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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

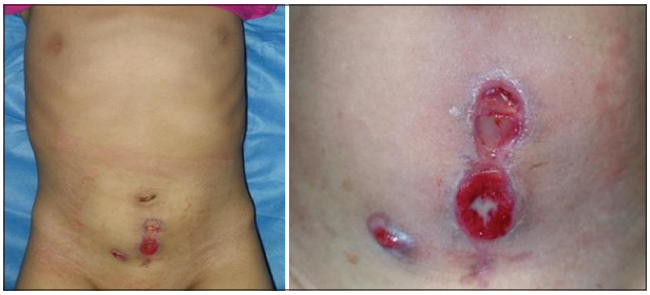

Figure 1: Lump and sores in the lower abdomen. Urine was leaking through the wound

anemia (Hemoglobin 12.2 g/dL). On ultrasound, the pelvic ultrasound revealed a tumor of 7.17 × 5.90 cm in the right and front wall of the bladder (Figure 2).

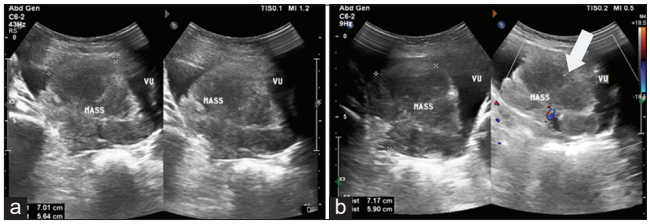

Figure 2: Pelvic ultrasound showed a mass in the right anterior wall of the bladder. (a) transversal and longitudinal view. (b) Color Doppler examination showed vascularity within the lesion (arrow)

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

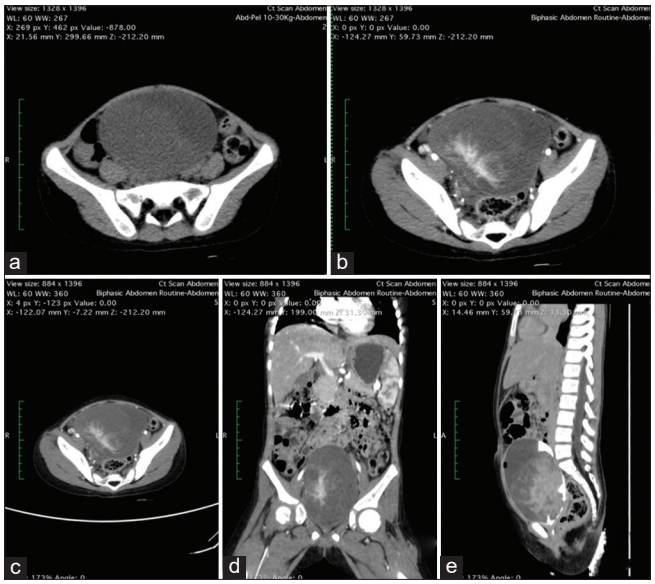

Figure 3: Pelvic CT-Scan with and without contrast media revealed enhancement within the mass (a and b). Axial, sagittal, and coronal view (c-e) pelvic CT-Scan showed an enhancing, heterogeneous tumor in the bladder

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, polyploid 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

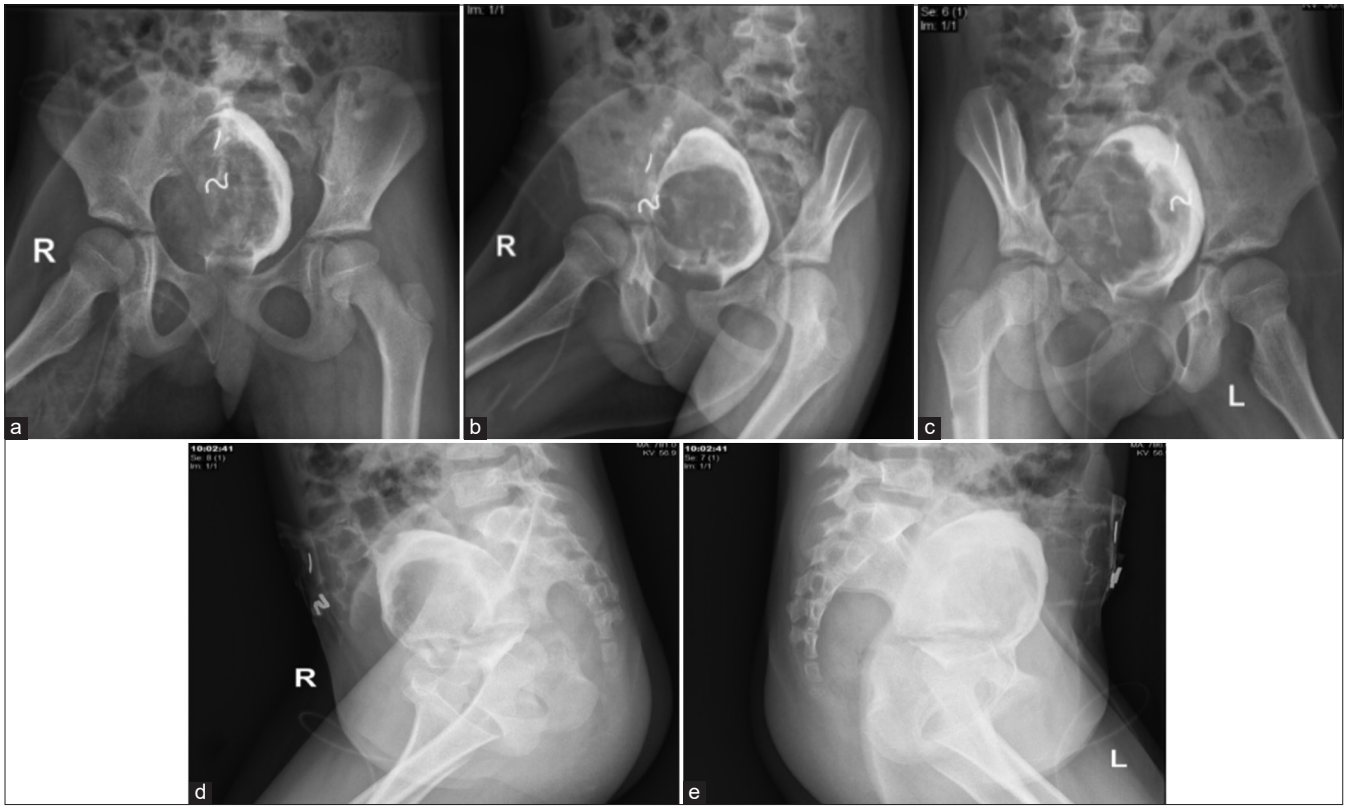

Figure 4: Cystography showed asymmetrical polypoid mass with filling defect at right anteroinferolateral side of the bladder wall in AP, right oblique and left oblique (a-c) and multiple vesicocutaneous fistula (arrow) at infraumbilical area with right and left lateral view (d-e)

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.

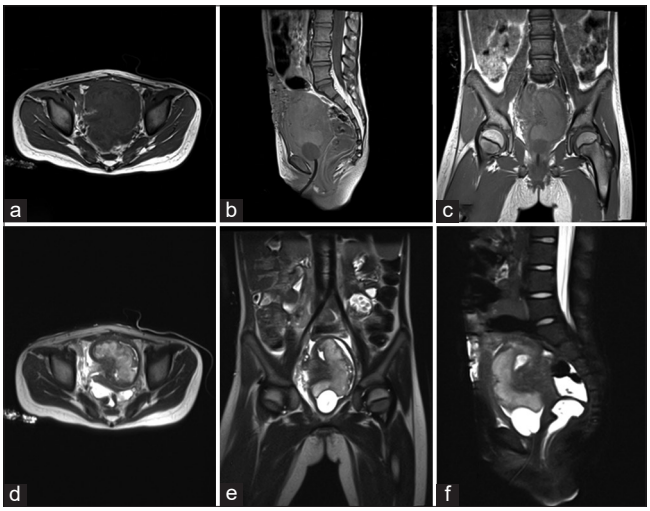

Figure 5: (a-f) Pelvic MRI showed polypoid mass at posterolateral side of bladder wall with low signal on T1-weighted images and heterogeneously high signal on T2-weigthed images. Infiltration of the surrounding area into the perivesical space, rectus abdominis muscle, and abdominal cutaneous dan subcutaneous tissue were present

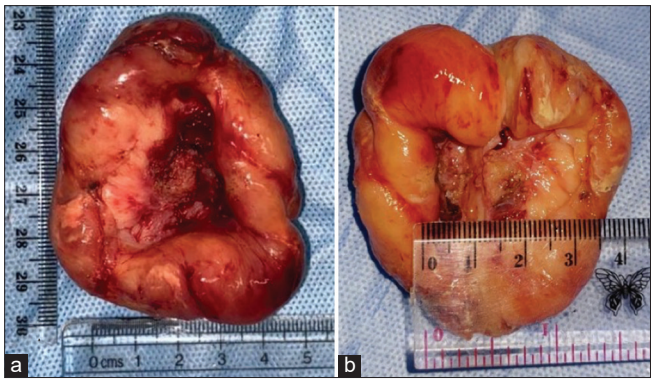

Figure 6: (a and b) Macroscopic appearance: A solid, polyploid mass 5 × 6 × 3 cm in size. The stalk, 2 cm in diameter, was connected to the right posterior bladder wall

Table 1: Symptoms, radiological findings, and differential diagnosis of the inflammatory myofibroblastic tumor's of the body [19]

Location	Symptoms	Imaging findings	Differential diagnosis
Orbit	Pain, redness, oedema, proposis Ptosis, Slalom: oar deficits, diplopia, swelling, mass-effect	Solid and heterogeneous enhancing. Low intensity on 12-weighted due to fibrotic composition. Associated retrobulbar fat or oedema	Granulomatous diseases. primary infection. sarcoid, Worn disease. Ornery and secondary tumours of the orbit, lymphoma and connective tissue diseases
Lung	Cough, chest pain, dyspnea, haeroomysis, fever, malaise and weight loss	Solitary well-circumscribed peripheral, predominance for the lower lobes, unusual calcifications. Heterogeneous or homogeneous enhancement pattern	Malignant lung masses: Primary bronchogenic carcinoma
Scrotum	Lump in the scrotum	Low intensity on 12-weighted images Solid and heterogeneous masses with internal vascularity on US. Well-defined, hypodense, tilde enhancement on CT. Small calcifications can be associated	Scrotum primary neoplasm
Hepatic	Unspecific, depending on the sine	Heterogeneous or peripheral enhanormem dining the arterial phase. Or homogeneous enhancement during the arterial phase and washout during the delayed phase on CT TI hypointense and T2 hyperintense with heterogeneous enhancement	liCC
Splenic	Left upper quadrant or cpiganrie pain	Low-density pattern. Low-intensity on the TI-and T2-weighted images. Low-intensity baton in the early phase alter gadolinium injection and high intensity in the delayed phase	Splenic hannangiome and other primary splenic neoplasms as lymphoma or picnic an mosarooma
Gastrointestinal tract and mesentery	Abdominal pain, intestinal obstruction dysphasia, anaemia	14p: dense, Heterogeneous, well-dclimited	GIST, inflammatory fibroid polyp, smooth muscle neoplasm peripheral nerve sheath tumour, solitary fibrous tumour. Fittromatosis, the follicular and whine cell sarcoma, lymphoma and admocareinoma
Musculoskeletal	Pain, oedema	Nonhomogencous, solid	Malignant lesions: Rabdomyosareana

CT: Computed tomography, US: Ultrasound.

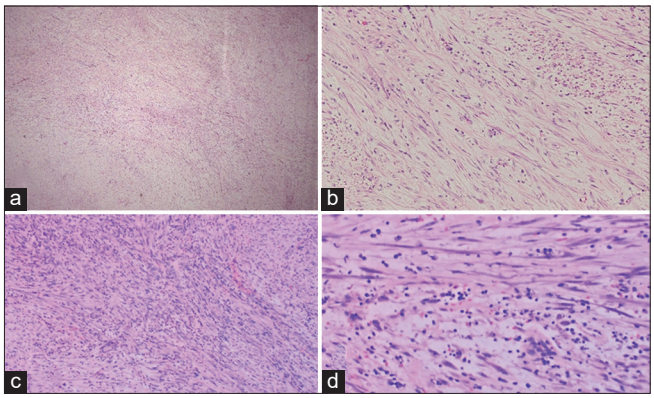

Figure 7: Hematoxylin-Eosin staining. The histologic feature revealed proliferative spindle cells arrange in haphazard pattern admixed with inflammatory cells. Objective 20× (a), 100× (b) and (c), 200× (d)

The final and definitive diagnosis can only be made by histopathological examination and immuno-histochemical or molecular study. Histopathological examination result is consistent with IMT. IMT

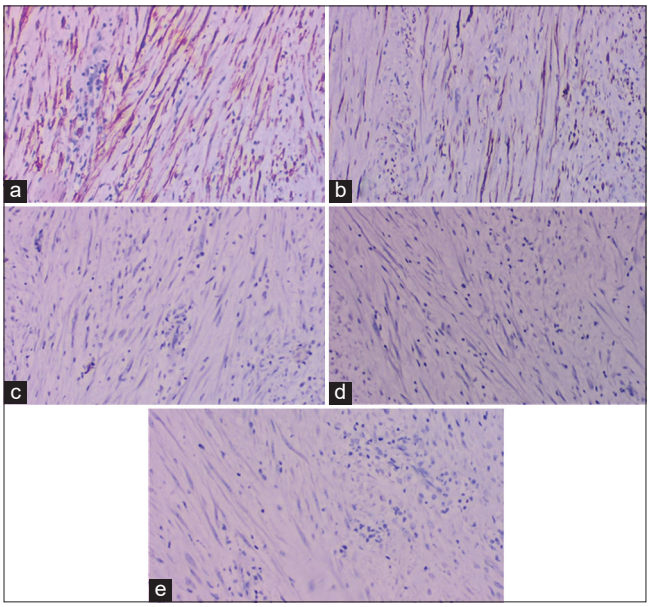

Figure 8: The immunohistochemistry staining was positive partial for smooth muscle actin (a), and desmin (b). myogenin, MyoD1, and ALK were negative (c-e)

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

Diagnosis	US	CT	MRI
Inflammatory pseudotumor	Exophytic or polypoid bladder mass with variable echogenicity	Ranging from hypo to hyperattenuating lesion. Early peripheral enhancement. Central areas may have late enhancement	Hypointense on Ti and T2-weighted images. Early peripheral enhancement. Central areas may have late enhancement
Urothelial carcinoma	Papillary hypoechoic masses or areas of focal wall thickening	Papillary or nodular mass or focal wall thickening. Tumoral calcification 5% of cases. Avid and early contrast enhancement	Masses with intermediate signal intensity on T2-weighted images. Avid and early contrast enhancement
Leiomyoma and leiomyosarcoma	Leiomyomas are smooth, solid, homogeneous masses. Leiomyosarcomas are more heterogeneous and infiltrative	Solid, homogeneous or heterogeneous masses. Variable contrast enhancement	Intermediate signal intensity on T1-weighted images and low signal intensity on T2-weighted images. Leiomyosarcomas are heterogeneous on T2-weighted images
Endometriosis	Wall thickening	Wall thickening. Endometriotic masses may enhance after contrast administration	Hemorrhagic foci with high signal intensity on fat-suppressed and non-fat suppressed TI-weighted images
Schistosomiasis	Wall thickening, calcifications, associated mass	Wall thickening, calcifications, associated mass	Wall thickening, associated mass
Tuberculosis	Diffuse irregular wall thickening. Reduced urinary bladder capacity in chronic phase	Thickened and contracted urinary bladder. Fistulas and sinus tracts. Ureteric dilatation	Thickened and contracted urinary bladder. Fistulas and sinus tracts. Ureteric dilatation

MRI: Magnetic resonance imaging, CT: Computed tomography, US: Ultrasound.

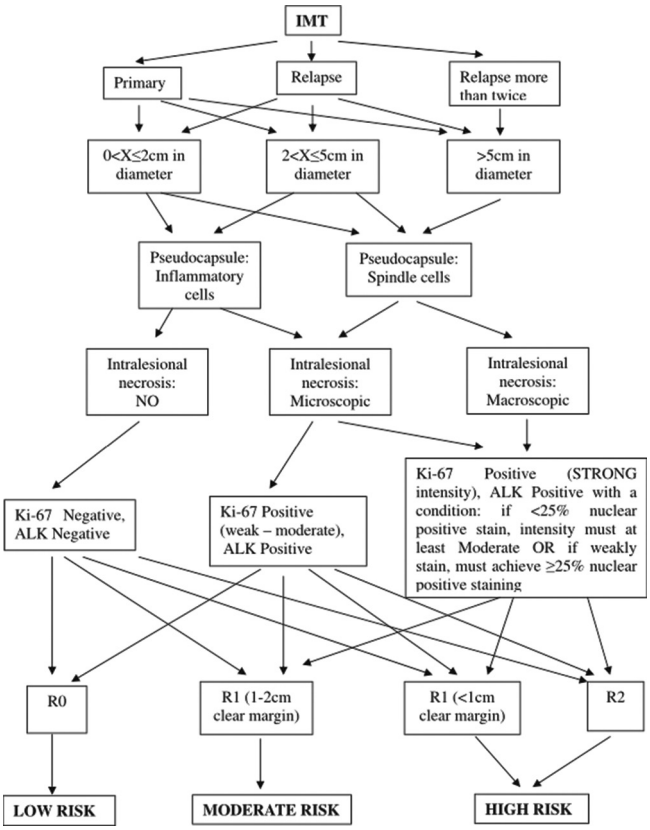

Figure 9: Risk factors stratifications for IMT [15]

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

1. Roth JA. Reactive pseudosarcomatous response in urinary bladder. Urology. 1980;16(16):635-7. https://doi.org/10.1016/0090-4295(80)90578-6
PMid:7445316
2. Ozsan I, Ozsoy M, Sahin E, Ortac R, Aydın U, *et al*. Inflammatory myofibroblastic tumor of the gallblader. Balkan Med J. 2016;30(3):323-6. https://doi.org/10.5152/balkanmedj.2013.8263
PMid:25207130
3. Teoh JY, Chan NH, Cheung HY, Hou SS, Ng CF. Inflammatory myofibroblastic tumors of the urinary bladder: A systematic review. Urology. 2014;84(3):503-8. https://doi.org/10.1016/j.urology.2014.05.039
PMid:23671533
4. Wei L, Jianbo L, Qiang W, Hai Y, Zhixiang L. Inflammatory myofibroblastic tumour of the bladder: Case report and review of the literature. Can Urol Assoc J. 2013;7(3-4):E237-40. https://doi.org/10.5489/cuaj.544
PMid:23671533
5. Kim HW, Choi YH, Kang SM, Ku JY, Ahn JH, Kim JM, *et al*. Malignant inflammatory myofibroblastic tumor of the bladder with rapid progression. Korean J Urol. 2012;53(9):657-61. https://doi.org/10.4111/kju.2012.53.9.657
PMid:23061006
6. Lott S, Lopez-Beltran A, Maclennan GT, Montironi R, Cheng L. Soft tissue tumors of the urinary bladder, Part I: Myofibroblastic proliferations, benign neoplasms, and tumors of uncertain malignant potential. Hum Pathol. 2007;38(6):807-23. https://doi.org/10.1016/j.humpath.2007.03.017
PMid:17509394
7. Patnana M, Sevrukov AB, Elsayes KM, Viswanathan C, Meghan L, Menias CO. Inflammatory pseudotumor: The great mimicker. AJR Am J Roentgenol. 2012;198(3):W217-27. https://doi.org/10.2214/AJR.11.7288
PMid:22358018
8. Song D, Jiao W, Gao Z, Liu N, Zhang S, Zong Y, *et al*. Inflammatory myofibroblastic tumor of urinary bladder with severe hematuria: A Case report and literature review. Medicine (Baltimore). 2019;98(1):e13987. https://doi.org/10.1097/md.0000000000013987
PMid:30608442

9. Süer E, Gülpınar Ö, Mermerkaya M, Burgu B, Celepli P, Sertçelik A, *et.al*. Inflammatory myofibroblastic tumor of the bladder in a 10-year-old girl. Urology. 2012;80:1138-40.
10. Rosado E, Pereira J, Corbusier F, Demeter P, Bali MA. Inflammatory pseudotumor of the urinary bladder. J Radiol Case Rep. 2015;9(1):36-42. https://doi.org/10.3941/jrcr.v9i1.2066
    PMid:25926919
11. Surabhi VR, Chua S, Patel RP, Takahashi N, Lalwani N, Prasad SR. Inflammatory myofibroblastic tumors: Current update. Radiol Clin North Am. 2016;54(3):553-63. https://doi.org/10.1016/j.rcl.2015.12.005
    PMid:27153788
12. Montgomery EA, Shuster DD, Burkart AL, Esteban JM, Sgrignoli A, Elwood L, *et al*. Inflammatory myofibroblastic tumors of the urinary tract: A clinicopathologic study of 46 cases, including a malignant example inflammatory fibrosarcoma and a subset associated with high-grade urothelial carcinoma. Am J Surg Pathol. 2006;30(12):1502-12. https://doi.org/10.1097/01.pas.0000213280.35413.1b
    PMid:17122505
13. Libby EK, Ellis LT, Weinstein S, Hammer RD, Murray KS. Metastatic inflammatory myofibroblastic tumor of the bladder. Urol Case Rep. 2018;23:10-2. https://doi.org/10.1016/j.eucr.2018.11.007
    PMid:30505686
14. Yagnik V, Chadha A, Chaudhari S, Patel K. Inflammatory myofibroblastic tumor of the urinary bladder. Urol Ann. 2010;2(2):78-9. https://doi.org/10.4103/0974-7796.65106
    PMid:20882160
15. Ong HS, Ji T, Zhang CP, Li J, Wang LZ, Li RR, *et al*. Head and neck inflammatory myofibroblastic tumor (IMT): Evaluation of clinicopathologic and prognostic features. Oral Oncol. 2012;48(2):141-8. https://doi.org/10.1016/j.oraloncology.2011.09.004
    PMid:22014665
16. Coffin CM, Alaggio R. Fibroblastic and myofibroblastic tumors in children and adolescents. Pediatr Dev Pathol. 2012;15(Suppl 1):127-80. https://doi.org/10.2350/10-12-0944-pb.1
    PMid:22420727
17. Ferrari A, Alaggio R, Meazza C, Chiaravalli S, Vajna de Pava M, Casanova M, *et al*. Fibroblastic tumors of intermediate malignancy in childhood. Expert Rev Anticancer Ther. 2013:13(2):225-36. https://doi.org/10.1586/era.12.180
    PMid:23406563
18. Kovach SJ, Fischer AC, Katzman PJ, Salloum RM, Ettinghausen SE, Madeb R, *et al*. Inflammatory myofibroblastic tumors. J Surg Oncol 2006;94(5):385-91. https://doi.org/10.1002/jso.20516
19. Cantera JE, Alfaro MP, Rafart DC, Zalazar R, Muruzabal MM, Barquín PG, *et al*. Inflammatory myofibroblastic tumours: A pictorial review. Insights Imaging. 2015;6(1):85-96. https://doi.org/10.1007/s13244-014-0370-0
    PMid:25519466