Inflammatory Myofibroblastic Tumor in Bladder with Multiple Vesicocutaneous Fistula in Pediatric Patient: A Rare Case Report

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Abstract

BACKGROUND: Inflammatory myofibroblastic tumor (IMT) is a rare tumor that occurs at any age from childhood to late adulthood and may have a slight male predilection.

CASE REPORT: A 7-year-old female presented with gross hematuria and a lump in the lower abdomen 1 month before admission. The complaints were also accompanied by weight loss, abdominal pain, and anemia. Ultrasound examination, Cystography, CT-Scan, and MRI showed that mass in the bladder. Histopathology examination confirmed diagnosis for IMT, thus, radiology plays an important role in supporting histologic examination for diagnosis and evaluation of IMT.

CONCLUSION: This case presents a 5-year-old girl with unspecific radiological findings that is later confirmed to be IMT by histopathological analysis. Diagnosis can be challenging due to nonspecific clinical presentation and variable anatomic locations of tumors. Clinicians need to consider IMT when patients present with a lower abdominal mass, as diagnosis is primarily by exclusion and confirmed by biopsy.

Introduction

An inflammatory myofibroblastic tumor (IMT) of the bladder is an uncommon benign tumor, which was initially reported by Roth in 1980 [1]. IMT is a rare neoplasm of mesenchymal origin, which presents as a benign mass composed of fibrous tissues and myofibroblasts, with marked infiltration of inflammatory cells, particularly plasma cells. IMT mainly appears in the lungs of children and adolescents; although it may also occur in any part of the body [2]. IMTs rarely occur in the urinary bladder. It is essential to distinguish this tumor from other malignant spindle cell tumors, such as the sarcomatoid variant of urothelial carcinoma and leiomyosarcoma. In a previous study, the patients are mostly young, with a mean age of 38.9 years old with a slight female predominance (51.7%) [3] although others also report male predominance of 9.8 [4]. The most common symptoms are hematuria, irritative or obstructive voiding, and lower abdominal pain [3], [5], [6]. Tumor size descriptions range from 1.5 to 13 cm [6]. IMT etiology is still unknown. It has been associated with prior history of surgery or instrumentation, trauma, and steroid use, but it remains uncertain due to its rarity [3], [5]. We report a case of IMT of the urinary bladder in a 7-year-old girl diagnosed by exploratory laparotomy with wide excision tumor and partial resection of the bladder.

Case Report

A 7-years-old Indonesian girl presented with gross hematuria and a lump in the lower abdomen one month prior to admission. The complaints were also followed by weight loss, abdominal pain, and anemia. There was no previous relevant medical history. The patient had undergone an ultrasound examination, CT-Scan, and biopsy in another hospital. After the biopsy, there were sores in the lower abdomen accompanied by urine that came out through the wound (Figure 1). The patient was then referred to our hospital. A blood test ordered at our hospital showed...
anemia (Hemoglobin 12.2 g/dL). On ultrasound, the pelvis ultrasound revealed a tumor of 7.17 × 5.90 cm in the right and front wall of the bladder (Figure 2).

Figure 2: Pelvic ultrasound showed a mass in the right anterior wall of the bladder. (a) transversal and longitudinal view. (b) Color Doppler examination showed vascularity within the lesion (arrow)

Pelvic CT imaging revealed a 6.1 × 8 × 9.5 cm tumor in the bladder, with peripheral enhancement (Figure 3). Cystography showed an asymmetrical polypoid mass with a filling defect at the anterolateral right bladder wall and multiple vesicocutaneous fistula at the infraumbilical area (Figure 4). Pelvic MRI showed polypoid mass 3.71 × 5.48 × 6.46 cm in the right anteroposterolateral bladder wall with low signal on T1-weighted images and heterogeneously high signal on T2-weighted images with infiltration to the surrounding area into the perivesical space rectus abdominis muscle, abdominal cutaneous dan subcutaneous tissue and obliteration of the opening of the right uterovesical junction, causing widening of the right ureter (Figure 5).

Surgery was performed 12 days after admission. Wide excision with partial resection of the bladder, reconstruction, cystostomy, and insertion DJ stent on right kidney was performed with a total duration of 150 min. No complication after surgery (Figure 6). A solid, polypoid mass was resected at the right posterior bladder wall, 5 × 6 × 3 cm in size. Histopathology examination revealed proliferative spindle cells arranged in haphazard patterns admixed with inflammatory cells (Figure 7). The immunohistochemistry study was partially positive for smooth muscle actin and desmin. Myogenin, MyoD1, and ALK were negative. The diagnosis was an IMT of the urinary bladder (Figure 8). One month after surgery, the patient was advised radiotherapy 25 times. No complication after radiotherapy. Evaluation after radiotherapy will be able after 3 months.

Discussion

Inflammatory myofibroblastic tumor (IMT) is a rare neoplasm of mesenchymal origin, which presents as a benign mass composed of fibrous tissues and myofibroblasts, with marked infiltration of inflammatory cells, particularly plasma cells. IMT commonly appears in children and adolescents’ lungs, but it can also appear in other parts of the body [2]. In children, IMT of the bladder is unusual [7]. IMT etiology is still unknown and IMT has been associated with prior history of surgery or instrumentation, trauma, and steroid use, but it remains uncertain due to its rarity [3], [5]. Clinical presentation of IMT depends on the organ in which they arise, but they are frequently associated with general inflammatory symptoms such as fever or malaise. In the bladder, hematuria and dysuria are the customary clinical manifestations [8]. The radiological appearance of IMT is unspecific, and they are often misdiagnosed as malignant neoplasms. Many of them are incidentally discovered when an imaging technique (computed tomography, ultrasonography, or magnetic resonance imaging) is performed for any other reason. Their most common radiological presentation is solid, irregular, well-defined masses (Table 1).

In this case, clinically and radiologically, IMT of the urinary bladder is indistinguishable from other entities, it has a broad differential diagnosis ranging from reactive to neoplastic malignant lesions, comprising post-operative spindle cell nodule, embryonal rhabdomyosarcoma, leiomyosarcoma, or sarcomatoid urothelial carcinoma [2], [9]. Differential
diagnoses for IMT of the urinary bladder are listed in Table 2. When compared to urothelial bladder carcinoma, the most frequent bladder tumor, IMT usually presents at younger age and spares the trigone [10]. Cystography demonstrates a filling defect on lesion, a specific diagnosis or even differentiation between benign or malignant tumors cannot be made with these techniques and additional imaging or evaluation is required for more definitive diagnosis. Ultrasound findings are unspecific; mostly show an isoechoic to hypoechoic intramural mass with a variable degree of vascularity on color Doppler evaluation. CT and MRI are the most useful imaging tools in the evaluation of IMT. On CT IMT can present as intraluminal polypoid or submucosal mass, with variable density and, usually, early peripheral enhancement [10], [11]. Perivesical fat stranding can also be present [11]. On MRI IMT exhibits low signal on T1-weighted images and heterogeneously high signal on T2-weighted images, enhancement characteristic is similar to those seen on CT.
The final and definitive diagnosis can only be made by histopathological examination and immunohistochemical or molecular study. Histopathological examination result is consistent with IMT.

Table 2: Differential diagnosis table for inflammatory pseudotumor of the urinary bladder

<table>
<thead>
<tr>
<th>Diagnosis</th>
<th>US</th>
<th>CT</th>
<th>MRI</th>
</tr>
</thead>
<tbody>
<tr>
<td>Inflammatory pseudotumor</td>
<td>Exophytic or polypoid bladder mass with variable echogenicity</td>
<td>Ranging from hypo to hyperattenuating lesion. Early peripheral enhancement. Central areas may have late enhancement</td>
<td>Hypointense on T1 and T2-weighted images. Early peripheral enhancement. Central areas may have late enhancement</td>
</tr>
<tr>
<td>Urothelial carcinoma</td>
<td>Papillary hypoechogenic masses or areas of focal wall thickening</td>
<td>Papillary or nodular mass or focal wall thickening. Tumoral calcification 5% of cases. Avid and early contrast enhancement</td>
<td>Masses with intermediate signal intensity on T2-weighted images. Avid and early contrast enhancement</td>
</tr>
<tr>
<td>Leiomyoma and leiomyosarcoma</td>
<td>Leiomyomas are smooth, solid, homogeneous masses. Leiomyosarcomas are more heterogeneous and infiltrative</td>
<td>Solid, homogenous or heterogeneous masses. Variable contrast enhancement</td>
<td>Intermediate signal intensity on T1-weighted images and low signal intensity on T2-weighted images. Leiomysarcomas are heterogeneous on T2-weighted images</td>
</tr>
<tr>
<td>Endometriosis</td>
<td>Wall thickening</td>
<td>Wall thickening. Endometriotic masses may enhance after contrast administration</td>
<td>Hemorrhagic foc with high signal intensity on fat-suppressed and non-fat suppressed T1-weighted images</td>
</tr>
<tr>
<td>Schistosomiasis</td>
<td>Wall thickening, calcifications, associated mass</td>
<td>Wall thickening, calcifications, associated mass</td>
<td>Wall thickening, associated mass</td>
</tr>
</tbody>
</table>

CT: Computed tomography; US: Ultrasound.
expressing mainly smooth muscle actin, desmin, and anaplastic lymphoma kinase (ALK-1) on immunohistochemical study. In our case, the smooth muscle actin and desmin were positive, myogenin and myoD1 were negative hence ruled out sarcomas based on previous diagnostic guidelines for sarcoma [12], [13], [14]. IMT was of neoplastic origin based on chromosomal rearrangement at 2p23 for those with ALK-1 positive staining. Overexpression of ALK shows more aggressive phenotype. We adapt the risk factor stratification by Ong et al. to (Figure 9) determine the treatment for the patient. Since in our case the tumor size was within 5 cm and ALK-1 staining was negative, we decided radiotherapy for the patient [15]. Surgical excision is the treatment of choice for patients with IMT, and chemotherapy is useful for treating those with non-resectable tumors [16], [17], [18]. Recurrence is seen in about 25% of cases. Distant metastases to the lungs, brain, liver, and bone are possible in <5% of cases (Figure 9).

**Conclusion**

Inflammatory myofibroblastic tumors (IMT) of the bladder in children are a rare occurrence and risk factors for this condition is yet to be discovered due to its rarity. This case presents a 5-year-old girl with unspecific radiological findings that is later confirmed to be IMT by histopathological analysis. Diagnosis can be challenging due to nonspecific clinical presentation and variable anatomic locations of tumors. Clinicians need to consider IMT when patients present with a lower abdominal mass, as diagnosis is primarily by exclusion and confirmed by biopsy.

**Ethical Clearance**

Informed consent from patient’s parent and the ethical clearance of this publication has been approved by the Ethical Committee of Dr. Hasan Sadikin Hospital.

**References**

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