



A Case of Xanthogranulomatous Pyelonephritis with Nephropleural Fistula Formation: Role of MRI in Diagnosis and Treatment

황색육아종성 신우신염의 신장-흉강 누공 형성의 증례: 진단과 치료에서의 자기공명영상의 역할

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Xanthogranulomatous pyelonephritis (XGP) is a rare type of chronic bacterial nephritis, which rarely involves the invasion of adjacent organs or the formation of fistulas due to tissue-destructive granulomatous reactions. Although the invasions of various adjacent organs have been reported in several cases of XGP, MRI data on their features are limited. MRI has a better soft-tissue resolution than CT. Thus, it can identify the extent of extrarenal involvement in advanced XGP, and the findings can be used in treatment planning. Herein, we report a rare case of XGP with nephropleural fistula formation diagnosed using CT and MRI.

Index terms Pyelonephritis, Xanthogranulomatous; Magnetic Resonance Imaging; Fistula; Kidney

INTRODUCTION

Xanthogranulomatous pyelonephritis (XGP) is a rare but well-known type of chronic pyelonephritis with a destructive granulomatous process. In terms of imaging modalities, CT scan and MRI can reveal the characteristics of the condition and the extension of the lesion.

XGP has two morphologic subtypes, namely, the focal and diffuse forms, which may extend outside the kidney and can be complicated with fistula formation connecting the urinary tract with the proximate organs (1). The occurrence of fistulas connecting

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the kidney to the upper and lower gastrointestinal tract, bronchus, diaphragm, psoas muscle, and skin has been reported (2, 3).

Herein, we present a rare case of XGP with nephropleural fistula formation diagnosed via contrast-enhanced CT scan and MRI and successfully managed with surgery in a patient who presented with cough.

CASE REPORT

A 55-year-old woman with diabetes and hypertension visited a local clinic due to cough. Non-enhanced chest CT scan revealed enlargement of the left kidney with pelvocalyceal dilatation and consolidation with pleural thickening in the left lower lobe (Fig. 1A). She was then referred to our facility for further evaluations. The patient had an elevated white blood cell count at 12720/mL and C-reactive protein level at 0.87 mg/dL. Based on the urinalysis results, the white blood cell or bacteria count was not significant. CT urography revealed a signifi-

Fig. 1. Xanthogranulomatous pyelonephritis with nephropleural fistula formation in 55-year-old women.

A. Initial chest CT reveals peripheral consolidation with pleural thickening in the left lower lobe (arrowheads). An enlarged left kidney with multiple cystic areas is noted (arrow).

B. Coronal contrast-enhanced CT shows an enlarged left kidney with a multilocular appearance of dilated calyces and cortical thinning with diminished parenchymal perfusion. Staghorn calculi in the pelvocalyceal system obliterate the ureteropelvic junction (arrows), and the renal pelvis is contracted. An extrarenal infiltrative inflammatory lesion is noted (arrowhead).

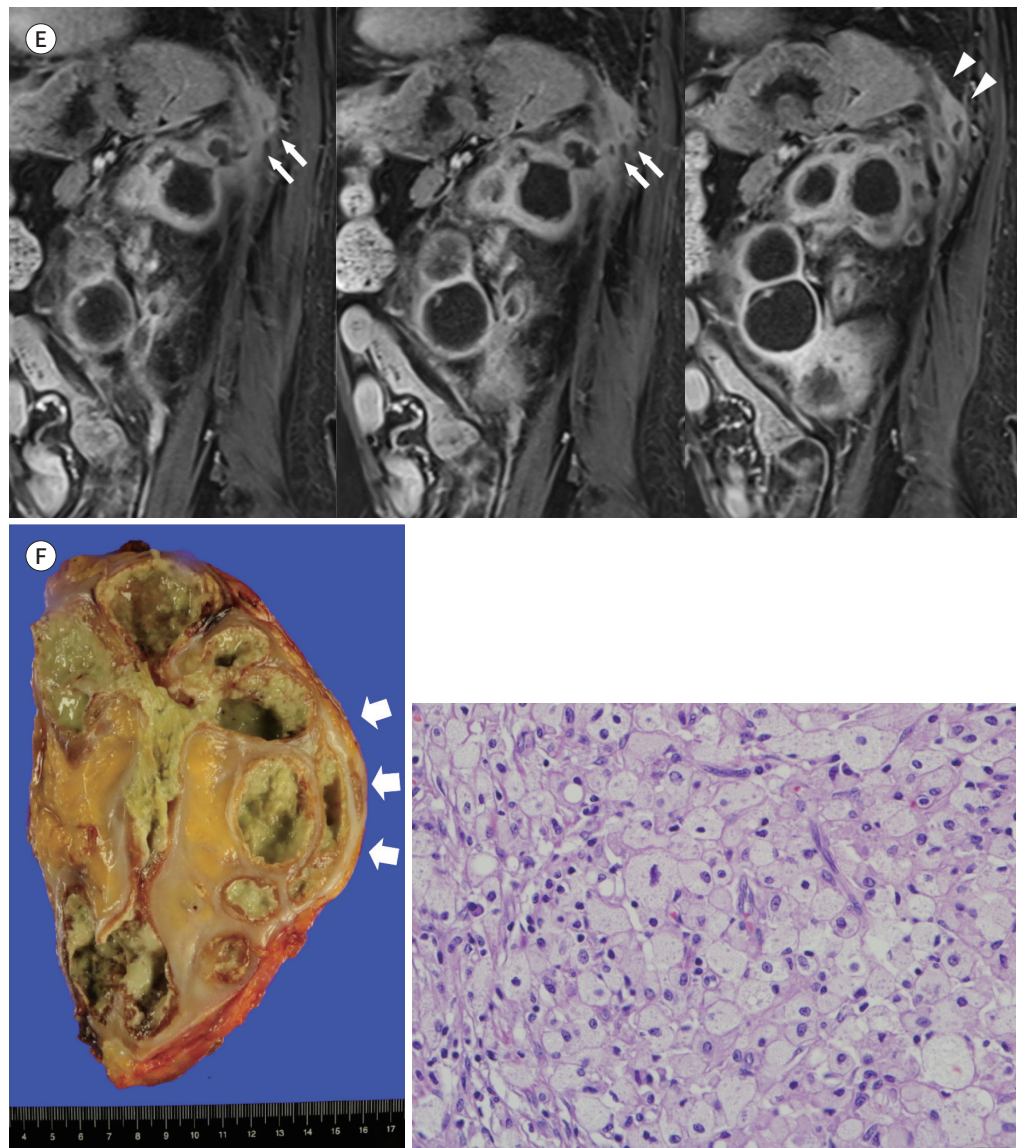
C. The extrarenal inflammatory lesion extends to the left pleural space via the left hemidiaphragm (arrows), resulting in empyema and consolidation in the left lower lobe (arrowhead). This CT scan image does not clearly show the fistula formation.

D. Sagittal contrast-enhanced fat-suppressed T1-weighted image shows a strong rim enhancement at the border of the cavitory lesions of the kidney. Perirenal inflammatory lesions and abscess (arrows) are shown.



cantly enlarged left kidney with diminished perfusion. Moreover, there were staghorn calculi in the pelvicalyceal system obliterating the ureteropelvic junction, contraction with thickening of the renal pelvis wall, and multilocular appearance of dilated calyces with cortical thinning, and these findings were consistent with XGP (Fig. 1B). Abscess and inflammatory lesion were characterized by a lobulated cystic lesion with an enhancing rim and an adjacent enhancing soft tissue lesion in the upper perirenal space, respectively. The inflammatory le-

Fig. 1. Xanthogranulomatous pyelonephritis with nephropleural fistula formation in 55-year-old women.
E. Serial sagittal contrast-enhanced fat-suppressed T1-weighted images show the small trans-diaphragmatic nephropleural fistula formation (arrows). Moreover, small empyema and atelectasis were identified (arrowheads).
F. Gross specimen shows a cystic appearance of the dilated calyces filled with pus and atrophied renal parenchyma. Extrarenal abscess formation within a thickened Gerota's fascia is shown in the left lateral aspect of the specimen (arrows). Photomicrograph reveals the microscopic features of chronic inflammation, including several xanthomatous histiocytes with foamy cytoplasm, neutrophils, lymphocytes, and plasma cells (hematoxylin & eosin stain, $\times 400$).



sion was suspected to infiltrate the left pleural space via the left hemidiaphragm, thereby resulting in empyema and consolidation in the left lower lobe (Fig. 1C). Nephropleural fistula was also considered. However, it was not clearly observed on CT scan.

For a more accurate diagnosis, contrast-enhanced MRI of the kidney was performed. A strong enhancement in the border of the cavitory lesions of the kidney and perirenal infiltrative inflammatory lesion was evident (Fig. 1D). Moreover, an extremely small fistula tract between the cystic area of the left kidney and the left pleural space via the left diaphragm was identified (Fig. 1E). The surrounding perirenal infiltrative inflammatory lesion had a strong enhancement. Thus, based on the abovementioned findings, the patient was diagnosed with XGP with trans-diaphragmatic nephropleural fistula formation in the left kidney.

The surgeons planned to conduct an elective radical nephrectomy. However, as the trans-diaphragmatic fistula was extremely small on MRI, diaphragm repair was not considered. Then, the use of oral broad-spectrum antibiotics (Cefditoren pivoxil) was initiated, and the drug was continuously administered for 3 weeks before surgery. She ultimately underwent a successful left laparoscopic radical nephrectomy. During the surgery, it was found that the perinephric inflammation and adhesion were extremely severe. Thus, the nephropleural fistula was difficult to identify, and kidney dissection was challenging to perform.

The pathology was XGP with lipid-laden macrophages (xanthoma cells), giant cells, and plasma cells (Fig. 1F). The patient did not present with significant postoperative complications, and she then recovered from the symptoms and was discharged 9 days after the surgery.

DISCUSSION

To date, there have been no reports on the MRI appearances of XGP with nephropleural fistula. In our case, the extrarenal extension of XGP was observed on CT scan, however, the nephropleural fistula was not obvious. Further evaluation with contrast-enhanced MRI provided an additional information about the small fistulous tract and adjacent anatomy. This finding indicates that MRI is superior to CT scan in identifying precise lesion characteristics. Since extrarenal extension of the disease is important in surgical planning, MRI is considered better than CT scan in visualizing the actual extent of XGP.

XGP is a chronic and severe inflammatory disease, which is a subset of pyelonephritis, and it results in the destruction of the renal parenchyma and complete loss of function in the affected kidney. It is more common in middle-aged women. The symptoms are often nonspecific, which include malaise, low-grade fever, flank pain, and weight loss (4). Moreover, the condition is often caused by prolonged renal infection due to obstructive uropathy, such as staghorn calculi or renal calculi, in several sites of the urinary tract. The commonly isolated organisms are *Proteus* species and *Escherichia coli* (5).

According to radiologic findings, XGP is classified into two, namely, the diffuse and focal forms, which account for 85% and 15% of cases, respectively (6). Diffuse XGP is staged as follows: Stage I is limited within the kidney. Stage II extends to the renal pelvis or the perirenal fat within the Gerota's fascia, and stage III extends beyond the Gerota's fascia into the other organs, retroperitoneum, or both. Extrarenal inflammation may invade and spread to adjacent structures, the most common of which are the gastrointestinal tract, urinary tract, and

skin. Involvement of the diaphragm, psoas muscle, small bowel, duodenum, colon, and lung with fistula formation has also been observed (7, 8).

To obtain a definite diagnosis of XGP, histopathological examination and assessment of lipid-containing macrophages (xanthoma cells) in tissue specimens are required. However, the use of classical imaging findings can help obtain a noninvasive diagnosis. CT scan remains the imaging modality of choice for XGP. The combination of a non-functioning enlarged kidney with loss of normal kidney contour, contracted renal pelvis with dilated collecting systems (bear paw sign) with or without central calculus, and inflammatory changes in the perirenal area are indicative of XGP (4, 8). Extrarenal inflammation with abscess formation and involvement of the adjacent organs can also be observed. On MRI, morphological features are equally displayed. As inflamed tissues have a hypervascular appearance, a bright enhancement of not only the borders of the cavities but also the extrarenal extension, such as thickened perirenal fascia, can be identified (1). Not only these MRI findings, but also nephropleural fistula was seen in our patient.

The therapeutic plan for XGP is based on the extent of the disease. Radical or partial nephrectomy is the treatment of choice. However, when diagnosed at an early stage, focal XGP can be treated with antibiotics in some cases (9). Advanced-stage XGP or that complicated by the involvement of other organs may require subsequent repair. Baydarian and Ludwig (10) have reported a patient with diffuse XGP involving the left diaphragm and left lower lobe of the lung, and the condition was treated with nephrectomy, partial diaphragmatic resection, diaphragmatic fistula repair, and left lower lobectomy. In our case, since the trans-diaphragmatic nephropleural fistula was small on preoperative MRI, the surgeon did not perform additional diaphragmatic repair. The use of MRI can help in a more accurate identification of the disease, and the findings are useful in establishing a surgical plan.

In conclusion, nephropleural fistula formation in XGP is a rare complication. CT scan is still considered the primary imaging modality for XGP, however, MRI can help in a more accurate identification of the extent of extrarenal involvement and fistula formation in other organs. Moreover, the findings of this imaging modality are useful for physicians in making treatment plans, including surgery.

Author Contributions

Conceptualization, all authors; investigation, L.G.Y.; project administration, L.J.W., M.S.K.; supervision, L.J.W.; writing—original draft, L.G.Y.; and writing—review & editing, L.J.W., Y.M.

Conflicts of Interest

The authors have no potential conflicts of interest to disclose.

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황색육아종성 신우신염의 신장-흉강 누공 형성의 증례: 진단과 치료에서의 자기공명영상의 역할

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황색육아종성 신우신염은 드문 형태의 만성세균성 신감염으로, 조직 파괴적인 육아종성 반응으로 인해 드물게 인접한 장기를 직접적으로 침범하거나 누공을 형성할 수 있다. 황색육아종성 신우신염이 인접 장기를 침범한 다양한 경우가 보고되어왔으나 자기공명영상의 보고는 매우 드물다. 자기공명영상은 전산화단층촬영보다 우수한 연부조직 해상도를 가져 황색육아종성 신우신염의 파급 범위를 정확히 파악하여 치료 계획을 세우는 데 도움이 될 수 있다. 우리는 전산화단층촬영과 자기공명영상으로 황색육아종성 신우신염의 신장-흉강 누공 형성을 진단한 드문 증례를 경험하였기에 이를 보고하고자 한다.

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