ESC European Society of Cardiology

European Heart Journal - Case Reports (2022) **6**, 1–3
https://doi.org/10.1093/ehjcr/ytac314

CASE REPORT
Cardiac imaging

Infected thoracic aortic graft in a woman with Darier disease: a case report

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Received 23 December 2021; first decision 18 January 2022; accepted 26 July 2022; online publish-ahead-of-print 29 July 2022

Background Patients with Darier disease often present with staphylococcal skin infections and are at risk for complications when they undergo cardiothoracic surgery, such as acute aortic dissection repair.

Case summary A 39-year-old woman with hypertension and Darier disease suffered an acute type A aortic dissection, requiring emergency operation with a Dacron graft. Twenty-five days post-operatively, she developed pneumonia and staph hominis was isolated in blood cultures and Bronchoalveolar Lavage. Following completion of antibiotics, multiple relapses occurred during a 6-month period, each time treated with appropriate antibiotic therapy. An 18F-fluorodeoxyglucose positron emission tomography computerized tomography showed persistent graft uptake and re-operation was performed. At 22 months of follow-up, the patient remains asymptomatic and the 18F-FDG PET/CT shows significant reduction in FDG uptake.

Discussion Graft infection is a rare but serious complication. Antibiotic therapy is often inadequate and re-operation is needed. As staphylococcal skin infections often occur in patients with Darier disease, adequate preprocedural skin preparation and sterilization are very important in these patients.

Keywords Darier disease • Staphylococcus hominis • Graft infection • 18F-FDG PET/CT • Case report

ESC Curriculum 2.1 Imaging modalities • 2.5 Nuclear techniques • 7.5 Cardiac surgery • 9.1 Aortic disease

Learning points

- Graft infection must be suspected when multiple episodes of fever occur post-operatively, despite appropriate antibiotic therapy.
- 18F-fluorodeoxyglucose positron emission tomography computerized tomography is the imaging modality of choice for diagnosis and follow-up.

Introduction

Darier disease[1] is a rare cutaneous disease with autosomal dominant inheritance. Patients with Darier disease often present with staphylococcal skin infections.[2] Longstanding hypertension predisposes to acute aortic dissection. Repair of the aorta for acute dissection is associated with significant morbidity and mortality as well as remote complications, including true or false aneurysm formation, graft infection, and recurrence of dissection.

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Handling Editor: Parham Eshtehardi
Compliance Editor: Megha Agarwal
Supplementary Material Editor: Jonathan Senior

Timeline

Presentation	39-year-old woman with hypertension and Darier disease
	Acute type A aortic dissection
Index procedure	Aortic repair with dacron graft
Post-operative infection	25 days post-op fever
Investigations	Blood cultures and BAL positive for staph hominis
Management	Ciprofloxacin for 4 weeks
Recurrence of infection	Recurrence of fever a week later, ciprofloxacin plus vancomycin for 4 weeks
Investigations	18F-FDG PET/CT shows graft uptake
Further relapses	Multiple febrile episodes, each time, after completion of daptomycin
Re-operation	Bovine graft 9 months after the 1st operation and 6 weeks of daptomycin post-op
Follow-up	22 months after the 2nd operation the patient is asymptomatic and the FDG uptake significantly reduced.

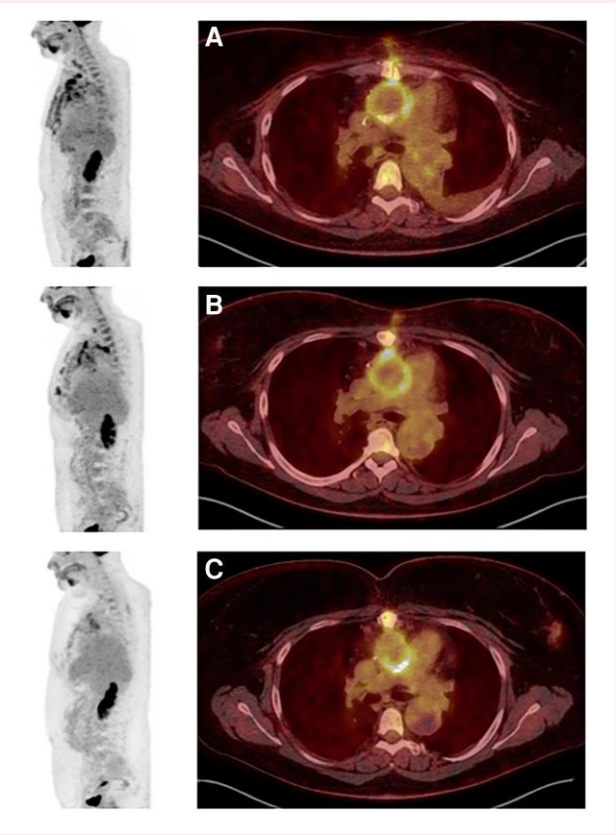

Figure 1 Full body 18F FDG PET/CT scan (left) and transverse section at the level of pulmonary artery bifurcation (right), after initial antibiotic therapy (*A*), after multiple courses of antibiotics (*B*), and at 22 months post aortic graft re-operation (*C*), showing persistent hypermetabolic activity in (*A*) and (*B*), which is largely improved at (*C*).

Case presentation

Our patient is a 39-year-old Caucasian woman, with family history of hypertension and with hypertension herself since her late 20s, well controlled on amlodipine 10 mg once daily. In addition, her non-cardiac history includes keratosis follicularis (Darier disease), with frequent skin infections, treated empirically with minocycline. She suffered an acute aortic dissection type A 3.5 years ago and underwent surgery for aorta repair with a 34 mm Dacron graft. Her immediate post-operative course was uncomplicated and she was discharged a week later.

Twenty-five days after surgery she presented to our institution with fever; clinical examination was remarkable for red keratotic papules of forehead, neck, and presternal area,[3] in addition, fine left lower lobe crackles were present. Her fundi were normal. Leucocytosis with granulocytosis was observed; renal and liver functions tests were within normal limits. A chest X-ray revealed left lower lobe pneumonia with ipsilateral pleural effusion and an echocardiogram showed moderate pericardial effusion. Her blood cultures and Bronchoalveolar Lavage were positive for staph hominis and ciprofloxacin 500 mg BID was administered for 4 weeks. One week after completion of antibiotic therapy, she was re-admitted with fever and malaise and had negative blood cultures, but PCR was positive for Coagulase Negative Staphylococci and ciprofloxacin 500 mg BID and vancomycin 1 g IV BID were given for 4 weeks. With the suspicion of graft infection,[4,5]—relapse of fever, despite previous appropriate antibiotic treatment, soon after urgent surgery, in a patient with skin disease—an 18F-fluorodeoxyglucose Positron Emission Tomography Computerized Tomography (FDG PET-CT scan)[6,7] was done and showed uptake in the aortic graft (*Figure 1A*). Daptomycin 500 mg IV OD was initiated for another 6 weeks and fever and inflammatory markers subsided. During a 6-month period, however, multiple relapses occurred, each time following completion of antibiotics and a subsequent FDG PET-CT scan showed increased metabolic activity in the graft, pleura, pericardium, sternum and substernal fat, lymph nodes and spleen (*Figure 1B*). The decision to re-operate was then made; the infected Dacron graft was removed and a bovine pericardium graft was inserted, 9 months after the first operation. Methicillin Resistant *Staphylococcus epidermidis* (MRSE) was isolated in graft's cultures and IV daptomycin was administered for 6 weeks post-operatively. Patient remained asymptomatic at 22 months follow-up after second surgery and 18F-FDG PET/CT scan (*Figure 1C*) showed significant reduction in FDG uptake.

Discussion

Bacterial, particularly staphylococcal, skin infections are common in patients with Darier disease. Therefore, if an operation or invasive procedure is indicated, especially on an emergent basis, adequate pre-procedural skin preparation and sterilization are very important. Patients with longstanding hypertension are prone to aortic aneurysm formation or acute dissection. When a graft is inserted, either electively or urgently, the risk of graft infection is not negligible. Clinicians should have, in patients with appropriate clinical presentation, a high suspicion of this complication and consider 18F-FDG PET/CT imaging, which is the most accurate method for diagnosis.

Lead author biography

Dr Sbarouni studied Medicine in Greece. She was trained in Cardiology in the UK. She is currently practicing Interventional Cardiology in Athens, Greece.

Supplementary material

Supplementary material is available at *European Heart Journal – Case Reports* online.

Slide sets: A fully edited slide set detailing these cases and suitable for local presentation is available online as Supplementary data.

Consent: The authors confirm that written consent for submission of the case report including images and associated text has been obtained from the patient in line with COPE guidance.

Conflict of interest: None declared.

Funding: The authors report no specific funding related to the article.

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