Symptomatic congenital saccular aneurysm of the inferior vena cava associated with a circumaortic left renal vein

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Congenital saccular aneurysms of the inferior vena cava (IVC) are extremely rare, with 23 cases reported so far. We present a case of a 55-year-old woman with an acute episode of back pain that mimicked aortic dissection. Computed tomography ruled out aortic dissection but demonstrated a saccular aneurysm of the infrarenal IVC and a circumaortic left renal vein. The saccular aneurysm was excised, and the postoperative anatomopathologic examination revealed that it was congenital in nature. Surprisingly, preoperative symptoms of recurrent palpitations, dyspnea, and positional abdominal discomfort resolved after resection. This is the first reported case of a symptomatic congenital saccular aneurysm of the IVC with an associated circumaortic left renal vein. (J Vasc Surg Cases 2015;1:9-12.)

The development of the inferior vena cava (IVC) takes place at 6 to 8 weeks of gestation through a complex mechanism that begins with formation of three pairs of embryonic veins (posterior cardinal, subcardinal, and supracardinal veins). These veins initially interconnect and then partially regress to eventually form the definitive IVC, in addition to the entire venous network of the thorax, abdomen, and pelvis.[1] Any point in this sequence constitutes a potential area of malformation. However, the same complex network invariably substitutes some form of venous flow through an alternative route.

Unlike arterial aneurysms, the term "venous aneurysm" lacks a precise definition in the literature. Some reports suggest that the normal diameter of the infrarenal IVC in adults ranges from 1.5 to 3.7 cm when measured by computed tomography (CT) scan.[2] Any fusiform dilatation larger than this diameter on CT is regarded as an IVC aneurysm (IVCA). Saccular aneurysms, on the other hand, are different. According to the Gradman and Steinberg classification,[3] type 4 IVCAs (miscellaneous) encompass all saccular aneurysms, regardless of their location or association with congenital anomalies. It is important to note that this classification was proposed in 1993 based on review of only 13 cases.

We conducted an extensive PubMed literature search using the keywords inferior vena cava, congenital, aneurysm, saccular, and symptomatic. To the best of our knowledge and based on this systematic search of all published reports, only 30 cases of nontraumatic IVCAs have been reported.[4]

In this report, we describe a rare case of a congenital saccular IVCA in a 55-year-old woman with acute back pain and a background history of recurrent chest pain, shortness of breath, and palpitations. This IVCA was associated with a circumaortic left renal vein (CLRV), another rare anomaly. Written informed consent was obtained from the patient for publication of this case report and accompanying images. To the best of our knowledge, this is the first reported case of saccular aneurysm of the IVC associated with a CLRV.

CASE REPORT

A 55-year-old woman presented to the emergency department with a 3-hour history of severe back pain radiating to the chest. The pain was followed by a syncopal episode lasting 2 minutes with prodrome. She gave no history of cardiac disease or any syncopal attack, except for recurrent mild shortness of breath, palpitations, and chest pain, which she attributed to normal stresses of life. Her background history included type 2 diabetes mellitus, dyslipidemia, depression, and polycystic ovarian syndrome.

The patient's heart rate, blood pressure, and respiratory rate were all within normal reference ranges, and the results of chest and abdominal examinations were normal, except for mild abdominal tenderness. The results of blood investigations were within normal reference ranges except for mild transaminasemia (aspartate transaminase, 200 U/L; alanine transaminase, 120 U/L), which reverted to normal within a few days.

Results of CT brain, echocardiogram, and 24-hour Holter monitoring were normal. An abdominal ultrasound assessment suggested an intimal flap in the abdominal aorta concerning for

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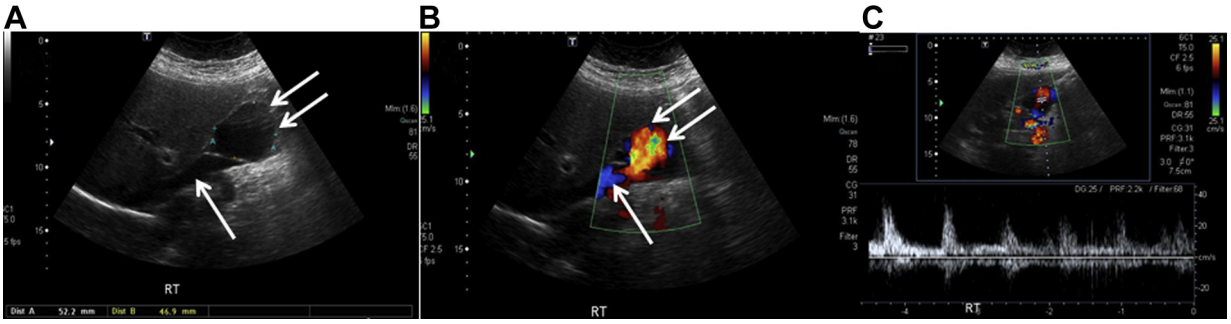

Fig 1. Abdominal ultrasound **(A)** B-mode imaging and **(B)** color-flow imaging demonstrates a vascular structure anterior to the aorta and inferior vena cava (IVC). **C,** Duplex demonstrates a "ying-yang" appearance on Doppler flow. The *single arrow* points to the IVC, and the *double arrows* point to the IVC aneurysm (IVCA).

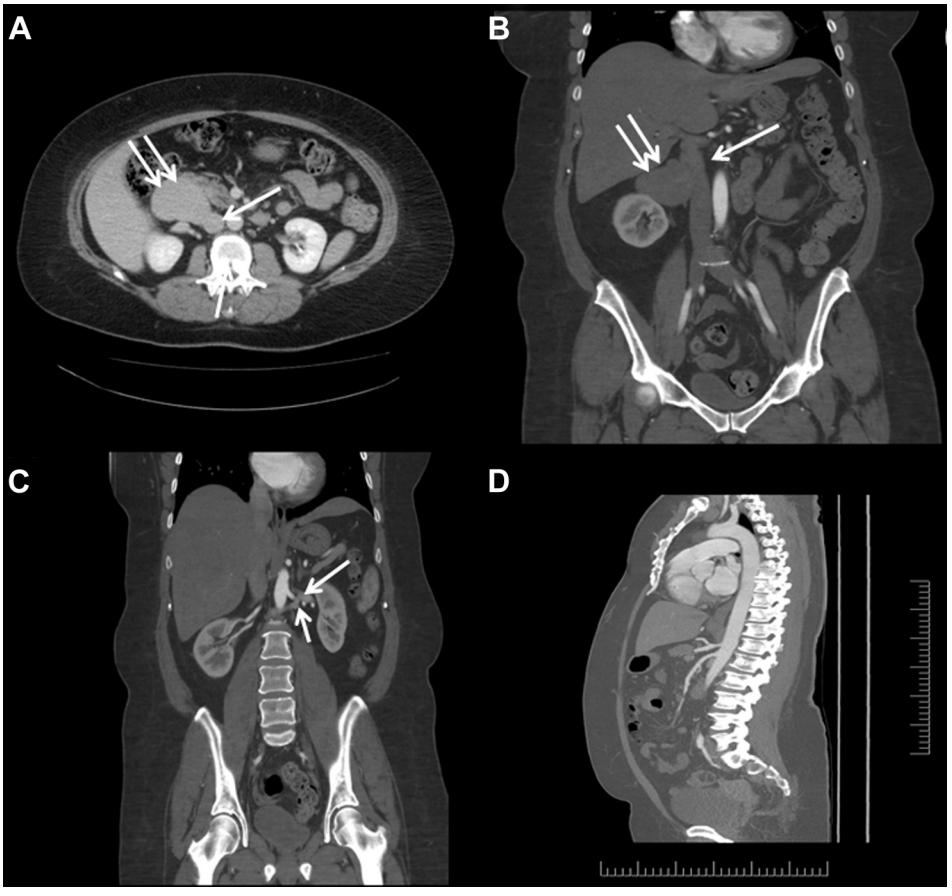

Fig 2. Abdominal computed tomography (CT) scan (venous phase) in **(A)** axial and **(B)** coronal views demonstrates a saccular inferior vena cava (IVC) aneurysm (IVCA). The *single arrow* points to the IVC and the *double arrow* points to the IVCA. The images show **(C)** the circumaortic left renal vein (CLRV) between the two *single arrows* and **(D)** a sagittal section of the thoracoabdominal aorta with no dissection.

aortic dissection and also described a vascular structure adjacent to the infrahepatic IVC, likely representing an aneurysm (Fig 1). Interestingly, thorax, abdomen, and pelvis CTs ruled out aortic dissection, but reported an "outpouching" from the lateral aspect of the IVC just below the right renal vein measuring 6.3 × 4.9 cm (Fig 2).

Because her back pain resolved and she remained stable over the days after admission, the patient was discharged with a follow-up CT scan in 8 weeks. Although no structural changes were demonstrated on the repeat CT scan, the patient reported recurrence of her symptoms of chest pain, shortness of breath, and palpitations and was anxious to have the aneurysm excised.

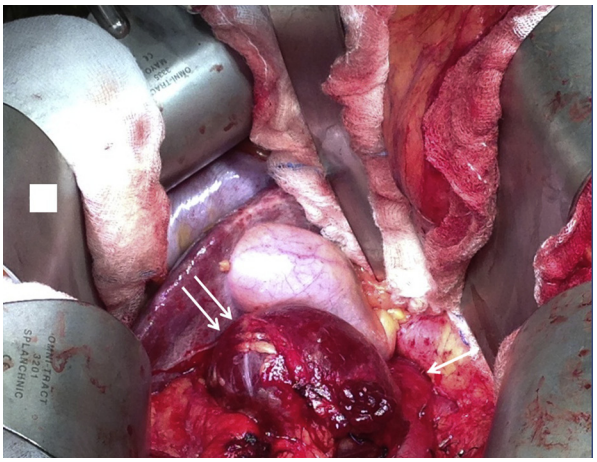

Fig 3. Operative photograph of the midline laparotomy with a self-retaining retractor shows large saccular aneurysm of the inferior vena cava (IVC). The *single arrow* points to the IVC, and the *double arrow* points to the IVC aneurysm (IVCA).

Her decision was felt to be reasonable. All possible risks were discussed in addition to operative strategies, such as deep vein harvest, in case excision and an interposition graft was needed.

At laparotomy, using right-sided medial-visceral rotation, a large purple mass (Video) showing characteristic systemic venous pulsations was found adjacent to the lateral wall of the infrarenal IVC. The IVC below the mass was dissected first, followed by the suprarenal IVC and the right renal vein. In addition to a normal-appearing left renal vein, a large accessory retroaortic left renal vein was encountered and preserved. The final steps of the dissection enabled clear identification of a saccular aneurysm with its communicating 2-cm neck. (Fig 3).

The patient was systemically heparinized before placement of a side-biting clamp on the IVC and the right renal vein to exclude the base of this saccular aneurysm. A small vessel loop was applied for better control of the right renal vein. The aneurysm was then decompressed using a small needle to facilitate precise excision down to the normal lateral wall of the IVC. The aneurysm was excised, and the defect was closed primarily using a double layer of continuous 4-0 polypropylene suture. The excised aneurysm contained no thrombus, and the subsequent anatomopathologic report confirmed its congenital nature, being a true aneurysm lined with intima, media, and adventitia.

On her first follow-up clinic visit 2 weeks postoperatively, and contrary to the contextual pathophysiology, the patient stated that all of her symptoms of recurrent chest pain, shortness of breath, and palpitations had resolved completely. At 6 months, the patient remained asymptomatic, with complete resolution of the saccular aneurysm on the postoperative CT scan.

DISCUSSION

Our current knowledge regarding IVCAs is mostly based on isolated case reports, an unfortunate fact that makes the exact natural history difficult to interpret. IVCAs, whether fusiform or saccular, symptomatic or asymptomatic, have been managed conservatively or surgically without a clear understanding of the manners in which they progress. A prospective registry for all nontraumatic IVCAs is greatly needed to further understand their behavior and design appropriate management strategies for this rare entity.

Davidovic et al[4] reported an IVCA and reviewed 29 others. Within the total of 30 IVCAs, they identified 23 saccular aneurysms, of which 16 were symptomatic, including their own patient. Those symptoms were mainly lower limb swelling, abdominal pain, or their combination. Only one report, by De Bree et al,[5] of those analyzed by Davidovic et al[4] described back pain as the presenting symptom, and none of the 30 reported IVCAs was associated with a circumaortic or retroaortic left renal vein. De Bree et al[5] listed thrombosis of the IVC and both iliofemoral veins as the underlying cause of the back pain. No IVC thrombus was identified in our patient, which suggests that there was a thrombus that embolized and caused the chest pain radiating to the back or, possibly, there was a recent expansion of the saccular aneurysm that caused her back pain.

The potential for bleeding and thromboembolic complications should be considered when surgery is contemplated. In contrast to fusiform aneurysms, full excision of the entire IVC segment containing the saccular aneurysm with interposition graft is rarely required. Surgical resection of a saccular aneurysm is commonly followed by lateral venorrhaphy to close the defect, and this can be accomplished by simple primary suturing,[6] as in our case, or by using a bovine pericardial patch[7] if the neck of the saccular aneurysm is too wide.

Because no other etiologic factor was identified on the clinical or radiologic assessment, our patient's IVCA was initially labelled as congenital, and this was postoperatively confirmed by the presence of all three layers: intima, media, and adventitia. The appearance therefore was that of a true aneurysm.

CLRV is a rare congenital anomaly and was found in 200 of 2839 cadavers donated to an anatomy laboratory.[8] The reported incidence in clinical practice is 1.8% (287 of 16,221). Three types of CLRV have been described.[8] The presence of a circumaortic or retroaortic left renal vein has a very important clinical and surgical significance, especially in open aortic surgery or surgery around the IVC, as in our patient.

CONCLUSIONS

IVCAs constitute a rare congenital anomaly, and their association with a retroaortic left renal vein has never been described. A prospective registry of all reported cases of IVCAs is needed. Given the small number of reported cases, very little is known about the natural course of IVCAs, and without this knowledge, making clinical decisions regarding treatment is difficult. In our case, the patient was possibly symptomatic and very keen to have it repaired. However, the treatment of this category of patients remains controversial in the absence of characterized natural history.

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