To the Editors:

A case of Behcet's disease complicated with intra cardiac thrombus and Budd Chiari syndrome

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Mr. C, a 21-year old male, who had painful oral and genital ulceration for two years, came to us with a left blind eye due to panuveitis, generalised body swelling and painful abdominal distention of 1 week duration. He was diagnosed to be having Behcet's disease with pathergy test confirmation. He was anicteric. His cardiovascular, respiratory and neurological examinations were normal. He had tender ascites and extensive scrotal ulceration. Ultrasound scan of the abdomen revealed an enlarged liver with coarse echopattern with reduced portal venous flow velocity (<10 ml/sec). Central hepatic veins showed no blood flow. A contrast CT abdomen revealed inferior vena caval thrombosis involving all three hepatic veins. The portal vein appeared normal and the inferior vena cava reformed just above the right renal vein. These features confirmed Budd Chiari syndrome. A 2D echo showed a 30 mm elongated right atrial thrombus attached to the inter atrial septum. His serum AST was 58 u/l, ALT was 44 u/l, INR was 1.83 and APTT was 30 sec. Anti-nuclear antibody test and VDRL test were negative with no evidence of protein C and S deficiency.

He was started on enoxaparin 40 mg and warfarin 3 mg daily aiming for an INR of 2-3. Oral prednisolone was started and tailed off with the introduction of azathioprine. A repeat 2D echo was performed after one month of treatment, and the right atrial thrombus had resolved.

Figure 1. Hepatic vein thrombosis and hepatomegaly.
Poncet’s disease or tuberculous rheumatism, first described by Poncet in 1897, is a rare form of polyarthritis resulting from visceral tuberculosis without any direct involvement of the joints [1].

A 16-year old girl had low to moderate grade fever and joint pain for 3 months. Her joints were swollen without signs of inflammation. This involved almost all joints of the body both small and large. At times the joint pain was severely incapacitating.

On examination, she was anicteric and had pallor. The liver was marginally enlarged. Her joints, particularly the knee and small joints of hand, were painful on palpation. Chest examination was normal except for occasional crepitations in the right infrascapular region.

Her investigations were as follows: haemoglobin – 8.7 g/dl, total leukocyte count – 7.7×10 g/l neutrophils – 52%, lymphocytes – 44%, platelet count – 255×10g/l, ESR – 80 mm in 1st hour, marginally raised liver enzymes and raised serum globulin 50g/l against a normal serum albumin 38g/l. The chest X-ray showed few suspicious shadows in the left apical area and abdominal ultrasonography showed marginally enlarged liver without intra abdominal lymphadenopathy. Routine urine examination, culture and serum creatinine values were normal.

Though the sputum for acid-fast bacilli was negative, sputum was sent for tuberculosis culture. Rheumatoid factor, serum uric acid, HIV serology and ASO were normal. The CRP was only marginally elevated (1.72). Mantoux test was highly reactive (18 mm) at 72 hours. Synovial fluid analysis from the left knee joint was normal. Considering the elevated ESR, positive Mantoux test raised serum globulins and ALP and suspicious infiltrates on chest X-ray with negative serology a presumptive diagnosis of tuberculous rheumatism was made. The patient was started on antitubercular therapy. The patient stayed in hospital for

Intracardiac thrombi usually involve the right side of the heart [2]. Its presence confirms a poor prognosis [3]. Anticoagulants and anti inflammatory drugs are effective in treating it [4].

References

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