Combined Xanthogranulomatous Urachitis and Bullous Cystitis
- A Case Report -

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The urachus is a vestigial remnant of the embryonic allantoic duct, which extends from the dome of the bladder to the umbilicus. The tubular urachus normally involutes before birth and remains as a fibrous band with no known function. However, persistence of an embryonic urachal remnant can give rise to various clinical problems, not only in infants and children, but also in adults. Urachal carcinoma and infected urachal cysts are the most frequently reported urachal lesions. Xanthogranulomatous urachitis is a very rare entity and only a few cases have been reported. We report here on a case of xanthogranulomatous urachitis with combined bullous cystitis and we discuss its histopathologic and ultrastructural features.

CASE REPORT

A 31-year-old woman presented with abdominal pain, urinary frequency and dysuria for 3 months, and all these problems had developed after a normal full term delivery. Physical examination detected a palpable mass in the suprapubic area. No abnormal findings were detected on the routine serologic findings. The urinanalysis showed a trace of proteinuria and hematuria. Cystoscopic examination showed bullous protrusions of the bladder mucosa (Fig. 1A). MRI of pelvic cavity revealed a single cystic mass with peripheral enhancement. The mass originated from the urachus and it involved the urinary bladder dome and it extended up to the umbilicus (Fig. 1B). The urachus, including the mass and a cuff of the bladder dome, was resected. The patient has been in good health with no recurrences for two years after the operation. Grossly, the cystically dilated urachus measured $7 \times 3 \times 2$ cm. The cyst was filled with yellowish greasy substance (Fig. 2A). Microscopically, the cyst contents consisted of xanthogranulomatous exudate. The walls of the urachus and the urinary bladder were infiltrated with inflammatory cells. The epithelial lining of the urachal remnant was lost. The main cells of the urachal mass were foamy histiocytes with a small proportion of lymphocytes and eosinophils (Fig. 2B). No organisms were seen on the Gram, GMS, and PAS staining. The xanthogranulomatous inflammation also involved the perivesical adipose tissue and the entire urinary bladder wall; this extend-
Fig. 1. (A) Cystoscopic examination shows bullous masses on the surface of urinary bladder (arrows). (B) Computed tomography reveals a cystic mass originating from the dome of the urinary bladder and extending to the umbilical level (between arrows).

Fig. 2. (A) The urachal mass extends from the dome of urinary bladder (arrow indicates partially resected bladder wall) up to the obliterated median umbilical ligament (arrowhead). (B) Sheets of foamy histiocytes and mixed inflammatory cells are noted in the xanthogranulomatous areas. (H&E, × 200) (C) Xanthogranulomatous cystitis. Xanthogranuloma splits the detrusor muscle layers and creates submucosal mass (arrows). Adjacent mucosa shows edematous bullous cystitis (arrowheads). (H&E, × 10) (D) Ultrastructural study reveals numerous histiocytes with electron lucent multiple droplets.
ed up to mucosal surface, causing xanthogranulomatous bullous cystitis with surface erosion (Fig. 2C). On the ultrastructural examination, multiple electron lucent multiple droplets without limiting membrane were noticed in the histiocytes (Fig. 2D). The size of the droplets was 0.5–2.0 \( \mu m \) in their maximum diameters.

**DISCUSSION**

Urachus-derived lesions can be categorized as congenital or acquired.\(^1\) A congenital patent urachus is derived either from the persistence of an open lumen or the failure of the bladder to descend into the pelvis. In patient with acquired disease, the urachus closes normally after birth, but it partially reopens under pathologic conditions. Acquired urachal disorders include umbilical urachal sinus, vesicourachal diverticulum, urachal cyst and alternating sinus.\(^1\) Urachal tract remnants that abnormally remain patent are subject to infection. These infected remnants are frequently confused with a wide spectrum of midline intra-abdominal or pelvic inflammatory disorders on clinical examination, or they are confused with malignant tumors.\(^2\)–\(^4\)

The present case describes typical histopathologic and ultrastructural features of xanthogranulomatous urachitis. Xanthogranulomatous lesions are rare forms of chronic mixed inflammatory processes that are characterized by the presence of lipid-laden histiocyte aggregation and mass formation. Such lesions have been described in many other sites such as kidney, urinary bladder, gallbladder, appendix, mandible, retroperitoneum, third ventricle, choroid plexus, orbit, vagina, lung, stomach, pericardium and ovary.\(^5\) Urinary tract involvement is well documented and most commonly as xanthogranulomatous pyelonephritis. Several cases of xanthogranulomatous cystitis have also been reported.\(^5\) Yet xanthogranulomatous involvement of the urachus has been rarely reported\(^6\)–\(^9\) and most of these articles discussed the clinical and radiological features without description of the histopathological or ultrastructural findings. According to those studies, the clinical and radiological findings of xanthogranulomatous urachitis were similar to urachal carcinoma and so making the correct differential diagnosis of solid urachal masses should be needed.

The ultrastructural features of xanthogranulomatous lesion are not well known. The greater the content of the unsaturated fatty acid and the higher the degree of unsaturation, the more electron dense lipid are seen because that lipid tend to better bind to osmium.\(^10\) Thus, the appearance of the lipid droplets in the present case suggests that these lipid droplets in xanthogranulomatous inflammation are formed of saturated fatty acids, which explains their resemblance to the neutral fat (triglyceride) droplets accumulated in the hepatocytes.

The pathogenesis of xanthogranulomatous inflammation is not well known. Defective lipid transport, immunological disorders, low virulent infections, reactions to specific infectious agents and lymphatic obstruction have been proposed as putative mechanisms.\(^11\) McVey and McMahon suggested that xanthogranulomatous inflammation is related to a longstanding or recurrent inflammatory process.\(^12\)

Additionally, the present case was combined with xanthogranulomatous bullous cystitis. Xanthogranulomatous cystitis is also rare. It was first described for in 1932 by Wassiljew and since then only about 20 cases have been reported.\(^13\) The location was mainly at the dome of the urinary bladder or near the dome. To the best of our knowledge, there have been no reported cases of combined xanthogranulomatous urachitis and bullous cystitis.

In conclusion, xanthogranulomatous urachitis should be included in the differential diagnosis of urachal masses. Because of the lack of specificity for radiological methods used in making the differential diagnosis of solid or cystic urachal masses, careful pathological study is very helpful to optimize the surgical approach and to avoid unnecessary radical surgery.

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