology underlying other HES-associated end-organ damage. Therefore, venous aneurysms can also be considered a sign of end-organ damage in HES, though very exceptional.

Thromboembolic complications occur in about 25% of patients with HES and 5–10% of HES patients die from these complications. Currently, it is believed that most thromboembolic complications in HES consist of intracardiac thrombus and secondary pulmonary and arterial emboli. However, venous thrombus formation, in particular in a large saccular venous aneurysm,⁵ may cause (potentially fatal) venous thromboembolism in these patients as well. The co-occurrence of HES and

venous aneurysms might be underestimated in these patients.

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Conflict of interest

None declared.

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