Full Inferior Vena Cava Agenesis Causing Acute Abdominal Symptoms: Case Report and Literature Review

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Abstract

Absence of the entire inferior vena cava caused acute abdominal symptoms, and the patient, a twenty-year-old man, was operated on on an emergency basis. Subsequent ascending venography, abdominal computed tomographic scan, intraarterial digital subtraction angiography, and intraosseous phlebography revealed full inferior vena cava and iliac venous system agenesis, up to and above the level of the hepatic veins, venous return from the lower limbs and the abdominal viscera being through a series of multiple collateral channels and the azygos-hemiazygos system.

Introduction

The following developmental anomalies of the inferior vena cava (IVC), which arises from various segments, have been distinguished: (1) left-sided position (0.5%), (2) duplication (3%), (3) anomalous insertion into the left atrium, (4) agenesis, and (5) stenosis and occlusion.1,2 “Absent IVC” designates a well-defined abnormality with absence of the prerenal (supra-renal) segment of the IVC.3,6 Various abnormalities of the postrenal (infrarenal) IVC have been reported, but absence of the postrenal segment is indeed very uncommon.4,6 “Full IVC agenesis,” an extremely rare condition, may be compatible with normal adolescence,9 and such a congenital abnormality has never to our knowledge caused acute abdominal symptoms.

Case Report

A twenty-year-old white man was admitted owing to gradually deteriorating constipation, left abdomen pain, pyrexia (38.5°C), and prostration. On examination he was lying still on the
bed, he was apprehensive, his color was normal, his breath was diaphragmatic (25/min), and his pulse rate was 100/min. His lower extremities were large but not edematous (no pitting edema). He stated that at times his lower extremities would become even larger, especially following vigorous exercise, since childhood. No varicose veins, caput medusae, or distended abdominal wall veins were seen, but on inspection the abdomen was slightly prominent in the left iliac fossa. His medical history was unremarkable.

His bowel sounds were diminished, his abdomen was tender and rigid especially on the left side, and rebound tenderness could be elicited. On rectal examination a palpable, soft, tender mass was felt. Hematologic and biochemical reports, urinalysis, chest x-ray, abdominal x-ray, and ECG were within normal reference ranges. An abdominal computed tomographic (CT) scan before and after injection of contrast material disclosed: (1) a collection of fluid in the left prerenal and left retroperitoneal space, (2) a mass having a tube-like appearance beginning just below the left renal vein and extending to the level of the left iliac crest, (3) numerous rounded masses in the pelvis and abdomen quite apart from the opacified bowel, (4) dilatation and significant lateral displacement of the left ureter, and (5) displacement of the urinary bladder to the right and cephalad. During the thirty-six-hour investigation in the hospital the patient fainted twice when he attempted to stay upright.

With the possible diagnosis of an abscess and under combination chemotherapy, the patient was operated on on an emergency basis. Upon entrance to the peritoneal cavity through a midline incision, about 200 mL of blood was found, the sigmoid colon was displaced medially, and the retroperitoneal space was bulging anteriorly, having the appearance of a large, black, grape-like structure and giving the impression of having bled recently. On palpation this structure was soft, but some regions were solid. An attempt to sever the retroperitoneum caused remarkable hemorrhage. Following laborious hemostasis a tube drain was left in place and the abdomen was closed. The next day the patient was feverish (39°C) but stable, and so intravenous pyelography (IVP) was performed, which disclosed obstruction of the left ureter at the level of its lower third (Fig. 1).

On the second postoperative day the patient developed extensive edema of both lower extremities extending to the level of the lower abdomen, and he remained pyrexial (39°C). Phlebothrombosis of both lower extremities was confirmed by duplex ultrasonography, and the patient was placed on full-dose heparin regardless of having been operated on recently and the coexistent spontaneous retroperitoneal hemorrhage.

On the third postoperative day the patient continued to be feverish and prostrated, and it was decided to have him reexplored in order to relieve the lower third obstruction of the left ureter, for it was thought that this was the most possible cause of his pyrexia. At that time percutaneous renal pelvis drainage was judged to be unsuitable for the patient. Just before reoperation there was heparin reversal with protamine sulfate, and all bleeding parameters were normal. Heparin was reinstituted immediately after operation. During reoperation the left ureter was mobilized and made free of the constricting venules, a stent was placed in it because it had had to be repaired during mobilization, and there was considerable blood loss.

Two days later the temperature returned to normal and the antibiotics were discontinued on the seventh postoperative day, but anticoagulant therapy continued with warfarin. The first signs of improvement of the deep phlebothrombosis were seen on the sixth postoperative day,
and on the twentieth day the patient’s lower extremities had returned to their usual size.

The patient was discharged on a warfarin regimen in good condition on the thirtieth postoperative day, and bilateral ascending venography (Fig. 2), intraosseous venography (Fig. 3), intraarterial digital subtraction angiography (DSA), a new abdominal CT scan (Fig. 4), and IVP (Fig. 5a, 5b) were scheduled.

These investigations proved that the patient has total agenesis of his inferior vena cava (IVC) and iliac venous system, up to and above the level of the hepatic veins, venous return from the lower limbs and the abdominal viscera being through a series of multiple collateral channels and the azygos-hemiazygos system.

Three months later he developed extensive venous collaterals on the lateral aspects of his lower abdomen, especially on the left side (left superficial lower epigastric vein and thoracoepigastric vein). The medial and lateral saphenous veins on both sides were dilated. These new clinical findings were compatible with those already known from the ascending phlebogram of his lower extremities (Fig. 2).

Six months later the patient complained of lower back pain on exertion (quick walking on uphill ground) ceasing three to five minutes following rest. This has been ascribed to venous blood pooling in the retroperitoneal collaterals during exercise due both to still obstructed channels and inadequate superficial abdominal wall collaterals. The patient has been instructed to reduce the magnitude of exertion, and his problem has now been solved.
FIG. 3. Intraosseous phlebogram of pelvis and abdomen showing neither evidence of patent external or common iliac veins nor of IVC. Very enlarged azygos and hemiazygos veins are seen (These two photographs were joined together for better understanding of the condition).

FIG. 4. Abdominal CT scan at the level of the liver showing vessels in front of the vertebral column representing the aorta (middle) and the azygos and hemiazygos veins (on either side of the aorta) but no IVC.

Discussion

The development of the IVC involves fusion, regression, and establishment of midline anastomoses between parts of three paired fetal venous systems: the two posterior cardinal veins, the two subcardinal, and two supracardinal veins. The changes in the abdominal venous systems that produce the final IVC cover a long period from day 25 to day 50, mainly between days 29 and 41. Depending on the embryonic origin, four parts of the IVC have been recognized: (1) hepatic segment, (2) prerenal segment, (3) renal segment and, (4) postrenal
“Absent IVC” normally designates absence of the prerenal segment of the IVC only, with azygos continuation being the most common variety of an interrupted IVC. “Full IVC agenesis” should cover those cases in which all four parts of the IVC and the iliac venous system are absent and blood return is accomplished by one or both of the following pathways: (1) vertebrolumbar pathway (anterior external vertebral plexus, ascending lumbar veins, and azygos and hemiazygos veins), and (2) superficial anterior abdominal wall collaterals. This definition is exemplified by the present case and other ones previously reported.

The incidence of an absent IVC in the general population is very small, whereas this ranges from 0.6% to 1.3% among patients with malformations of the heart and increases to 20% in situs inversus and to 46% in the asplenia syndrome. These conditions have in common their dependence on a blood supply that is changing from being symmetrical to asymmetrical. Any uncertainty about whether the situs is to be normal or inverted may cause bilateral superior vena cavae or an absent IVC.

Computed tomography is the investigation of first choice, before cavography, which demonstrates the associated venous anomalies (retro-aortic and para-aortic space-occupying structures representing the enlarged collaterals draining the lower half of the body). Exact diagnosis and classification can be made also via intravenous and intraarterial DSA and magnetic resonance imaging. Most of the reported cases have been recognized as a result of angiocardiography when the advancing catheter via the femoral vein encountered difficulty.
Some cases have been diagnosed accidentally on a posteroanterior chest film by an opacity, in
the right tracheobronchial angle corresponding to the very dilated azygos vein.\textsuperscript{7,17,23,25}

In making the differential diagnosis the following conditions should be taken into account:

1. Pathological mediastinal lesions and other vascular anomalies, when the IVC agenesis
   presents as a mediastinal abnormality on the chest radiograph, to avoid mediastinoscopy or thoracotomy\textsuperscript{6,7}
2. Causes of dilatation of the azygos-hemiazygos venous system\textsuperscript{13}
3. The syndrome of obstruction of IVC in childhood\textsuperscript{3}
4. Abdominal wall venous collaterals and varicose veins because excision of these veins
   would interrupt a vital venous return or at least result in intractable ulcers\textsuperscript{6}
5. Retroperitoneal and para-aortic space-occupying structures\textsuperscript{13,19}
6. Causes of difficulty or prevention of catheterization of the heart from the IVC\textsuperscript{4,11}
7. Other causes of acute abdominal symptoms (although it should be last in the list)
8. Causes of ureteric obstruction
9. Causes of retroperitoneal hemorrhage
10. Causes of lower back pain
11. Causes of Klippel-Trenaunay syndrome\textsuperscript{26}

There are at present no known therapeutic measures for this very rare anomaly.\textsuperscript{19} In pure
IVC agenesis disclosed accidentally (eg, by chest x-ray),\textsuperscript{7,18,23-25} those cases revealed during
investigation of patients with congenital heart abnormalities,\textsuperscript{4,11,27} and those in symptomatic
patients,\textsuperscript{6,19} the management should be focused on the prevention of complications, such as
thromboses in the distal drainage channels.\textsuperscript{19}

On the other hand, knowledge of this condition is important for surgeons to avoid ligation
of the azygos vein in the course of pulmonary or cardiac operation because prevention of the
only major route of venous drainage from below the diaphragm may result in death.\textsuperscript{4,7,28}
In complicated cases, eg, hypertension due to a hypoplastic kidney, or ureteric obstruction from
constricting venous collaterals as in the present case, surgery is unavoidable. When thrombo-
sis occurs,\textsuperscript{28,29,31} a life-threatening condition for this type of patient, vigorous anticoagulant
therapy has to be instituted promptly that will last until sufficient venous return has been
reestablished.

We believe that prolonged anticoagulant therapy has no benefits; on the contrary it in-
creases the risk of retroperitoneal hemorrhage. When thrombosis and retroperitoneal hemor-
rhage occur simultaneously, as in the present case, anticoagulant therapy should not be a
contraindication, because hemorrhage is the end result when blood cannot escape forward.
Note here a discrepancy between the extent of venous collaterals found on operation and seen
by CT scan and those shown by phlebography. It is evident that a large part of the retroperito-
neal venous collaterals is not operational. Obviously this is due to previous subclinical throm-
booses and subsequent development of new collaterals, which is in accord with the patient’s
history of having at times "large legs" since childhood. The clinical course of this case has
proved that dilated superficial abdominal wall veins do not appear until a critical number of
obstructed deep venous collaterals has been exceeded.

The prognosis of this rare congenital anomaly seems to be directly related to the presence
of thrombosis in the retroperitoneal venous collaterals and to any coexistent congenital anom}-
lies (cardiac and vascular defects, situs inversus). In “full IVC agenesis” thrombosis is the major cause of mortality \(^9,11\) and morbidity \(^6,12,19,29-31\) (paresthesias of lower legs and lower back pain on exertion, retroperitoneal hemorrhage, hypertension, ureteric obstruction, and emotional distress due to dilated superficial abdominal wall veins). Apart from this anomaly per se, \(^31\) implicating factors favoring thrombosis in this category of patients have been reported such as (1) oral contraceptives and cigarette smoking \(^30\); (2) increased intraabdominal pressure (constipation, tumors, pregnancy), and (3) dehydration, marasmus, sepsis, and exertion. \(^9\) Therefore, prevention of thrombosis must be our mainstay in helping these patients.

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**References**