Arteriovenous malformation causing obstructive uropathy: A different dimension to ovarian vein syndrome

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ABSTRACT

Ovarian vein syndrome is defined as obstructive uropathy caused by dilated ovarian vein with or without thrombosis. This is seen in the puerperal period as an acute condition with abdominal pain and fever and in multipara women with chronic recurrent abdominal pain. We report an ovarian vein syndrome caused by a true vascular anomaly in an 8-year-old child. Laparoscopic ureterolysis was performed with ligation of the arteriovenous malformation during the first operation. As ureterolysis was not effective, the patient was reoperated and ureteroureterostomy was performed after 3 months, which emphasizes the importance of removing the diseased segment even if it looks normal.

Key words: Arteriovenous malformation, gonadal vein syndrome, laparoscopy, ovarian vein syndrome, ovarian vein thrombosis, pelvic congestion syndrome, ureterolysis

INTRODUCTION

Ovarian vein syndrome is defined as ureteric obstruction caused by dilated ovarian vein with or without thrombosis and is a controversial and rare diagnosis. All cases reported so far are in adults where an abnormal gonadal vein compressed the ureter and caused obstructive uropathy mostly on the right side and lower ureter. Here we report the ovarian vein syndrome caused by an arteriovenous malformation affecting the left upper ureter in an 8-year-old child.

CASE REPORT

An 8-year-old girl presented with recurrent attacks of left flank pain of 6 months duration. She had no episodes of urinary tract infection. Ultrasound revealed left hydronephrosis with dilatation of the upper ureter. Intravenous pyelography (IVP) showed hydronephrosis with dilated left upper ureter with sudden tapering and normal lower ureter below the area of tapering [Figure 1]. She was diagnosed to have left upper ureteric obstruction and was taken up for laparoscopic exploration.

During surgery, we found that the upper ureter near the site of obstruction was surrounded by a sheet of vessels [Figure 2]. The ureter was sandwiched between a dilated periureteric vein and the gonadal vein. The gonadal vein instead of crossing from the lateral to medial side of the ureter, made a U shaped loop laterally, attached to the ureter at the apex of U and was seen draining into an abnormally placed lumbar vein with tributaries coming from above and below the confluence. The part of gonadal vein attached to the ureter was thrombosed with turbulent flow seen near the junction of gonadal vein with the lumbar vein. A small artery was seen entering the vein above the site of clot and causing turbulent flow in the vein. All the vessels were ligated proximally and distally laparoscopically and the parts of vessels attached to ureter were removed. The dilated gonadal vein attached to the left ureter was found to contain clotted blood. Complete ureterolysis was done at the site of obstruction. As ureter seemed normal, ureteroureterostomy was not done A line diagram is produced here for understanding the anomaly [Figure 3].

Postoperatively, Doppler study was done to uncover major vascular anomalies and was found to be normal. Double J (DJ) stent was removed after 15 days. Subsequently
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the patient continued to have abdominal pain again and a repeat IVP at 2 months showed similar obstruction as in the past. The patient was operated again and the obstructing segment was excised and ureteroureterostomy was performed. Ultrasound done after 3 months showed complete resolution of hydronephrosis and the patient remained asymptomatic.

DISCUSSION

Clarke described ovarian vein syndrome for the first time in 1964.[1] Following initial controversy, it was accepted as a clinical entity after further cases were reported.[2]

Two forms of the ovarian vein syndrome are now recognized. The first is a form of pelvic congestion syndrome analogous to varicocele in males. The right ovarian vein is most commonly involved. All patients are multipara, leading to the hypothesis that the dilatation of the ovarian vein is due to recurrent obstruction by the gravid uterus. However, as this condition is rare, it is always a diagnosis of exclusion. The patients usually present with recurrent abdominal, flank or pelvic pain, which increases in the lying down posture, mainly in the pre-menstrual period. The diagnosis is confirmed by imaging studies – ultrasound, Doppler, computed tomography and magnetic resonance imaging. The treatment consists of either percutaneous embolization or surgical ligation of the dilated vein.[3]

The second one is due to puerperal ovarian vein thrombophlebitis, which causes perivenous phlegmon formation and resultant periureteritis. The patients present with nonspecific abdominal pain and fever 2-3 days after delivery. The majority of patients respond to conservative measures with antibiotics, although some patients may require temporary DJ stenting or surgical ureterolysis with ligation of the ovarian vein.[4]

The present case adds a new dimension to the two types already described. This is the first case in a child, and is a true vascular anomaly. The abnormally placed gonadal vein was thrombosed and the resultant collaterals communicated with a branch of gonadal artery and caused this anomaly. The case also highlights the need for excising the obstructed segment of ureter along with ureterolysis in cases of complex vascular obstruction of upper ureter.

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Figure 1: Intravenous urography showing a ureter - sudden narrowing and tapering of the upper ureter with normal distal ureter

Figure 2: Intraoperative pictures showing vascular anomaly surrounding the upper ureter and its ligation (a) Ureter. (b) Dilated periureteric vein. (c) Thrombosed gonadal vein surrounding ureter. (d) Turbulent flow in gonadal vein. (e) Psoas major muscle. (f) Reflected colon. (g) Ligation of arteriovenous malformation above and below. (h) Arterial communication ligated

Figure 3: Line diagram showing the abnormal course of the gonadal vein and its communication with the lumbar vein (a) inferior vena cava. (b) Aorta. (c) Kidney. (d) Gonadal vein. (e) Gonadal Artery. (f) Communication of gonadal vein with a periureteric vein. (g) Dilated periureteric vein. (h) Thrombosed gonadal vein. (i) Arterial communication with the dilated gonadal vein. (j) Lumbar vein. (k) Ureter