

SUPERIOR VENA CAVA SYNDROME CAUSED BY BEHCET'S DISEASE

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Key-word: Behçet disease

Background: A-36-year-old man presented with a six-year history of Behçet's disease and superior vena cava (SVC) syndrome. He said his symptoms related with Behçet disease (recurrent genital and oral ulcers, joint tenderness) did start 20 years ago. He has dyspnea and generalized joint tenderness. Physical examination was normal except for generalized joint tenderness. Laboratory findings were unremarkable.

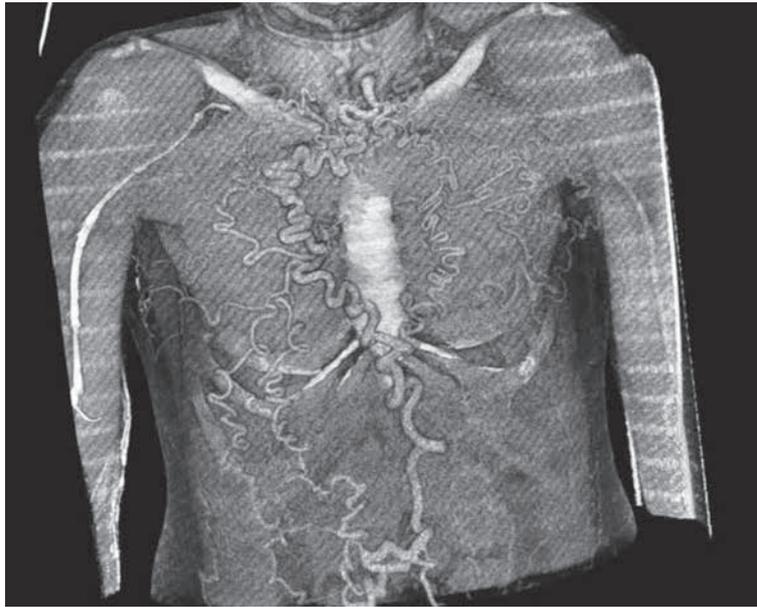


Fig. $\frac{1}{2}$

Work-up

On 3D-volume rendered MDCT angiography (Fig. 1), extensive superficial collateral blood vessels located at the chest wall and abdominal wall are demonstrated.

On coronal maximum intensity projection (MIP) image of the thorax, obtained during the arterial phase after administration of contrast medium (Fig. 2), narrowing of superior vena cava without thrombus is noticed (arrow). There is no evidence for lymphadenopathy nor associated mediastinal mass.

Radiological diagnosis

Based on clinical history and imaging features, the diagnosis was *superior vena cava syndrome caused by Behçet's disease* was made. Corticosteroids, cyclophosphamide and warfarin were given for treatment.

After treatment, partial improvement of the symptoms of the syndrome has been observed.

Discussion

The superior vena cava (SVC) syndrome is a clinical presentation caused by obstruction of the SVC or its major tributaries by intraluminal occlusion or by extrinsic compression and/or invasion from malignant and benign diseases. As a result the venous return of the head, neck and upper extremities is critically diminished.

The main causes of SVC syndrome are malignant intrathoracic tumors (such as bronchogenic carcinoma and lymphoma), iatrogenic factors (placement of central catheter, insertion of a pacemaker, etc.), and vasculitic syndromes such as Behçet's disease.

Behçet's disease is multisystemic vasculitis of unknown etiology. Its diagnosis is based on clinical criteria.

The disease is characterized by recurrent urogenital ulceration, cutaneous eruptions, ocular manifestations, arthritis and vasculitis.

Although SVC thrombosis is a rare but well-recognized manifestation of Behçet's disease, SVC syndrome due to vasculopathy, without evidence of thrombosis is very rare.

In the presented case, the SVC syndrome developed without evidence of thrombosis. Behçet's disease should be considered in the differential diagnosis of SVC syndrome.

Bibliography

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