Case: Eosinophilic cystitis presenting as a bladder mass in an 11-year-old girl

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Introduction

We report the case of an 11-year-old girl who presented with acute urinary retention and microscopic hematuria. Pelvic imaging revealed a bladder mass for which sarcoma was initially suspected. Surgical biopsies revealed a massive eosinophilic infiltration of the submucosa confirming a diagnosis of eosinophilic cystitis. Clinical evolution was favourable without treatment. Eosinophilic cystitis prognosis is usually benign, but rare severe complications, such as complete bladder fibrosis requiring bladder augmentation or cystectomy, may occur.

Case report

Eosinophilic cystitis was first described by Brown in 1960[1] and about 50 pediatric cases have been described in the literature since. We report the case of an 11-year old girl with eosinophilic cystitis presenting as painless acute urinary retention associated with microscopic hematuria and favourable evolution without treatment.

The patient was referred to Grenoble University Hospital's emergency room by her primary care physician for a two-day history of suprapubic pain, dysuria, and microscopic hematuria. She had no past medical history or allergies. She did not report any regular or sporadic medication intake, history of travel to tropical areas, or familial atopy manifestations.
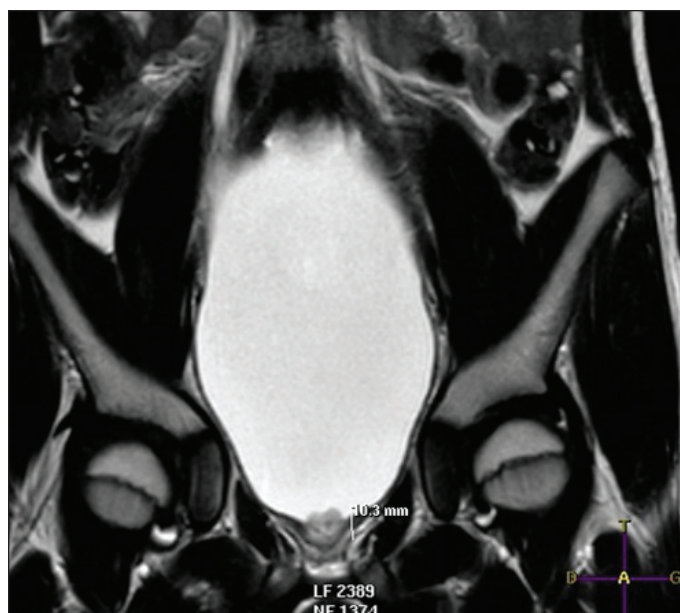
Symptoms were associated with acute fatigue, but she had no history of weight loss, diarrhea, fever, or rash. Clinical examination revealed an isolated, painless acute urinary retention without hepatosplenomegaly, fever, or cutaneous manifestations. Urinary labstix showed microscopic hematuria, automated urinalysis 10 000 red blood cells/mL, few white blood cells, and no eosinophiluria.

Complete blood count, blood eosinophils, C-reactive protein (<3 mg/L), and renal function were normal. Abdominal ultrasonography revealed a trigonal bladder mass of 10x10 mm with secondary urinary retention of 700 cc without upper urinary tract dilatation.

Urinary drainage by Foley catheter was performed and the patient was hospitalized. Alfa-foetoprotein, ß-human chorionic gonadotropin (hCG) and lactate dehydrogenase (LDH) bladder sarcoma markers were normal. Pelvic magnetic resonance imaging (MRI) showed a 10x10 mm bladder mass with a strong gadolinium-enhancing profile. Primary suspected diagnosis was rhabdomyosarcoma (Fig. 1). A positron emission tomography was performed and revealed intense fixation at the same anatomical site. No suspicious distant fixation foci were observed on this exam.

A cystoscopy under general anesthesia was performed and showed a flat edematous hypervascularized lesion with unclear borders localized at the bladder neck. Pathology colorations on biopsy tissue revealed a non-specific inflammatory infiltrate not allowing definitive diagnosis. Since rhabdomyosarcoma was suspected, close followup hospitalization with diagnostic laparotomy was scheduled seven days later.

A short suprapubic Pfannenstiel incision was performed, followed by a transvesical approach under cystoscopic control. The surgery was completed through pneumovesicoscopic approach allowing good and direct visualization of the lesion (Fig. 2). The pneumovesicoscopic approach revealed an edematous and hypervascularized lesion implicating the whole bladder neck. No induration or ulceration was seen. Six macrobiopsies, including the whole macroscopic lesion were taken. A bladder catheter was maintained until macroscopic hematuria resolution on Day 4 after surgery. A clamped suprapubic security catheter was also initially left in place. The patient's urinary function recovered completely without any post-void residue. Macroscopic biopsies showed an important eosinophilic infiltrate of the chorion and submucosa typical of eosinophilic cystitis (Fig. 3). Clinical evolution was favourable without specific treatment and no recurrence or compli-

Fig. 1. Pelvic magnetic resonance imaging. T2 injected sequence showing a gadolinium-enhancing bladder mass of 10.3 mm with urinary retention.

Fig. 2. Pneumo-vesicoscopy of the bladder neck showing a typical aspect of eosinophilic cystitis, with hypervascularized inflammatory edema in "medusa's head."

cations were observed at up to seven months' followup. Complementary infectious workup, including *Toxocara gondii* serology came back negative.

Discussion

Eosinophilic cystitis is a rare and exceptional condition, respectively, in adults and children. The median age at diagnosis in the pediatric population is 6.5 years old,[2] with a male to female ratio of occurrence at 3:1.[3,4] This case is therefore uncommon compared to the usual epidemiology. The clinical presentation is very diverse, usually associating dysuria, microscopic hematuria, and pollakiuria.[3,4] Active urinary tract infection is present in nearly half of the cases at time of diagnosis.[5]

Eosinophilic cystitis has been described in very diverse clinical contexts, such as postoperative, following a viral or parasitic infection, after intravesical instillation of mitomycin C, and during acute kidney stone disease, among others. It is therefore difficult to establish clear causal or risk factors for this condition.[2] In a recent review, association with other atopic features was only found in about 33% of patients.[4] An association with *Toxocara gondii* infection was described by Perlmutter and al, justifying a systematic serology.[6]
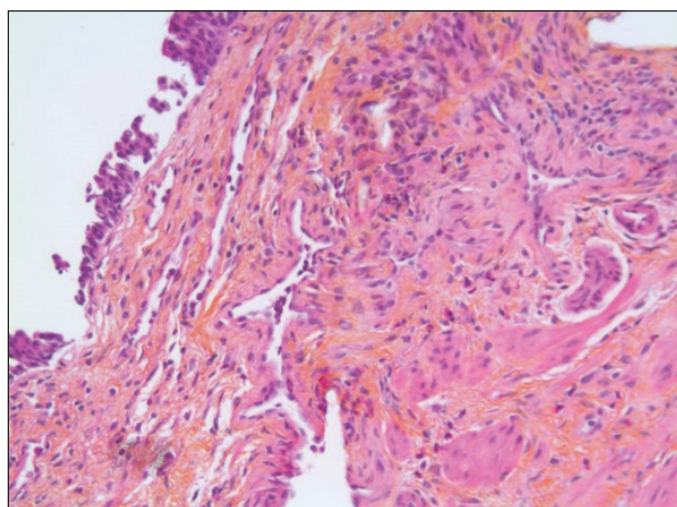
Our patient did not present any predisposing conditions or risk factors. About 40% of cases present with eosinophilia, which is an inconsistent finding, such as eosinophiluria.[7,8] The clinical and radiological presentation is frequently that of a bladder mass, resulting in initial high suspicion of in situ carcinoma in adults and rhabdomyosarcoma in children.[9,10] Widespread thickening of the whole bladder wall and uni-

lateral ureteral dilatation, present in one-third of cases, are other possible manifestations.[7]

Independent of the initial presentation, lesions are usually completely self-resolving. The initial malignancy suspicion often leads to a quick cystoscopy and biopsies, which are mandatory to establish the diagnosis.[3] Typical endoscopic aspects include submucosal hemorrhage, ulceration, edema, and hypervascularization,[8] which correspond to our patient's findings (Fig. 2). Proposed treatment algorithm by Ozdoğan in 2014 includes non-steroidal anti-inflammatories and anti-histamines as first-line agents, followed by corticosteroids in case of insufficient improvement after two weeks of treatment.[11] Alternatively, a "watchful waiting" approach, including clinical and ultrasonographic surveillance, was proposed

Fig. 3. Bladder mass biopsy showing important eosinophilic infiltrate of the submucosa on H&E colouration.

by Itano.[12] In this alternative algorithm, pharmaceutical treatment is reserved for cases in which symptoms persist after two weeks and after removal of potential antigenic stimuli (treatment of active urinary tract infection, antigen eviction if atopy is reported, treatment of toxocarosis).[12]

To our knowledge, no malignant evolution has been reported, but the frequency of recurrences and potential chronicity justifies close followup for at least one year after complete resolution.[7,8] Corticosteroids seem to be very efficient, allowing complete and sustained remission in two weeks compared to 8–10-week remission delays without treatment.[8] Immunosuppressive therapies, such as cyclosporine, have been proposed in exceptional cases of insufficient improvement under corticosteroids.[5,11] Ozdoğan proposes a regular followup, including clinical examination, ultrasonography, and control of initially elevated plasmatic eosinophil count. In cases of persistent ultrasonographic abnormalities under corticosteroid therapy, a control cystoscopy should also be performed. Several publications reported unfavourable evolution, such as complete bladder fibrosis with reduced compliance requiring aggressive surgical treatments, such as bladder augmentation or cystectomy.[5,7,12]

Conclusion

Eosinophilic cystitis is a difficult-to-recognize and probably underdiagnosed condition whose pathogenesis is still poorly understood. This case expands our knowledge and the existing literature with cystoscopy, radiology, and pathology visual material that may help urological surgery, radiology, and pathology teams when confronted with such atypical findings. This case also reinforces the need for endoscopic biopsies to confirm diagnosis and initiate optimal management of this rare pediatric pathology, which has good prognosis when optimally managed.

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