Case Report

Xanthogranulomatous cystitis masquerading as bladder tumor in a child

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ABSTRACT

Xanthogranulomatous cystitis affecting the urinary bladder is extremely rare, and only around thirty adult cases and two pediatric cases have been reported in the literature. The treatment is predominantly surgical as the lesion is mostly infiltrative and mimics malignancy. We report probably the third pediatric case, who presented with symptoms of urinary tract infection and urinary retention and was initially suspected as bladder tumor on imaging. The diagnosis was confirmed on histopathology, and the child responded well to aggressive antibiotic therapy alone.

INTRODUCTION

Xanthogranulomatous cystitis (XGC) affecting the urinary bladder is extremely rare, and only around thirty adult cases and two pediatric cases have been reported in the literature so far.[1] Conventionally, the treatment is surgical as the lesion is mostly infiltrative and mimics malignancy. We report the third pediatric case, who was initially suspected as bladder tumor, but he responded well to aggressive antibiotic therapy alone.

CASE REPORT

A 4-year-old male presented with straining during micturition, poor urinary stream, dysuria, and suprapubic pain for 3 months. After acute urinary retention, a week earlier, a bladder catheter had been placed. There was no history of fever, constipation, hematuria or weight loss or umbilical discharge. The general examination was unremarkable. On abdominal examination, a firm pelvic mass with an irregular border was palpable; per-rectal examination revealed a firm mass 2 cm from anal verge abutting on the right hemicircumference. The lumen was compressed, and the cranial extent was not ascertainable. Ultrasonography showed a thickened urinary bladder wall, with hypoechoic areas close to the bladder neck. Contrast-enhanced computed tomography scan confirmed a globally thickened urinary bladder (12 mm), with mucosal irregularities and bilateral hydroureteronephrosis [Figure 1a]. Laboratory investigations (leukocyte count - 17,100/cumm, urine microscopy - 30–35 pus cells/hpf, urine culture - *Escherichia coli* sensitive to piperacillin with tazobactam, co-trimoxazole, and nitrofurantoin) confirmed a urinary tract infection (UTI). A clinical diagnosis of bladder or – prostate rhabdomyosarcoma, bladder outlet obstruction, and urinary infection was made. He was initiated on intravenous piperacillin and tazobactam, and a trucut biopsy of the mass was obtained per rectally. The histopathology was a surprise. There were multifocal aggregates of foamy histiocytes admixed with epithelioid histiocytes and eosinophils. Few Touton type giant cells with focal areas of perivascular lymphocyte infiltration and fragments of fibrocollagenous tissue and smooth muscle bundle were noted [Figure 1b]. There were no well-formed granulomas, necrosis, or atypical cells. Special stains (acid–fast bacilli [AFB], periodic–acid

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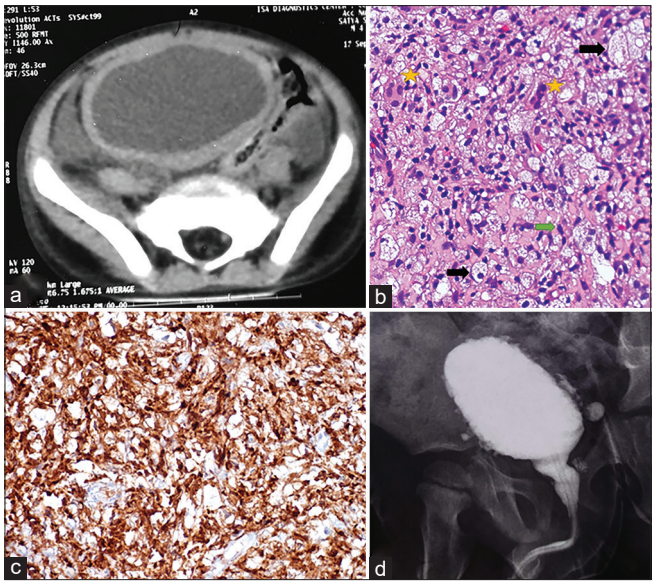

Figure 1: (a) Contrast-enhanced computed tomography (axial view) showing thickened bladder wall with mucosal irregularities and an ill-defined soft tissue mass posterolateral to it. (b) Photomicrograph showing foamy macrophages (black arrow), epithelioid cells (green arrow), and Touton type giant cell (black star) (H and E, ×400). (c) Immunohistochemistry showing diffuse histiocytes strongly positive for CD68. (d) Follow-up micturating cystourethrogram showing residual mucosal irregularity and multiple small diverticula in the bladder and prostatic utricular reflux

Schiff, Gomori methenamine silver) ruled out AFB or fungal organisms. On immunohistochemistry, histiocytes were diffuse and strongly positive for CD163 [Figure 1c] and CD68; smooth muscle actin, S100, synaptophysin, and CD1a did not identify tumor cells. A diagnosis of juvenile xanthogranuloma (JXG) was considered, and XG inflammation was the differential diagnosis. His symptoms settled with antibiotics, and the indenting mass was no longer palpable per rectally after 2 weeks. The dimercaptosuccinic acid scan showed bilateral preserved renal cortical function. After bladder catheter removal, he voided in a good stream with ease and was discharged on oral co-trimoxazole for another 2 weeks. At 6 months of follow-up, he was asymptomatic; the micturating cystourethrography showed residual irregularity of a part of the urinary bladder, multiple diverticulum, and contrast refluxing into the prostatic utricle [Figure 1d]. There was no vesicoureteral reflux or postvoid residue. A diagnostic cystoscopy was planned. However, as the child was asymptomatic and lived in a remote place, parents were not willing for further hospital admission.

DISCUSSION

JXG is a rare, self-limiting non-Langerhans cell histiocytic disorder. It is histologically characterized by benign-appearing histiocytes with or without lipid-laden multinucleated giant histiocytes (Touton giant cells). Limited to the first two decades of life, it usually presents as circumscribed cutaneous lesions that spontaneously regress in a few years. Extracutaneous manifestations of JXG are occasional.[2] The described clinicoradiological "mass" had Touton types of giant cells and immunopositivity to CD68 and was initially labeled as a pelvic xanthogranuloma. A review of the details (no cutaneous lesion, resolution with intravenous antibiotics alone) led us to a revised diagnosis of Xanthogranulomatous Cystitis rather than Juvenile Xanthogranuloma.

XG processes are rare, aggressive inflammatory conditions. The characteristic pathologic feature is the lipid-laden (foamy) macrophages or histiocytic cells.[3] XG processes are common in pyelonephritis and cholecystitis; however, it has been described in other viscera too. Thirty cases of XGC have been reported in the published English literature.[1] Most (85%) were adults and only two pediatric cases are documented.[1,4] The median age of presentation was 45 years without predilection for either sex. Most were located on the dome of the urinary bladder (85%) and associated with a urachal remnant (70%).[5] Abdominal pain and lower urinary tract symptoms including dysuria, frequency, and urgency were the common presenting symptoms. A few had hematuria or an umbilical discharge.[4,5] To the best of our knowledge, this is the youngest child with the entity reported till date.

The etiology of XGC remains unclear – chronic infection and inflammation with associated urachal remnant, immunological mechanisms, foreign body reaction, reaction to a local tumor, and abnormal lipid metabolism have been proposed.[1,4,5] Chronic Gram-negative UTI was in evidence here; no neoplasia was identifiable at histopathology. Like others,[3] we noted imaging findings of heterogeneous mass lesions with hypoechoic/hypodense areas. Yet, their asymmetric nature and deep pelvic location mimicked bladder/prostate neoplasia. Although benign, management options adopted for XGC have ranged from simple excision to partial or even radical cystectomy.[1,4,5] Medical management with antibiotics has been used as a postoperative adjunct earlier. In this case, the clinicoradiological resolution with antibiotics (intravenous for 2 weeks and oral for 2 weeks) alone suggests that the process was probably in the early stages following a chronic UTI.

CONCLUSION

XGC due to a chronic/recurrent UTI may mimic bladder/prostate neoplasia. Histology confirms the benign pathology, and medical management with appropriate antibiotics arrests the disease process in its nascent stages.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not

be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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