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Transcaval ureter: case report and a review of the literature

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(*) *in memoriam*

SUMMARY

The authors describe a case of post-mortem transcaval ureter. This is a rare congenital anomaly, where the inferior vena cava (IVC) forms a circle over the right ureter, in a vascular ring. In the bibliographic survey carried out through MEDLINE it was found that only 8 cases have been described in the worldwide literature as of December 2004.

Key words: Transcaval ureter – Postcaval ureter – Retrocaval ureter – Venous anomalies – Anatomic variations

INTRODUCTION

The formation of the inferior vena cava (IVC) has its origin in a complex venous system which develops by anastomosis between 3 pairs of embryologic veins. Variations in the development of the IVC result from the patency, atrophy or from the disappearance of those veins (Lopes et al., 1992).
The anomaly reported here was an occasional finding, which seems to be very rare, as demonstrated in our survey of the literature.

CASE REPORT

The corpse of an elderly white man, apparently around seventy, showed a rare anatomical variation of circumcaval ureter, in which the right ureter crossed a vascular ring formed by a duplication of the post-renal segment of the inferior vena cava (Figs. 1 and 2).
The right kidney had a normal anatomical appearance, even though it exhibited a pyelo-ureteral dilatation that extended until close to one of the sides of the ring of the inferior vena cava, shaped like an inverted "J".
To the left, the kidney and ureter did not exhibit any anatomical change.

DISCUSSION

The first case of retrocaval ureter was described by Hochstetter in 1893 (cited by Heslin and Mamonas, 1951). Owing of its position has been designated retrocaval ureter, postcaval ureter, circumcaval ureter, deflected ureter or even preureteric vena cava (Heslin and Mamonas, 1951).
It is important to stress that the transcaval ureter is a condition that has its origin in a venous anomaly and not a ureteral abnormality (Randall and Campbell, 1953).
Inferior vena cava variations are classified according to the region of their occurrence, namely: hepatic, prerenal, renal and post-renal. According to Huntington and McGure, the possible variations in the post-renal segment are as follows (cited by Heslin and Mamonas, 1951, and Lopes et al., 1992):

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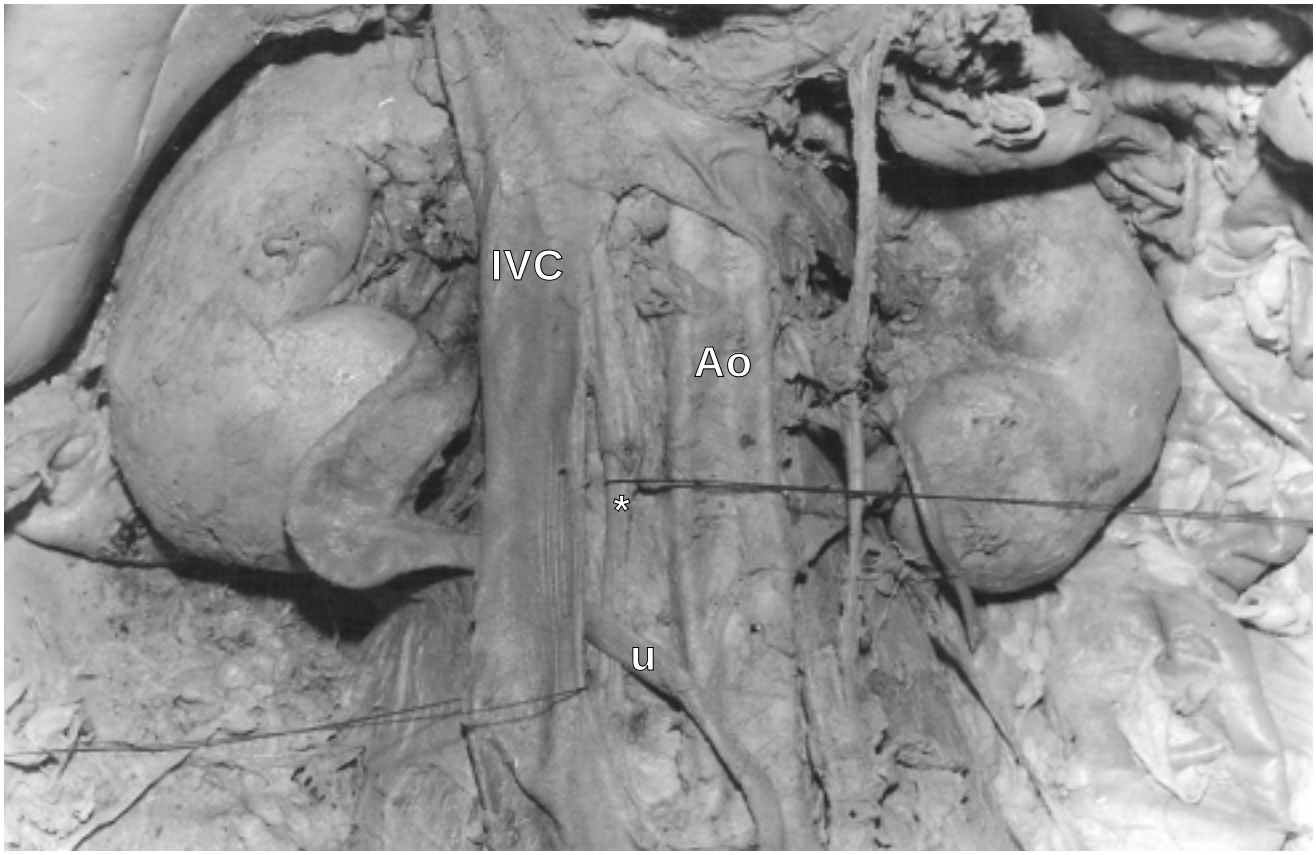

Figure 1.- Anatomic finding. Black lines are silk surgical sutures used to facilitate visualization. Ao: Aorta artery; IVC: inferior vena cava; *: accessory IVC; u: ureter.

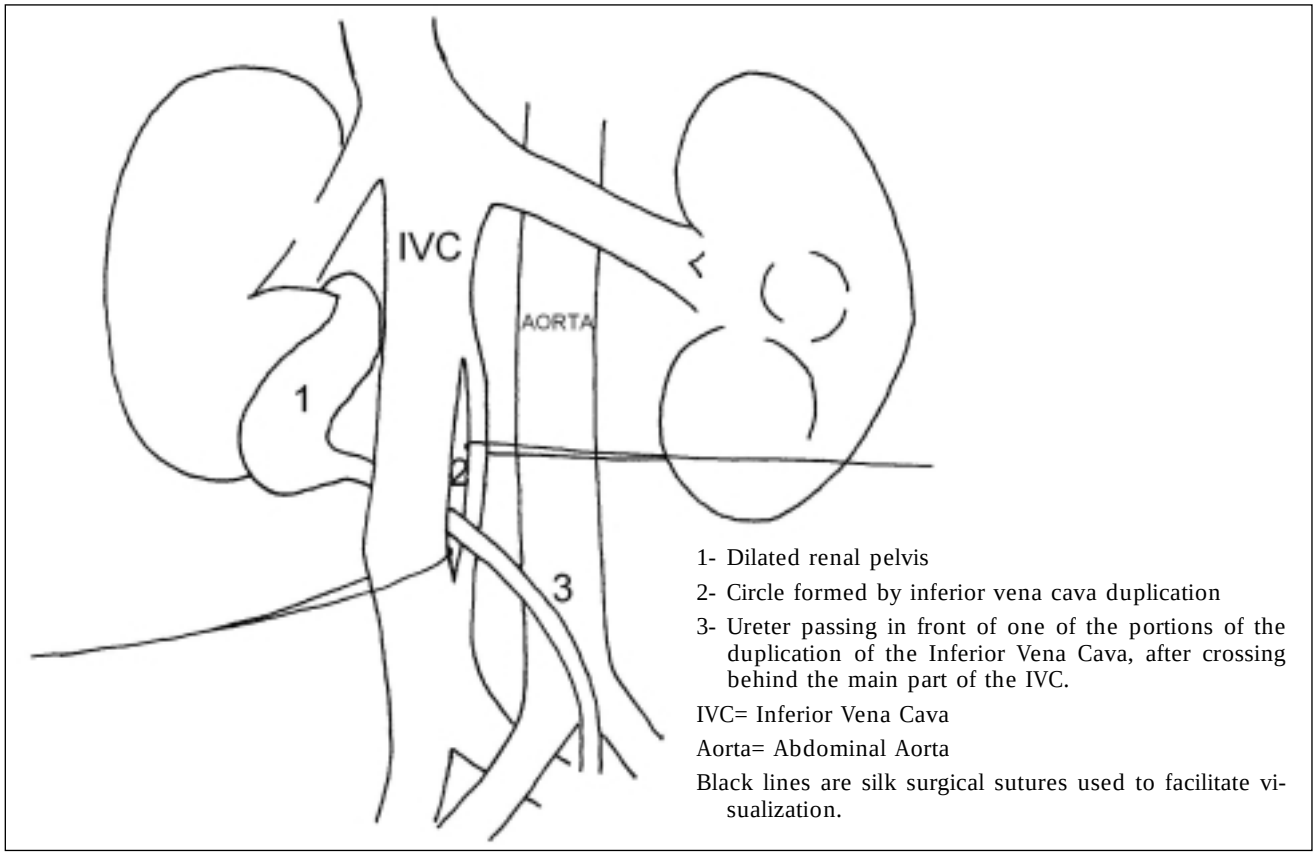

Figure 2.- Schematic drawing for figure 1.

- Type A: Retrocaval ureter
- Type B: Normal inferior vena cava
- Type C: Left inferior vena cava
- Type BC: Double inferior vena cava
- Type AB: Periureteral venous ring

The normally formed post-renal portion consists mainly of the supracardinal vein, dorsal to the ureter, which consequently does not deviate it or interfere in its function. The persistence of the dorsal posterior cardinal vein to the ureter causes an expressive medial dislocation and a restrictive compression when the ureter moves from a dorsolateral to a ventromedial position around the posterior cardinal vein, following the natural migration of the kidney towards its definitive position. This situation is called retrocaval ureter, although it would perhaps be better named "persistent pre-ureterical vena cava" (Heslin and Mamonas, 1951; Randall and Campbell, 1953).

The matter in hand represents an anomaly of the AB type. This is attributable to a unilateral persistence of the right posterior cardinal vein, together with the right supracardinal vein in post-renal position, forming a double inferior vena cava, both on the right side, with the ureter passing through the venous ring that it forms (Fig. 2).

A more specific classification for the circumcaval ureter was proposed in 1969, discriminating two types (Bateson and Atkinson, 1969):

– Type I: This is the most frequent. It has the radiological appearance of a fishhook or "S". The dilatation of the pyelo-ureteral junction is interrupted next to the margin of the IVC.
– Type II: The radiological appearance is that of a sickle up to the level of the obstruction. The hydronephrosis is not so severe and seems to end exactly at the level of the IVC wall.

Some cases are not susceptible to classification, since they seem to lie within the existing spectrum between the two above-mentioned types. Kenawi has demonstrated that of 162 cases analyzed approximately 93% of the circumcaval ureters could be included in the type I classification, and the remainder in type II, a proportion resembling that found in a Chinese study reporting a predominance of 94% of type I circumcaval ureters (Kenawi and Williams, 1976; Xiaodong et al., 1990).

In our survey of the literature, we found extreme variations in the incidence of circumcaval ureters (1:570 – Pick Anson to 1:4185 – DeCarlo, cited by Heslin and Mamonas, 1951), and probably none of the reports reflects the facts reliably, although several cases were identified in human anatomy laboratories, as we state for our own case. In the bibliographic survey carried out through MEDLINE it was found that only 8 cases have been described in the worldwide literature as of December 2004 (Gazaigne et al., 2002).

On the other hand, reports on the male/female ratio in humans coincide in the descriptions by different authors, with about 2.8 males to each female, in spite of the existence of reports in disagreement. One explanation for this male preponderance is the fact that there is a higher frequency of male corpses in anatomy laboratories. Nevertheless, in a study carried out on 34 patients 27 were men and 7 were women, resulting in a man/woman ratio of 3.9 (Xiao-dong et al., 1990).

The intensity of the symptoms vary, passing from complete absence to severe pain in colic with irradiation to the ureteral stretch. In a Chinese study (Xiaodong et al., 1990), the main clinical presentations were pain in the right flank in all of 34 patients; vesical irritation in 9 (26.5%), and hematuria in 24 (70.5%). During image examinations, the urography provided evidence of hydronephrosis on the right side in 29 patients (85%), being normal in only 3 of them.

Diagnosis is mainly accomplished by imaging studies (Randall and Campbell, 1953; Heslin and Mamonas, 1951; Bateson and Atkinson, 1969; Kenawi and Williams, 1976; Gefter et al., 1978; Murphy et al., 1987; Rosen et al., 1989; Xiaodong et al., 1990; Lopes et al., 1992; Gazaigne et al., 2002). The kidneys, ureters and the bladder are normal except if there is stone formation. Kidney function tests remain unchanged. The retrograde pyelogram shows the characteristics described above (Types I and II). The main differential diagnoses are retroperitoneal masses or fibrosis, achieved with radiological examination. Ultrasound and computerised tomography are of great use in the evaluation of this situation because they are less invasive than explorations using contrast (Gefter et al., 1978; Murphy, 1987; Lautin et al., 1988; Herman, 1991).

Treatment may be surgical or not, depending on the level and severity of the hydronephrosis, on the renal function deficit, and on the type of the abnormality. Clinical treatment consists of periodic examination, and is reserved for patients with no evidence of infection, no stone formation, and for those without or very mild hydronephrosis. Surgical treatment involves many procedures, such as section and reanastomosis of the ureter with its distal remainder or with the bladder; section and reanastomosis of the inferior vena cava; nephrectomy and finally, support of the inferior vena cava with pads (Lowsley, 1946; Heslin and Mamonas, 1951; Cathro, 1952; Bateson and Atkinson, 1969; Kenawi and Williams, 1976; Eidelman et al., 1978; Carion et al., 1979; Xiaodong et al., 1990).

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