Thrombotic Thrombocytopenic Purpura Associated with Staphylococcus Aureus Endocarditis in a Diabetic with Furunculosis

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Abstract

Thrombotic thrombocytopenic purpura(TTP) is an uncommon and potentially fatal syndrome which is due to either congenital or acquired deficiency of the ultra large von Willbrand's factor (ULVWF) cleavage protease. It is characterized by mechanical micro angiopathic hemolytic anemia, and organ dysfunctions.

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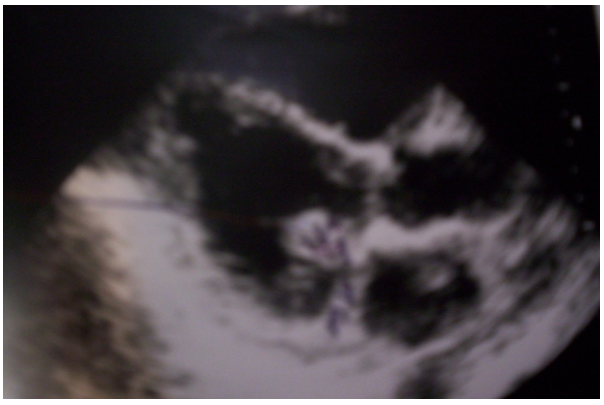
Case Report

A 48 yrs old man, with diabetes mellitus presented to our unit with a month history of intermittent high grade fever and excessive night sweat which started about a month prior to presentation. About the same time he developed pain in his back and the hip joints.

He also lost about 4 kg of his weight in 4 weeks. 4 days prior to presentation to the ER he developed abdominal pain which was associated with vomiting, diarrhea and jaundice. The stool had no blood nor mucous. Two days prior to presentation he became confused but there was no weakness of any of his limbs and he had no seizures. Apart from occasional bouts of cough he had no other cardiopulmonary symptoms. He had no history of any chronic illness except for the diabetes. And about three months to the start of his present illness he had furuncles that were treated with amoxicillin by his GP. He was on gliclazide at the time of presentation. The patient does not use illicit drugs but drinks alcohol in excess.

On examination he was acutely ill looking with a fever of 100.8 °F, he was pale and jaundiced with multiple splinter hemorrhages and minimal bipedal edema. His pulse rate was 118/min, BP=150/60mmHg, RR=24/min and oxygen saturation was 98% on room air. Systemic examination revealed a confused and restless man with flapping hand tremors. Murmurs of mitral and tricuspid valves regurgitation were heard in the precordium and he had hepatosplenomegaly. The planter reflex was extensor bilaterally and the fundoscopy was normal .

The WBC increased progressively during admission from 6.2k/ul to 20.7k/ul. And the hemoglobin dropped from 11 to 9.8 gm/dl .His platelet count was very low at 25k/ul at admission. The lactate dehydrogenase and urea were elevated at 890 U/l and 27 mg/dl respectively. The C-reactive protein was very high and the urinalysis revealed proteinuria, urobilinogen and microscope hematuria. The CXR showed an enlarged heart and the ECG showed sinus tachycardia. Echocardiography showed severe mitral and moderate tricuspid regurgitation with a very large vegetation on the anterior leaflet of the mitral valve (figure 1). Four bottles of blood culture grew staphylococcus aureus. The CT scan of his brain was normal. The patient was started on ceftriaxone, gentamycin and vancomycin for the bacteria endocarditis and the TTP like syndrome was treated with high dose methylprednisolone for 3 days. The patient improved clinically, he became well oriented and the flapping tremor disappeared in 2 days. The platelet count increased gradually to 330k/ul by day 5 and he was then started on low dose aspirin. However he continued to have low grade pyrexia and he developed biventricular failure. The heart failure was treated with frusemide ,digoxin, perindopril and spironolactone. He also developed arterial embolization to the left femoral artery which was demonstrated by ultrasound doppler. He was subsequently referred to a cardiothoracic centre in another hospital in the region where he had metallic mitral valve replacement and catheter embolectomy.



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Discussions

TTP is an uncommon and fatal multi systemic syndrome of uncertain origin that is characterized, by thrombocytopenia, fever, organ dysfunction (mostly involving the central nervous system) and evidence of microangiopathic hemolytic anemia (increase LDH, fragmented RBC, and urobilinogen)[1].

The pathogenesis of congenital and acquired TTP is said to involve deficiency of ULVWF cleaving protease ADAMTS-13, which lead to platelet aggregation and subsequent intravascular platelet thrombus formation. In acquired form of TTP due to infection as illustrated by the case been reported, the ULVWF protease, is inhibited by immune complexes and immunoglobulins produced during the infections[2].

The immune complexes and auto antibodies associated with bacterial endocarditis inhibits ULVWF cleaving protease as does the ADAMTS 13. This result in intra vascular platelet thrombus formation which lead to the microangiopathic hemolysis and infarction of the organs. The infarction in the brain and the kidneys result in neurological and renal dysfunction, but the former is more characteristic of TTP. Cardiac, cutaneous and gastrointestinal systems can also be involved[1-3].The cardiac involvement in TPP include, thrombotic endocarditis, myocardiac abscesses, pericarditis and cardiogenic shock. [1,4,5]. Some of these cardiac complications of TTP can be confused with bacteria endocarditis. The case presented have the features of both TTP and bacterial endocarditis. The thrombocytopenia, evidence of hemolysis, and neurological dysfunction in the presence of a normal brain CT strongly supported TTP. While the echocardiography and positive blood culture confirmed the bacterial endocarditis. This features makes it obvious that the TTP is a complication of the bacterial endocarditis in this case.

The treatment of TTP involves plasmapheresis and large volume plasma transfusion. While the treatment of underlying infection is said to be helpful in most cases of TTP associated with endocarditis. Steroid and anti platelet have also been shown to be useful. Vincristine and cyclophosphamide have been reported to increase the platelet count. It is pertinent to mention that platelet infusion can be dangerous and should not be used [3,4,7].

In conclusion we suggest that TTP should be sort for and treated in cases of bacteria endocarditis as the failure to recognized this fatal condition can impart negatively on the prognosis of an already potentially dangerous disease .

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