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CASE REPORT

Agenesis of the Inferior Vena Cava

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Introduction

Developmental anomalies of the inferior vena cava (IVC) are usually closely related to embryonal aberrations of the azygos and/or hemiazygos vein. Anomalies of the inferior vena cava are present in 0.6–4% of the population[1] and are due to the absence of a particular (often the hepatic) segment. Most frequently anomalies of the IVC present themselves with deep venous thrombosis of the iliac or femoral veins,[2] most certainly caused by impaired venous emptying. But also frequently the diagnosis is merely coincidental, the patient not having any clinical symptoms. This is because of early development of collaterals out of the remnants of the embryonal veins, which grow out to be the lumbar, azygos or hemiazygos system. Autopsy findings revealed that these collaterals could take over the function of the incomplete/deficient inferior vena cava, without causing any complaints to the patient.[1] The swollen (hemi-) azygos veins can cause diagnostic problems in the paravertebral and mediastinal area due to their tumour-like appearance. This dilemma can be solved by performing direct contrast opacification of the central venous system. Our observation documents one of such a rare findings and the diagnostic problems involved in it.

Case Report

A 22-year-old female was referred for further evaluation of a para-aortic tumour-like abnormality at the level of the lower part of the left kidney which was seen at ultrasonography, performed at the request of her general practitioner because of persisting complaints of low backpain.

Physical examination on admission revealed an healthy young woman, with isolated distinct percussion pain in the left lumbar region. No palpable abnormalities were found in the abdomen. Laboratory investigations showed no abnormalities. A non-contrast computed tomography of the abdomen and pelvis did show a solid para-aortic process medial to the left kidney, almost 15cm in diameter with a suspicion of a lymphoma. Several ultrasound guided needle biopsies of the process did not produce sufficient material for histological examination. An open biopsy was then performed. At lumbotomy a cystic structure was seen at the lower pole of the left kidney. Vein characteristics were missed by the surgeon and incision of the structure caused massive bleeding. It took a great effort to control this bleeding. A clear macroscopic diagnosis or sufficient material for microscopic examination could not be obtained. The surgical exploration was discontinued unsuccessfully. Considering the possibility of venous involvement, an ascending cavogram (Figs. 1 and 2) was attempted a few days later through both the left and right femoral vein. This revealed the true nature of the para-aortic mass. There was no visualisation of the inferior vena cava and the contrast medium flowed from the right iliac vein through large communicating veins into dilated and tortuous ascending lumbar veins. The phlebography also revealed an occlusion in the left iliac vein (Fig. 1).

Nucleair magnetic resonance imaging and re-evaluation of the computer tomography of the abdomen confirmed the diagnosis of agenesis of the inferior vena cava. The chest X-ray revealed a lobus vena azygos (Fig. 3), a sign of possible abnormal development of the central venous system.

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After this diagnosis, the patient was treated conservatively and was regularly seen on an outpatient basis. The low backpain, which exacerbated after the open biopsy and the massive bleeding, lessened, but never disappeared completely.

Discussion

The development of the inferior vena cava is a complex embryological process. It involves the total or partial retraction of three pairs of foetal veins. These are the posterior cardinal, the subcardinal and the supracardinal veins. The posterior cardinal veins lie posterolaterally in the foetus and are dominant at 6 weeks; the subcardinal veins lie ventromedially in the foetus and are dominant at 7 weeks, and the supracardinal veins, dominant at 8 weeks, lie dorsal to the aorta. These veins anastomose widely. The prerenal segment of the IVC is formed by the right subcardinal vein. The renal segment is formed by the anastomosis of the supra- and subcardinal veins. The right supracardinal vein together with part of the hepatic sinusoids, will form the postrenal (hepatic) segment of the IVC. The azygos and the hemiazygos system are formed out of the cranial part of the supracardinal veins (Fig. 4).

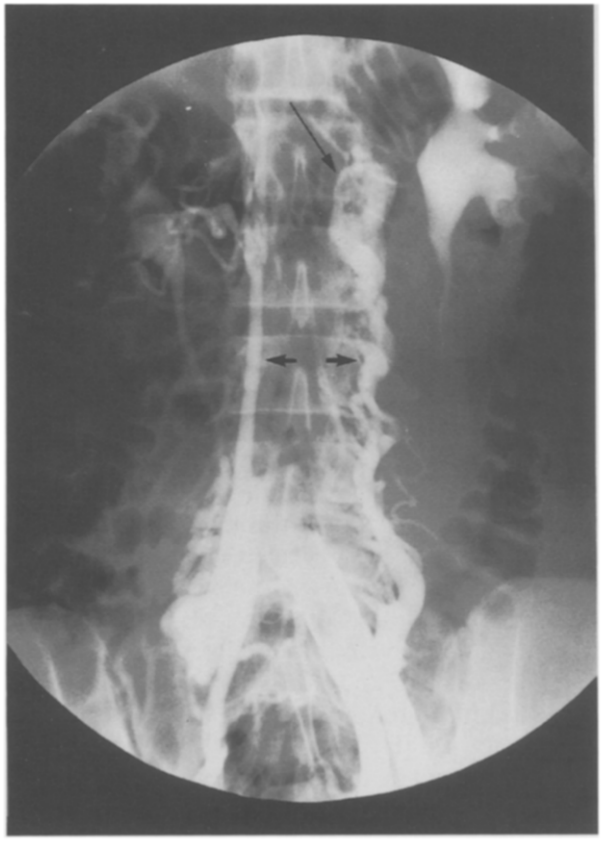

Fig. 1. Contrast medium is seen in the left and right iliac vein. A sudden stop is seen and continuation of flow is established through lumbar veins (small arrows). The paraortic mass is seen near the long arrow.

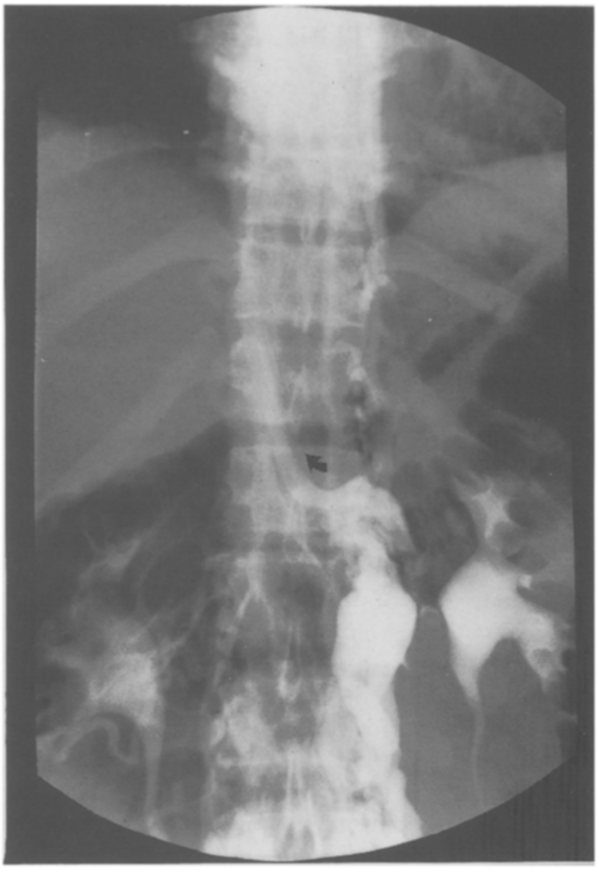

Fig. 2. From the paraortic mass the contrast flows through collaterals in the azygos vein (arrow).

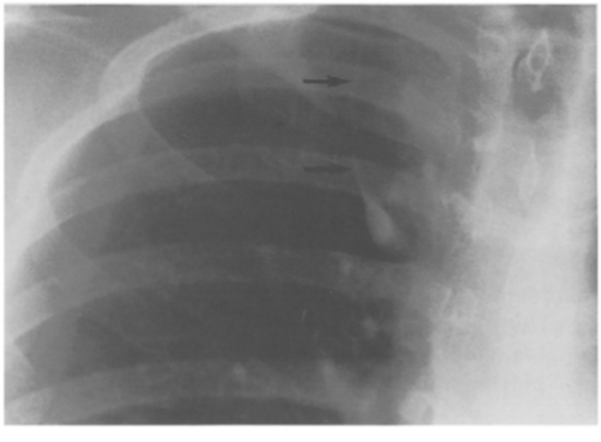

Fig. 3. X-chest reveals in the right top of the chest a lobus of the azygos vein (arrows).

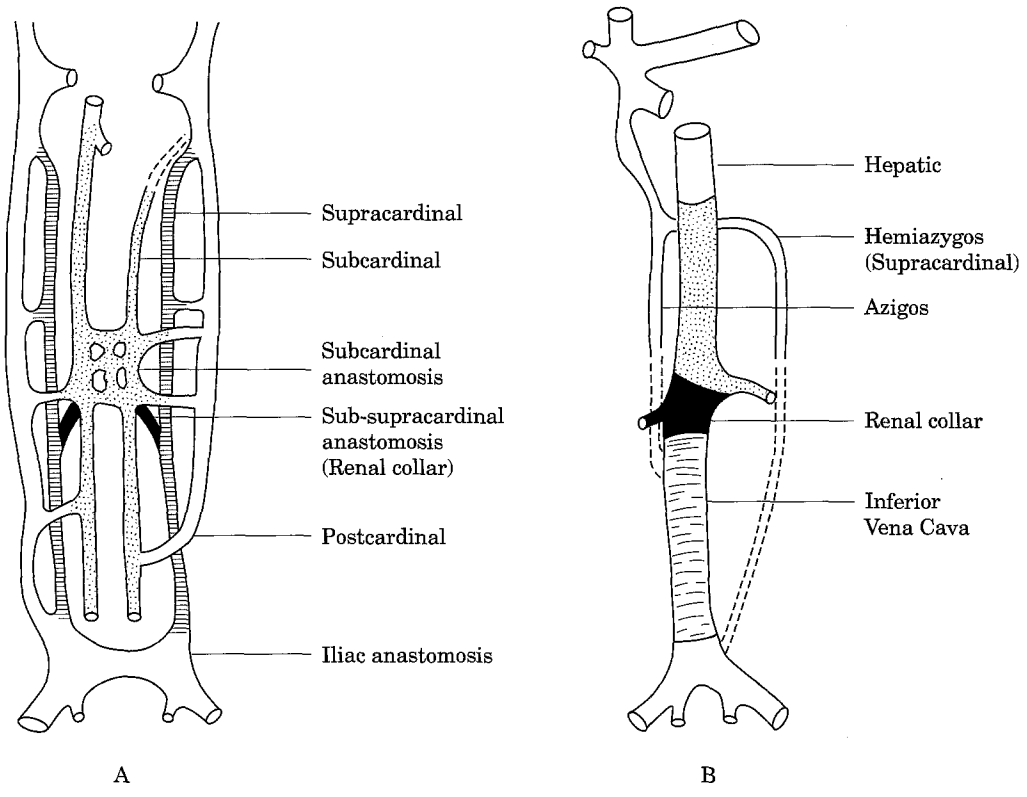

Fig. 4 Embryological development of the inferior vena cava at 7 weeks (A) and in the adult (B).

Considering the complexity of the development of the central venous system it is not difficult to envisage some possible abnormalities. A complete review is described by Anderson *et al.*[1] and Chuang *et al.*[3] The anomalies of the IVC and the closely related renal veins can be divided according to their segmental development.

The postrenal segmental anomalies constitute the following: (a) retroureteral IVC: persistence of the right posterior cardinal vein; (b) left IVC: persistence of the left and regression of the right supracardinal vein; and (c) double IVC: persistence of both supracardinal veins.

The renal segmental anomalies constitute renal vein anomalies and are as follows: (a) renal venous collar: persistence of both limbs of the circumaortic venous ring; and (b) retroaortic left renal vein: persistence of the dorsal and regression of the ventral limb of the circumaortic ring.

The prerenal and/or hepatic segmental anomalies arise if there is atresia or failure of union between the hepatic and right subcardinal veins. This anomaly has been described under a variety of names: infrahepatic interruption of the IVC with azygos and hemiazygos continuation, absent IVC, anomalous IVC with azygos drainage, absence of the hepatic segment of the IVC, persistence of the supracardinal vein, and continuation of the postcardinal vein. The blood from the postrenal IVC will return to the heart through the cranial portion of the supracardinal vein, which constitutes the azygos and hemiazygos veins.

If there is a multisegmental anomaly of the IVC, i.e. affecting both the postrenal and prerenal segments, shunts may develop with other veins in addition to the azygos and hemiazygos veins. These veins include the ascending lumbar veins, and the superficial abdominal veins.

The suspicion of agenesis of the IVC is usually raised indirectly by symptoms of iliac and femoral vein thrombosis,[2] enlargement of the vena azygos shadow at a plain chest X-ray,[2] and computer tomography scanning.[1,4] The anomaly should be considered especially in patients with congenital cardiac abnormalities like situs inversus.

In the reported case the findings at CT scanning should have led to a differential diagnosis including agenesis of the vena cava. Performance of a cavogram at that time would have revealed the diagnosis. The percutaneous and open biopsies could have been prevented. The initial complaints of the patient were probably caused by thrombosis of the left common iliac vein. The complaints disappeared probably due to recanalisation of this vein.

In short we state that the radiological findings, that

can alert us of the possible existence of agenesis of the inferior vena cava are: enlarged shadow of the azygos vein on a plain chest X-ray; the absence of the inferior vena cava shadow on a lateral view of the chest X-ray; and a cylindric (as opposed to round) paravertebral structure on subsequent CT images (in continuity with the azygos arch) and paravertebral masses in the abdomen as well as in the chest.

Familiarity with agenesis of the inferior vena cava may be helpful in case of differential diagnostic problems in paravertebral masses especially at the diaphragmatic level, thrombosis of unknown cause and diagnostic or therapeutic interventions concerning the IVC or its branches.

References

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