It Is Not Just Mouth Ulcers; Dilated Cardiomyopathy As a Rare Presenting Symptom For Behcit's Disease

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Type of submitter

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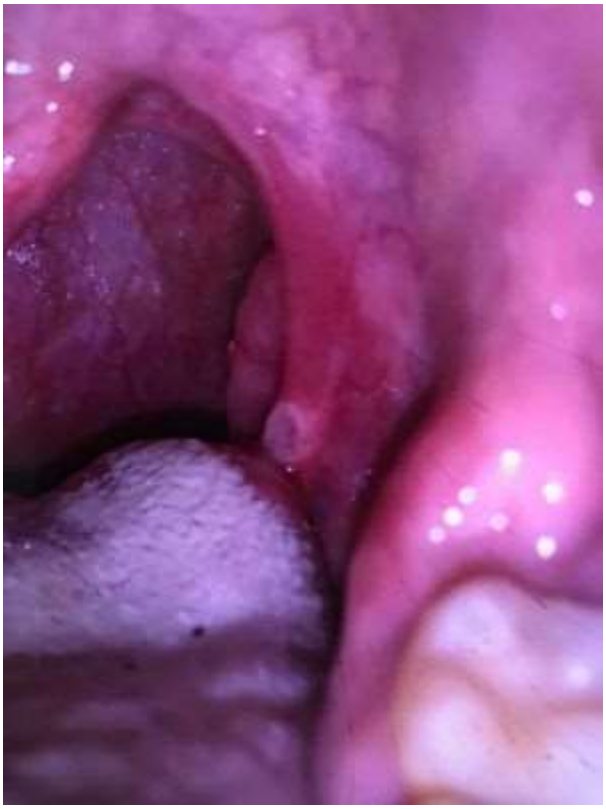
Abstract



Background:

Behcet's disease is a relatively rare entity, affecting 1 per 15,000 to 500,000 in developed countries. It is most commonly known to cause recurrent oral and genital ulcers alongside a wide variety of systemic

presentation, mostly related to generalized vasculitis. Here we present a case of orthopnea and intractable headache which was later attributed to severe heart failure with reduced ejection fraction as well as superior vena cava thrombosis as an initial presentation of Behcet's disease.

Case Presentation:

A 32-year-old gentleman with a past medical history of asthma presenting with generalized fatigue, orthopnea with intermittent fevers as well as recurrent sore throat over the course of 9 months that triggered multiple emergency room visits and for which he received the diagnosis of URTI. The patient, however, started to develop intractable headaches with facial and chest wall swelling for 1 week prior to his admission to our hospital. Patient's physical exam showed a positive Pemberton sign. He subsequently underwent CT angiogram that confirmed the presence of SVC thrombosis. The patient continued to have orthopnea despite initiation of heparin infusion and improvement of his SVC thrombosis symptoms for which he underwent transthoracic echocardiogram that revealed global hypokinesia with EF of 20-25%.

Upon further detailed history taking, the patient reported having mouth ulcers that used to erupt whenever he had a sore throat, he also reports unusual pimples in his legs that matched the description of pseudofolliculitis. He also reported a family history of Bechet disease in one of his distant family members. A diagnosis of Behcet was made and the patient was started on steroids.

The patient was also started on Sacubitril/Valsartan, Carvedilol, Aspirin, and Atorvastatin. He later underwent Myocardium Perfusion imaging that showed normal EF with no reversible ischemia. He eventually had a Cardiac MRI that showed Improvement of his EF to a low-normal with no evidence of myocardial scarring or fibrosis.

Discussion:

Behcet disease is a rare disease especially outside of the ancient Silkroad area. It is mostly diagnosed clinically, and that why a thorough history and physical is needed to uncover it. A diagnosis is made when a patient is found to have recurrent oral ulcers alongside two of the following; Recurrent genital aphthae, Eye lesions like anterior or posterior uveitis, Skin lesions like pseudofolliculitis, or a positive pathergy test

Cardiac involvement in Behcet's disease is rare but reported in the medical literature. The cardiac presentation can vary from inflammation of one or all of cardiac layers, endomyocardial fibrosis, coronary arteritis, intracardiac thrombus, conduction system disturbances, and valvular disease. Treatment of Bechet varies depending on the type of organs affected, ranging from topical steroids to Colchicine to systemic steroids and immunosuppressants. In our case patient was started early on steroids with a fast and rapid recovery of the myocardium.

Conclusion:

Recognizing the rare, but possibly grave, cardiac manifestation of Behcet's disease including dilated cardiomyopathy is essential as it might aid in making the diagnosis and avoid the burden of increased medical visits and over-testing.

Categories

Advanced Fellow: Case

Program/Institution Name

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Pause and Consider the Late Complications of Heart Transplantation

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Abstract

Introduction

Heart Transplantation is the gold standard treatment for patients with stage D heart failure. Survival after transplantation continues to improve; however, chronic complications impact outcomes and survival after transplantation.

Case

We present a 75-year-old female, 23 years status post orthotopic heart transplant presented with nausea, vomiting and an unexplained fall. She was followed routinely in the transplant clinic and maintained on Prednisone 2.5mg daily and Myfortic 720mg BID.

On hospital day two, she experienced sinus arrest lasting 14 seconds. A permanent pacemaker was emergently placed. An echocardiogram revealed normal graft function and workup for rejection was negative. She began complaining of worsening headaches, nausea, left arm numbness and right sided hearing loss. CT scan of her brain revealed a 2.7 cm ring enhancing lesion in the cerebellum; the mass was surgically resected. Pathology was consistent with a lambda restricted B-cell lymphoma favoring post-transplant lymphoproliferative disease (PTLD). Bone marrow biopsy showed no involvement and viral serologies including EBV were negative. A PET scan only localized disease to the cerebellum. Immunosuppression was reduced to Myfortic 360 mg BID and prednisone 5 mg. She was started a chemotherapy regimen of dexamethasone, rituximab, zidovudine and ganciclovir.

Discussion

PTLD is a heterogenous group of disorders presenting in immunosuppressed patients after organ transplantation. The most common sites of PTLD initial involvement were the gut, hilar lymph nodes and lung. The central nervous system (CNS), as in our patient, was a rare site of initial involvement. An analysis identified three cases of primary CNS-PTLD in 1674 heart and lung transplants with an incidence of 0.18% and all were driven by EBV. Our patient had no evidence of EBV in her tumor, bone marrow biopsy or serum.

The late onset nature of our patient's PTLD is unique. PTLD has a bimodal distribution with the majority of cases occurring in the first year after transplantation. The average time to diagnosis after heart transplantation has been reported as 5.5 years. Prior to this report, only two cases of PTLD were described twenty years post transplantation.

Beyond the characteristics of this patient's PTLD, the other novel aspect of this case is the bradycardia and asystole potentially of a neurologic etiology induced by the brain lesion. Although sympathetic reinnervation of the heart is accepted, parasympathetic reinnervation of the heart is controversial. Canine models have shown evidence of parasympathetic reinnervation as early as 6-12 months after transplantation. Human models, however, have not consistently shown parasympathetic control of donor hearts. .

Conclusion

Our patient presenting with an EBV negative primary CNS-PTLD 23 years after heart transplantation is a rare presentation of this immune system driven malignancy. Although conduction system delays in heart transplants are typically due to acute rejection, surgical anastomosis failure or chronic graft vasculopathy, there is data to suggest that reinnervation of the autonomic system the further a patient is from initial transplantation. We recommend pausing and considering neurally mediated etiologies, when evaluating asystole as a late complication of transplantation.

Categories

Advanced Fellow: Case

Program/Institution Name

Ohio State University Hospital

Reverse Takotsubo from Iatrogenic Stress Induced Cardiomyopathy

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Abstract

Introduction: Reverse Takotsubo is a version of stress cardiomyopathy in which the base and mid-ventricular segments of the left ventricle are akinetic. This is opposite of the classically described Takotsubo cardiomyopathy in which there is apical ballooning from systolic dysfunction of the apex.

Case: A 63 year old woman with no previous cardiac history presents after suffering a fracture. Her course was complicated by septic shock requiring hemodynamic support. Shortly after starting norepinephrine, she developed tachycardia, severe chest pain and dyspnea. An EKG showed atrial flutter with RVR thus she was started on IV Amiodarone. Troponin-T was elevated and EKG was remarkable for new T-wave inversions. During preparation for cardioversion, an error in the norepinephrine pump was noticed and thus stopped. The patient was successfully cardioverted into sinus rhythm. Her chest pain and dyspnea subsided. A prior echocardiogram showed normal wall motion in all territories and ejection fraction of 55%. The echo done during this episode demonstrated normal function of the apex but hypokinesis of the base. One week later, repeat echocardiogram was remarkable for complete resolution of the basal hypokinesis.

Discussion: Due to an error in pump setup, the patient received 10 mcg/kg/min (700 mcg/min) rather than 10 mcg/min of norepinephrine. This case brings up many important learning points including the management of iatrogenic stress induced cardiomyopathy. The patient received 70 times more norepinephrine than intended resulting in cardiac complications. These symptoms and laboratory findings were all a result of the iatrogenic stress induced cardiomyopathy. Stopping norepinephrine infusion was essential. The error was correctly recognized in a timely manner, preventing the need for unnecessary and potentially harmful procedures.

Conclusion: Reverse Takotubso is thought to result from the relative density of adrenergic receptors at the apex compared to the base of the heart. The reverse pattern typically causes more myocyte damage and higher troponins. Cardiac adrenergic receptors cause a negative inotropic effect when stimulated by excessive amounts of catecholamines.

Categories

Advanced Fellow: Case

Program/Institution Name

Ohio State University Hospital

Program/Institution Name If NOT listed in the prior question.

Electrophysiology Fellowship