



# A Case Report of Large Tailgut Cyst Located from the Perirenal to the Perivesical Spaces

콩팥 주위와 방광 주위에서 발생한 커다란 원장미부 낭종의 증례 보고

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Tailgut cysts are known to originate from the remnants of the embryonic hindgut. They occur exclusively in the retrorectal and presacral spaces. There have been limited reports of tailgut cysts occurring in the left perirenal space. The present case features a huge tailgut cyst extending from the right perirenal to the perivesical space. We believe that this case report will help to further elucidate the characteristics of perirenal and perivesical tailgut cysts.

**Index terms** Congenital; Cysts; Hamartoma; Tomography, X-Ray; Magnetic Resonance Imaging

## INTRODUCTION

Tailgut cysts are rare congenital lesions and occur in the retrorectal/presacral space. There are few tailgut cysts that occur in the perirenal space (1-3). Only three cases of tailgut cysts in the perirenal space have been reported. In addition, there is no previous report of tailgut cysts occurring in the perivesical space. However, the present case features a tailgut cyst that located from the perirenal to the perivesical space. We believe that this case will help us to better understand perirenal and perivesical tailgut cysts.

## CASE REPORT

An 83-year-old female patient visited the hospital with right-leg swelling, which began 1 month prior. She was suspected of having deep vein thrombosis (DVT) and underwent lower extremity veno CT. Cystic mass was seen in the retroperitoneal space,

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and the right external iliac vein was compressed by the mass. DVT was not observed in the examination.

A huge multiloculated 30.0 cm × 11.1 cm × 11.4 cm cystic mass was observed in the right perirenal space, retrorenal plane, posterior pararenal space, along the psoas muscle, and the pelvic extraperitoneal space (Fig. 1A). The margins were smooth and had a lobulated contour. This cystic mass had a thick, uneven wall, and there was no enhancing solid component. The renal pelvis was slightly dilated due to the ureter compressed by the cystic mass (Fig. 1B), and the urinary bladder was deviated to the left (Fig. 1C). But there was no urinary symptoms.

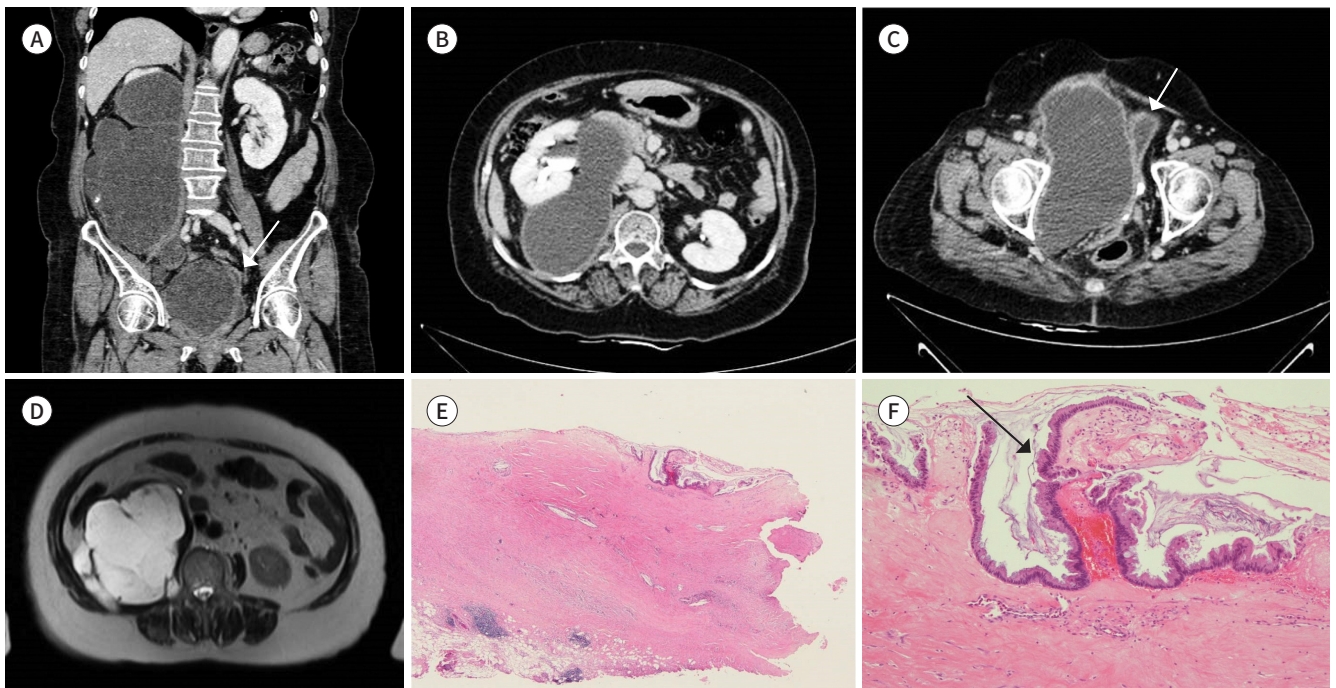
On abdominal MRI, this huge cystic mass was hypointense at T1-weighted images, hyperintense at T2-weighted images, and had a thick, uneven wall (Fig. 1D). There was no enhancing solid component or diffusion-restricted lesion. Therefore, this cystic mass mimicked a complicated cystic mass, such as a complicated lymphangioma.

The patient underwent retroperitoneal mass excision. However, there was severe adhesions around the cystic mass, so only a partial resection was performed.

The mass consisted of grayish white membranous soft tissue. The tissue was obtained in several pieces, and the largest piece was 3.5 cm × 2.0 cm × 0.2 cm. On microscopic pathology, a thick-walled cyst, smooth muscle bundle, and epithelial lining were observed (Fig. 1E, F). The tissue was fibrous and included the glandular mucinous epithelium, myxoid degenera-

**Fig. 1.** A large tailgut cyst located from the perirenal to perivesical spaces in an 83-year-old female.

- A.** The size of the cystic mass is 30 cm in diameter at its longest point and it is located from the perirenal to the perivesical space on CT (arrow indicates the urinary bladder).
- B.** The renal pelvis is slightly dilated due to the compressed ureter by the cystic mass, but the urinary passage is patent on CT.
- C.** The urinary bladder (arrow) is compressed by the cystic mass and deviated to the left side on CT.
- D.** T2-weighted image shows a multiloculated cystic mass with multiple septations and an uneven thick wall.
- E.** Photomicrograph shows a thick-walled cyst with smooth muscle bundles and lymphoid cells (× 2; hematoxylin and eosin stain).
- F.** Photomicrograph shows the cyst, lined by a ciliated columnar epithelium (arrow) (× 100; hematoxylin and eosin stain).



tion, and a cholesterol cleft, which are suggestive of a tailgut cyst. Some of the tissue had lymphoid aggregates, indicating inflammation. After surgery, the patient had improvement of her symptoms and she was discharged from the hospital.

This study was approved by the Institutional Review Board of our institution and the requirement for informed consent was waived (IRB No. E2021-038).

## DISCUSSION

The embryo has a true tail at about 28–35 days during the gestational process. This tail differentiates into many complex organs. After that, the tail completely regresses, but if a remnant remains, it can become a presacral tailgut cyst (4). A tailgut cyst forming in an unusual location, such as the perirenal space, is extremely rare, and it is assumed that the tailgut remnant moves along with the ascending kidney during development (1). In this case, the tailgut cyst spanned the perirenal and perivesical spaces. However, it is not clear where the origin was. Considering the main location of complicated cyst and no report on perivesical tailgut cyst, the cyst more likely originated from the perirenal space before extending into the pelvic extraperitoneal space.

There have been only three cases of tailgut cysts in the perirenal space. Two cases occurred exclusively in the perirenal space, and one case occurred concurrently in the perirenal and retrorectal spaces. These cysts were less than 20 cm in size, and they appeared on the left side (1-3). In contrast, present case showed huge cyst 30 cm in size occurring on the right side. Additionally, in previous cases, the main symptoms were abdominal pain or discomfort. In contrast, present case differs in that right-leg swelling was the main symptom, which was occurred by right iliac vein compression because of its unique location.

Generally, a retrorectal tailgut cyst is observed as a multicystic or multiloculated well-margined cystic mass with variable attenuation on CT. If there are no accompanying complications, tailgut cysts are observed to be homogeneously hypointense at T1-weighted images and hyperintense at T2-weighted images on MRI (2). The imaging features of the present case did not differ from other images of retrorectal tailgut cysts. However, the size and location were unique. Due to its large size, it had a mass effect on the right kidney and urinary bladder.

A differential diagnosis for perirenal tailgut cysts include retroperitoneal cystic tumors, such as cystic lymphangioma, cystic teratoma, and epidermoid cysts. Cystic lymphangioma typically appears as a large, thin-walled, multi-septated cystic mass (5, 6). Its attenuation varies from fluid to fat. An elongated shape and a crossing from one retroperitoneal compartment to an adjacent one is characteristic of the mass (5). A mature teratoma of the retroperitoneum manifests as a complex mass containing a well-circumscribed fluid component, adipose tissue, and calcification (7). Epidermoid cysts are commonly observed as thin-walled unilocular cystic masses of different fluid densities (8, 9), but their imaging findings are not specific. Other differential diagnoses include mucinous cystadenoma, schwannoma with cystic degeneration, and urinoma. It is difficult to distinguish tailgut cysts from other cystic masses through imaging. Therefore, histologic analysis is essential to establishing a definitive diagnosis of a tailgut cyst.

Malignant changes in tailgut cysts are rare, but some have been reported to occur. A num-

ber of adenocarcinoma and neuroendocrine tumors have been reported to arise from tailgut cysts. Nodular thickening of the cyst wall significantly increases the relative risk of the presence of cancer (10). In the present case, there was no evidence of cancer during the 3-year follow-up.

In conclusion, we believe that this case will help to further elucidate the characteristics of tailgut cysts occurring in unusual locations, such as in the perirenal and perivesical spaces.

#### Author Contributions

Conceptualization, K.D.M.; data curation, all authors; investigation, K.D.M., C.J.S., O.J.S.; project administration, K.D.M.; resources, K.D.M., O.J.S.; supervision, K.D.M.; visualization, O.J.S.; writing—original draft, O.J.S.; and writing—review & editing, all authors.

#### Conflicts of Interest

The authors have no potential conflicts of interest to disclose.

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## 콩팥 주위와 방광 주위에서 발생한 커다란 원장미부 낭종의 증례 보고

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원장미부 낭종은 태생기 원장미부의 잔유물에서 발생한다고 알려져 있다. 이들은 직장 후부 및 천골 전부에서 발생하는데, 좌측 콩팥 주위에서 발생한 증례가 매우 드물게 있었다. 이 증례는 우측 콩팥 주위에서 방광 주위까지 발생한 원장미부 낭종이다. 우리는 이 증례 보고가 콩팥 주위와 방광 주위에서 발생한 원장미부 낭종을 이해하는 데 도움이 될 것이라 생각한다.

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