Perforation of a ureteral diverticulum: a case report and review of the literature

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Abstract

The ureteral diverticulum represents a rare pathology. It is infrequently symptomatic and even more infrequently it manifests itself as an acute event. To our knowledge this is the only case described in the literature of perforated ureteral diverticulum with consequent uroperitoneum.
Key words: Ureteral diverticulum, perforation, false acute abdomen.

Introduction

The ureteral diverticulum represents a rare chapter in the pathology of the urinary system. In the majority of the cases the diagnosis is incidental. Infrequently it manifests itself with a proper symptomatology and more infrequently as an acute event. In the following case the diagnostic work up and the therapeutic approach were very elaborated due to the rarity of the pathology.
For this reason we present the case along with a thorough reviewed of the literature.

Riassunto

IL DIVERTICOLO URETERALE PERFORATO: CASO CLINICO E REVIEW

Il diverticolo ureterale rappresenta una patologia di raro riscontro, eccezionalmente caratterizzata da una sintomatologia propria ed ancora più eccezionalmente con un esordio legato ad una complicanza acuta. Il caso descritto dagli AA è probabilmente l'unico ad avere presentato una manifestazione complessa legata alla perforazione del diverticolo stesso con formazione di un uroperitoneo.
Parole chiave: Diverticolo ureterale, perforazione, falso addome acuto.

Case report

C.M.R. is a 22 years old female who presents to an outside hospital emergency room with a sharp and continuos pain to the left flank with radiation to the pelvis. Fever and signs of peritoneal irritation were absent. A sonographic examination revealed a left pelvic mass and approximately 1,000 cc of free intraperitoneal fluid.
The computed tomography confirmed the presence of free intraperitoneal fluid and showed irregularities of the left adnexa with a normal uterus. It was also noted the presence of left hydroureter to the sacral portion of it. No local lymphadenopathy or other intra-abdominal pathology was described.
The patient decided to go to a prestigious University of central Italy and was admitted under the Obstetrics and Gynecology service. The admission blood work showed hypochromic anemia, a CA-125 level of 105.4 U/ml (ref. <35) and air-fluid level on the abdominal x-ray. The chest x-ray, CEA, a-fetoprotein and b-HCG were within normal limits. The parasitic screening of vagina and feces as well as the urinalysis were negative. The intravenous pyelogram revealed ectasia of the left ureter with flattened calices, normal cortex without evidence of nephrolithiasis. The right excretory system was normal. These findings were attributed to extrinsic compression and possible infiltration from a pelvic mass.
The patient underwent diagnostic laparoscopy. The uterus and both adnexa were normal. Multiple biopsies of the ovaries and peritoneum as well as cytology and culture of the peritoneal fluid were obtained. All the results came back negative.

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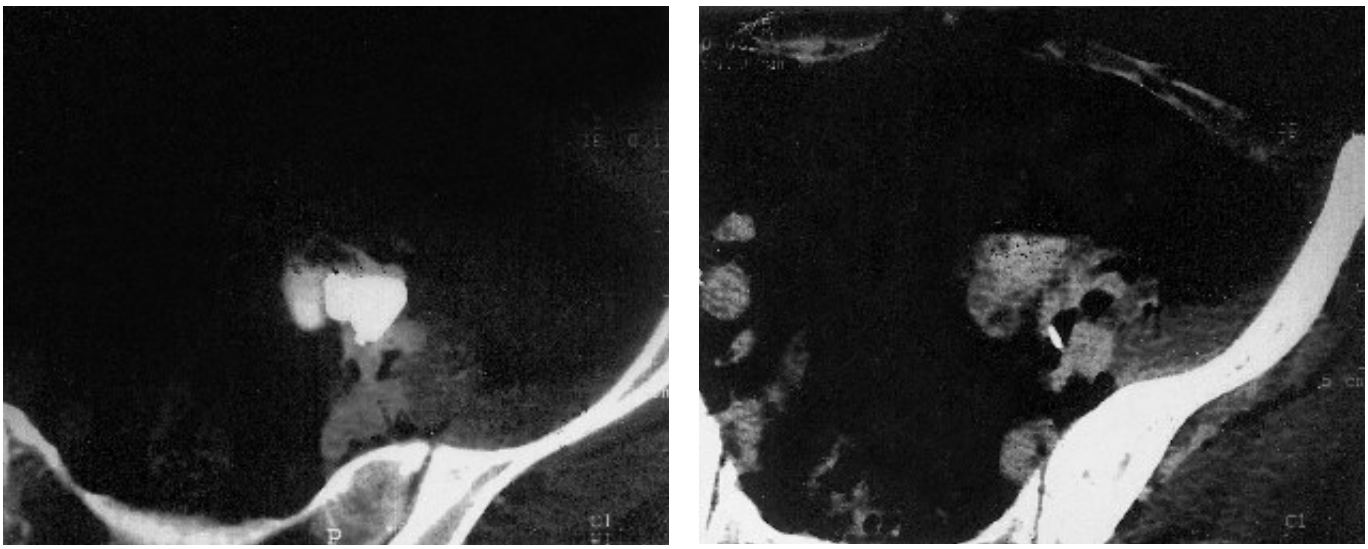

Fig. 1 a and b: Contrast enhanced CT scan confirmed the presence of ecstatic left ureter and contrast filled saccular structure representing either a diverticulum or megaloureter.

After the discharge the patient temporarily improved for few weeks, but then she had recurrence of the same symptomatology with progressive worsening.
After two months, due to the severe compromise of her overall condition with weight loss, dyspnea and ascites, the patient was admitted to a local Division of Obstetrics and Gynecology. A computed tomography was again obtained the main findings of which were: ascites, marked mesenteric edema, moderate ectasia of the left ureter to its proximal third of the pelvic portion associated with hydronephrosis. In the left pelvis was again demonstrated a 3 cm heterogeneous hypodense mass probably arising from the left adnexa. The distal ureter was normal in caliber. Once again no lymphadenopathy was demonstrated.
Subsequently the patient was transferred to our Division of General Surgery. On admission the patient was emaciated, deconditioned. Her abdomen was distended, moderately tender on deep palpation in the left lower quadrant and hypogastrium.
The admission blood work was significant for a moderate leukocytosis (12,000) with 95% neutrophils and CA-125 of 404 U/ml.
The abdominal ultrasound showed free peritoneal fluid, megaloureter with non-specific intraluminal echodensities, normal iliac vessels on the right but not visualized on the left as well as lack of visualization of the inferior vena cava. An additional CT scan of the abdomen and pelvis was obtained which confirmed the presence of intraperitoneal free fluid extending around the liver and spleen, retroaortic left renal vein, ecstatic left ureter (anteroposterior diameter 2.5cm) and contrast filled saccular structure behind the urinary bladder representing either a diverticulum or megaloureter. The pre intramural portion of the left ureter was not clearly visualized. (Fig. 1 a, b).

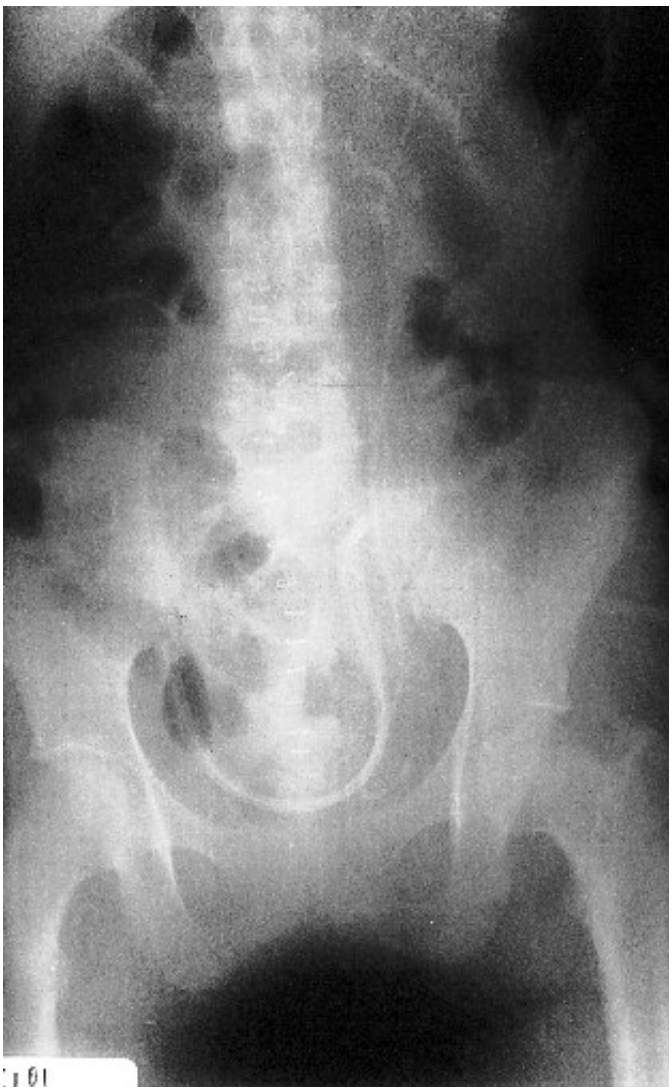

Fig. n. 2: Abdominal X ray on post-operative day 4.

The patient was then taken to the operating room for exploratory laparotomy. After a midline incision was made, 300 cc of serous fluid was aspirated. There was significant inflammatory reaction in the pouch of Douglas. Furthermore a nut size soft mass was noticed in correspondence to the crossing of the left ureter and the iliac vessel. After mobilization of the left colon and sigmoid, an inflammatory mass was unroofed from the retroperitoneum and urine like fluid was coming from it. The left ureter was isolated and its distal part had a divertricular enlargement. After resection of a 4 cm segment of the ureter an end-to-end anastomosis was carried out over a pigtail stent with 4-O monofilament suture. A closed drain was left in place (Fig. 2). The histological examination revealed a perforated ureteral diverticulum without evidence of malignancy (Fig. 3).
The patient was discharged to home on postoperative day 6. At her 40 month follow-up the patient was asymptomatic and the ultrasonographic and pyelographic evaluation confirmed the normal morphologic and functional status of the urinary tract (Fig. 4).

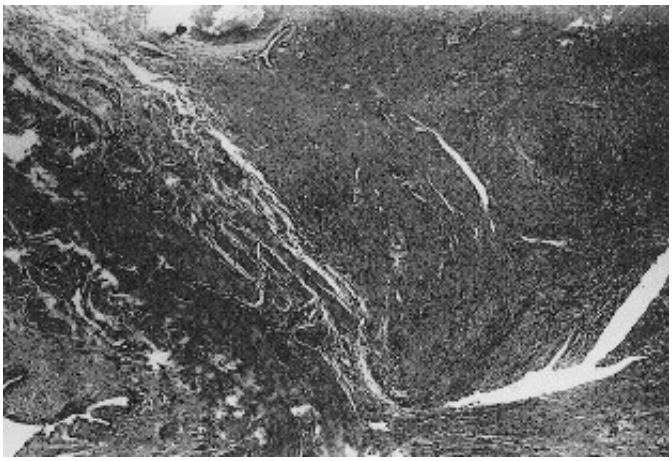

Fig. 3: Optical microscopy 50X, EE staining: fistulous tract in the ureteral periconnectival tissue lacking of epithelial lining and delineated by granulation tissue, fibrosis and recent hemorrhage (top left); ureteral wall and lumen (top right).

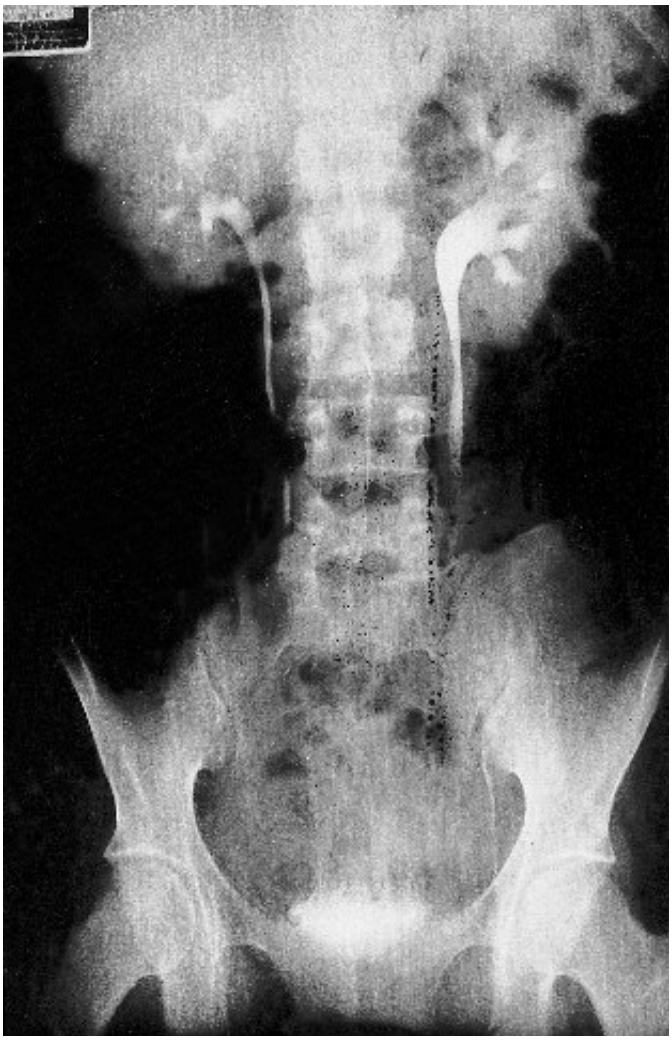

Fig. 4: 40 month follow-up: pyelography shows the normal morphologic and functional status of the urinary tract.

Discussion

The ureteral diverticulum is a rare pathology, affecting both sexes equally. Although it is more common in individuals 40 years old and above (1), it has been described in the pediatric population and even prenatally (2), which supports the congenital origin theory (1).
Although the diverticulum can affect the ureter at any level, the distal third is more commonly involved (8 out of 12 in the Mayo Clinic series), with the left side being 3 times more frequent and exceedingly rare the bilateral involvement (1). In 1947 Culp proposed a classification of the diverticula in congenital, in which all the layers of the ureter were present and directly communicating with it by a distinct opening and acquired, constituting simple mucosal protrusions (3).
Lawhon underlined the difference between the ureteral diverticula and the blind division of the ureter bifidus. The latter in fact is at least twice as long as its diameter and empties in the ureter on an acute angle (4).
Some Authors (5, 6) believe that the two entities share a common etiology: a very small ureter bifidus could change shape and size over time due to altered hydrodynamic states or concurrent pathologies (4, 7, 8).
In any case the solitary ureteral diverticulum has to be differentiated from both the pseudodiverticulosis and the multiple diverticula of the ureter. In the former the acquired sacciform or spicular extraflexion are made of mucosa only and are mostly present in the superior third of the ureter. Its pathogenesis has to be attributed to the epithelial hypertrophy deepening in the ureteral submucosa. The pseudodiverticulosis is frequently associated with transitional cell carcinoma and for such reason some authors consider it a precancerous condition requiring dedicated clinical and radiologic screening (9).

The multiple diverticula can be congenital or acquired, secondary to trauma, surgery or chronic ureteritis (10-11). The diverticulum is generally asymptomatic and the diagnosis is incidental during the work up or surgical intervention for different urogenital pathologies. In the Mayo Clinic series only two cases were symptomatic, one being caused by obstructive hydronephrosis due to a large infected diverticulum (1). In the other cases the diagnosis was incidental in patients with recurrent urinary tract infections with or without associated nephrolithiasis, prostatism, vescico-ureteral reflux, pyelonephritis or hydronephrosis due to a vascular malformation in a 14 year old female (12).

In the above reported case the clinical picture was quite complex and that explains the difficult diagnostic process. The symptomatology derived from a complication not described in the literature. The diverticulum became inflamed and after its perforation in the retroperitoneal cavity, fistulized into the peritoneum creating persistent uroperitoneum misinterpreted as malignant ascites from ovarian malignancy. In retrospect the chemical analysis of the ascitic fluid was consistent with urine mixed with inflammatory peritoneal fluid. The pathophysiology also explains why during the exploratory laparoscopy the gynecologic surgeon could not find any adnexal or intraperitoneal pathology but only free intraperitoneal fluid.

The rarity of the pathology made the first set of radiologists think that the ectasia of the ureter was secondary to extrinsic compression from adnexal pathology.

The absence of hydronephrosis and urinary tract infection did not warren the use of more specific diagnostic modalities such as retrograde pyelography or retroperitoneal laparoscopy, which have been reported not particularly helpful anyway in some series (12). The specific experience of the last radiologists was a key factor in the interpretation of the most recent CT scan helping in the formulation of the final diagnosis then confirmed by the intraoperative and histopathologic findings.

In conclusion, the knowledge of the multiform clinical manifestation of this rare entity facilitates the formulation of the differential diagnosis of unclear pictures of peritoneal and retroperitoneal (false acute abdomen) pathologies.

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Commento Commentary

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Con la definizione di addome acuto si intendono genericamente tutti i quadri clinici composti da segni e sintomi addominali tali da indurre ad un intervento in urgenza.
La disponibilità di indagini non invasive e di rapida esecuzione ha fatto sì che molti pazienti che nei decenni trascorsi sarebbero stati sottoposti ad interventi in urgenza, vengono attualmente prima sottoposti alle indagini necessarie a chiarire la natura della loro malattia.

Nonostante ciò è ancora possibile che una non corretta interpretazione dell'imaging comporti l'esecuzione di indagini invasive di inutili interventi o interventi inadeguati, esponendo il paziente al progredire della malattia ed alle sue complicanze.
Un caso esemplare è quello del falso addome acuto in una giovane donna riportato da Cappellani e Col. in cui fattori pregiudiziali come l'eccessiva valorizzazione data alla concordanza fra immagine di "neoformazione annessiale", "ascite" e innalzamento dei markers tumorali, ha condotto la paziente ad una inutile laparoscopia che non aveva né chiarito la diagnosi né impedito la progressione della malattia.
Interessante ed istruttivo il lavoro di Cappellani e coll. Induce a riconsiderare il work up preoperatorio e a rivedere la nostra condotta nella valutazione dei soggetti con addome acuto non accontentandosi della soluzione più probabile e tenendo presente, quando la prima ipotesi venga esclusa, anche soluzioni a prima vista eccezionali o mai descritte.
Anche in quest'ottica il lavoro in questione è di grande interesse essendo, a nostra conoscenza, il caso di perforazione spontanea di diverticolo ureterale con uroperitoneo, l'unico fin oggi descritto in letteratura. Esemplare descrizione e la dovizia di immagini lo rendono di grande interesse non solo per i chirurghi generali ma anche per radiologi ed urologi.

Generically the definition of acute abdomen points out all the clinical pictures composed by signs and abdominal symptoms that require the execution of an intervention in urgency.
The availability of investigation not invasive and of rapid execution has done so that many patients, that in the post decades would have undergone to interventions in urgency, currently are first submitted to the necessary investigations to clarify the nature of their illness.
Despite this it is still possible that a not correct interpretation of the imaging involves the execution of useless invasive investigations or inadequate interventions, exposing the patient to progress of the illness and to its complications.
An exemplary case is that of the false acute abdomen in a young woman brought by Cappellani and Coll. in which previously factors as the excessive exploration given to the agreement among image of «adnexal mass», «ascites» and raising of the tumor markers has conducted the patient to an useless laparoscopy that didn't have neither clarified the diagnosis neither prevented the progression of the illness.
Interesting and instructive the job of Cappellani and Coll. it induces to reconsider the preoperatory work up and to see our behavior in the evaluation of patients with acute abdomen not being satisfied with' the most probable solution and keeping in mind, when the first hipothesis is excluded, also solutions at first sight exceptional or ever described.
Also in this optics the report is of great interest being, to our knowledge, the case of spontaneous perforation of ureteral diverticulum with uroperitoneum the only one until today described in literature.

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