Coarctation of Inferior Vena Cava in Situs Inversus Totalis

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Abstract

A woman aged 28 years with situs inversus totalis presented with chronic Budd-Chiari syndrome with symptoms of 3 years' duration. The left sided inferior vena cava was occulted below the diaphragm above a patent morphologic right hepatic vein. A dorsal cavalatrial bypass was attempted but abandoned due to bleeding from extensive collaterals.

The occurrence of coarctation of the inferior vena cava in situs inversus totalis lends support to the view that it is a congenital condition and occurs due to disturbance in fusion of the hepatic segment of the inferior vena cava and the hepatocardiac channel. (Indian J Gastroenterol 1992; 11: 89)

Key words: Budd-Chiari syndrome.

A 28 year old woman was hospitalized for anorexia, loss of weight and abdominal pain for 3 years, oliguria and irregular menstrual cycles for 1 year and gross ascites. Distended veins were seen over the front and back of the abdomen with flow from below upwards. The liver was firm and palpable 3 cm below the left costal margin. Spleen was palpable just below the right costal margin.

Investigations: Hemogram and biochemical investigations including liver function tests were normal. Sex hormone profile revealed: serum testosterone 0.40 ng/mL (normal 0.31±0.17), estradiol 300 pg/mL (108±7), prolactin 580 mU/mL (284±56), follicular stimulating hormone 4.8 IU/L (4.8±0.8) and leutinizing hormone 6.0 IU/L (8.58±1.43).

Chest X-ray and electrocardiogram revealed dextrocardia. Plain X-ray abdomen showed features of situs inversus. Esophagogastroscopy showed grade I esophageal varices. Functional hepatogram done through the left eighth intercostal space showed the 'right' hepatic vein draining freely into the inferior vena cava (IVC) which was filling from above downwards. There was no antegrade flow in the IVC. The IVC was completely occluded below the diaphragm immediately above the entry of the 'right' hepatic vein.

A dorsal cavalatrial bypass was attempted through a left thoracotomy. The procedure was abandoned due to excessive bleeding from extensive subdiaphragmatic venous channels in the region overlying the IVC. The patient refused reexploration.

The embryogenesis of the IVC is complex, making it prone to developmental anomalies. When union of the hepatic segment of the IVC and the hepatocardiac channel is not properly established, atresia or stenosis at this level could result, causing coarctation of the IVC. This theory has been disputed because of varied age of onset of symptoms. However, many congenital disorders manifest symptoms in later life. The occurrence of situs inversus totalis with coarctation of IVC in our patient lends support to the view that the coarctation is congenital in origin.

These lesions have been treated by a variety of procedures. Cavoatrial bypass graft is useful but failed in the present case because of technical difficulties in exposing the obstructed segment.

References