Unusual case of nephrocutaneous fistula — Our experience

Rajeev Thekumpadam Puthenveetil*, Debajit Baishya, Sasanka Barua, Debanga Sarma

Department of Urology, Gauhati Medical College Hospital, Guwahati, Assam, India

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Abstract A rare case of nephrocutaneous fistula due to spontaneous expulsion of renal calculi is described. A 45-year-old man presented with urinary leakage from an ulcer over the left lumbar region for the last 3 months after a history of spontaneous expulsion of stones from this area. Ultrasonography abdomen revealed a small contracted kidney with multiple calculi in the kidney and renal pelvis, sinus tract from the lower pole of the left kidney with a ruptured calyceal calculus in the sinus tract. CT urography revealed a non excreting left kidney with multiple renal calculi, with hyperdense collection in the renal parenchyma extending to the subcutaneous tissue and left lung suggesting a xenthogranulomatous pyelonephritis (XGP). We performed a left-sided simple nephrectomy with excision of the fistulous tract. Histopathological examination revealed XGP. There have been a few case reports of XGP forming nephrocutaneous fistula in the back.

1. Introduction

Xanthogranulomatous pyelonephritis (XGP) is an atypical, chronic inflammatory disease of the kidney. Although the pathogenesis of XGP is still unclear, the primary factors are urolithiasis, urinary tract obstruction and infection [1]. Proteus species and Escherichia coli are the organisms most commonly isolated in XGP [2]. Although the inflammatory process is usually diffuse and can extend beyond the kidney, nephrocutaneous fistula formation is a very rare presenting sign [3]. Gastrointestinal system, adjacent urinary organs and skin are the most commonly involved structures. Nephrectomy and primary excision of fistula is usually curative.

2. Case history

A 45-year-old male came with a history of intermittent discharge from an ulcer over the left-side of back since the
last 3 months. This was preceded with on and off pain in the
left flank, fever with chill and rigor for 6 months which
progressed to thinning and reddening of skin over left-side
of the back and sudden bursting out with expulsion of 2–3
small calculi, following which he got relieved of his symp-
toms leaving behind an ulcer over that region through
which clear urine leaked intermittently.

Per abdominal examination revealed two ulcers of size
1 cm × 1 cm and 1.5 cm × 2 cm on the left-side of the back,
just lateral to posterior axillary line, below the 12th rib. It
was pale red in color and did not bleed to touch (Fig. 1).
The margins were inverted and occasional discharge of
clear urine could be seen.

Laboratory investigation revealed a normal leukocyte
count (8.89 × 10^9/L), haemoglobin of 83 g/L. Urinalysis
showed the presence of more than 10 leukocytes per high-
power microscopic field, but no bacterial growth was
observed. Culture from the fistulous discharge showed no
growth.

Ultrasonography abdomen (USG–KUB) revealed a small
contracted kidney with multiple calculi in the left kidney and
renal pelvis, sinus tract from the lower pole with a ruptured
calyceal calculus in the sinus tract (Fig. 2). CT-IVU was done
which revealed a non excreting left kidney with multiple
renal calculi, with hyperdense collection in the renal pa-
renchyma extending to the subcutaneous tissue and the left
lung suggesting XGP (Fig. 3). Preoperative ulcer biopsy
showed increased granulation and collagenous tissue.

Nephrectomy was done by thoracolumbar incision
(Fig. 4). Intraoperatively, it was noticed that the renal fis-
tula extended to the lung parenchyma and skin. Fistulec-
tomy was successfully performed with excision of the
surrounding granulation tissue involving the renal and lung
parenchyma. A 24 Fr chest tube was placed and removed
subsequently after full expansion of the left lung on the 5th
postoperative day. Histopathological examination revealed
chronic granulomatous inflammation of the left kidney with
diffuse infiltration of lipid-laden histiocytes, consistent
with the diagnosis of XGP.

3. Discussion

XGP is a rare chronic inflammation of the kidney and con-
stitutes less than 1% of chronic pyelonephritis. It was first
described by Schlagenhauser in 1916 [4]. It is characterized
by destruction of renal parenchyma and its replacement
with granulomatous tissue, abscesses and collection of lipid
laden macrophages (xanthoma cells) [5,6]. Females are
more affected than males with a ratio of 1:4, and with a
mean age varying from 45 to 55 years [6,7]. Though the
exact mechanism of XGP is not clear, a number of predis-
posing factors have been implicated. The two most com-
mon predisposing factors are obstruction and infection of
the genitourinary system. Calculi are frequently staghorn
type (47%–100%). The organisms most commonly isolated in
XGP are Proteus species and E. coli [1,2]. Although many
XGP patients have shown pyuria, bacterial growth in their
urine has only been demonstrated in two-thirds probably
because urinary obstruction blocks contaminated urine to
reach the bladder. Rather, the unidentified organism could
be revealed by renal tissue cultures taken during surgery.

There are two forms of XGP. The diffuse or global form
(85%) is more common than the localized, focal or
segmental form (15%). Based on the severity of the disease
XGP has been described in three stages [8]. In stage I the
lesion is confined to kidney, in stage II the lesion extends to
Gerota’s fascia and in stage III extends to the paranephric space and other retroperitoneal structures.

Fistula formation is an unusual presenting sign in XGP. Gastrointestinal system, adjacent urinary organs and skin are the most commonly involved structures. There is a tendency of perirenal inflammation to spread superiority along the lines of fusion of renal fascial planes that tend to direct the exudates within the retroperitoneal compartment. The lumbocostal triangle, a relatively weak area of diaphragm, can transmit infection into the thoracic cavity which may cause nephrobronchial fistulas and lung abscess [9]. Cutaneous fistula occurs mostly into ipsilateral flank region, but unusual localizations such as knee were previously reported [10]. XGP presented with spontaneous expulsion of renal calculi is extremely rare.

XGP itself is an infiltrating disease and together with an obstructing stone in the renal pelvis may cause pyonephrosis and perinephric abscess. Later, it may burst out through the overlying skin.

Radiological diagnosis of XGP can be challenging as it is often difficult to differentiate it from primary renal neoplasms, pyonephrosis and other retroperitoneal mesenchymal tumors. USG–KUB is usually the first investigation during evaluation of such cases. CT is the imaging modality of choice as it not only suggests the diagnosis but also shows the extrarenal extent of the disease.

Nephrectomy and primary excision of fistula is curative in diffuse renal destruction [10].

**Conflicts of interest**

The authors declare no conflict of interest.

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