



Pediatrics

Hemangioendothelioma of the Bladder: The Youngest Case Report in A Child



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ABSTRACT

Hemangioendothelioma is a vascular tumor that commonly found in soft tissue, lungs, heart, liver, and bones, but it is very rare in bladder. We report a case of pediatric hemangioendothelioma of the bladder in a 2 years 7 months-old boy that treated with total excision of the bladder and bilateral ureterocutaneostomy. This case is the second case in pediatric patients, and the youngest case that reported in a child. Our patient is doing well post operatively. But unfortunately interferon a-2b is not available in our hospital. At 3 months follow up, there was progressive progression of the tumor. Recurrent Abdominal mass was confirmed by Abdominal CT-Scan. The patient was died one week later. Interferon a-2b is might be an effective regimen on this tumor but further study is needed to confirm this statement.

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Introduction

Hemangioendothelioma is a vascular neoplasm that was first identified in soft tissues by Weiss and Enzinger.¹⁻³ It is very rare found in the bladder.^{1,3}

We report the second case of hemangioendothelioma in pediatric patient.

Case report

A 2 years 7 months-old boy presented with painless, intermittent, gross hematuria, and mass in suprapubic region in 6 months duration. He was coming to hospital with severe malnutrition, kidney failure, and obstructive ileus.

Respiratory rate was 30 times/minute. The other vital signs were within normal limit. The abdomen was blunt and mass was palpable in suprapubic region. There was abscess in umbilical region. Laboratory findings showed a hemoglobin level of 5,0 g per deciliter, 23.300 white blood cells per microliter, 757.000 platelets

per microliter, blood urea nitrogen of 121 mg per deciliter, and creatinine level was 2,34 mg per deciliter with natrium level was 129 milliequivalents per liter and potassium level was 5,9 milliequivalents per liter.

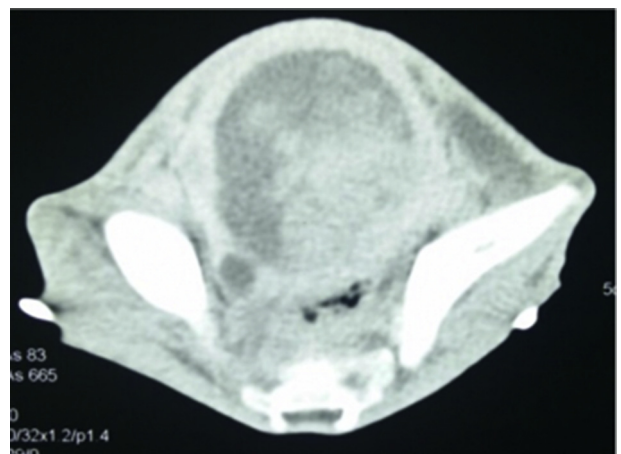


Figure 1. Computed tomography scan showing a large mass that almost fills all of the bladder with thickened wall.

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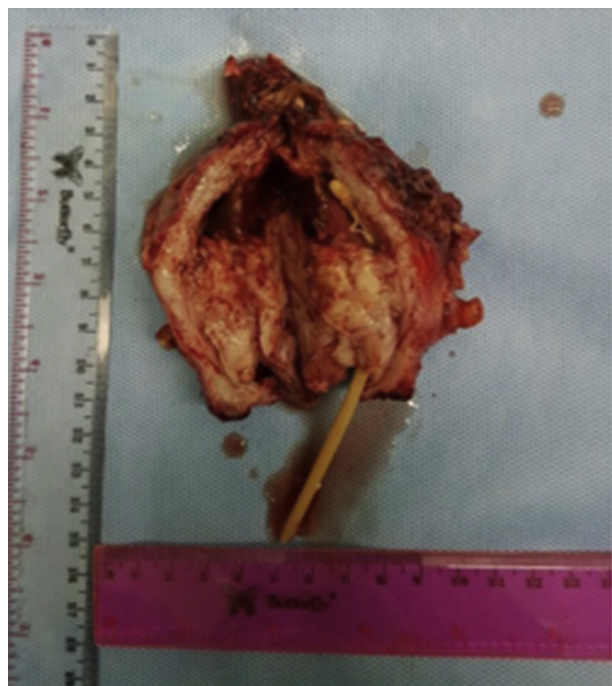


Figure 2. The result of the surgery. The large mass inside the bladder after excision.

Computed tomography revealed a 7,33 × 7,27 × 9,07 cm mass that filled almost of the bladder (Fig. 1). No regional lymphadenopathy was found. The kidneys showed hydronephrosis. No pulmonary metastatic was found.

The patient underwent rigid cystoscopy which revealed a huge bladder mass, normal bladder mucosa cannot be identified.

Biopsy of the mass revealed fibromuscular connective tissue with lymphocytes and histiocytes. Within those, there was endothelial cells proliferation. The nuclei were polymorph, hyperchromatic, mitotic figures, and wide necrosis area. Immunohistochemistry of the tumor cells were positive for CD 34 and negative for CK. And the result was hemangioendothelioma. Bilateral percutaneous nephrostomy was performed to improve general condition of the patient.

The patients was planned to excise the tumor. At exploration, there was umbilical abscess that caused severe adhesion to anterior abdominal wall. Dome of the bladder attached to ileum. There was infiltration to ileum and mesoileum. The left ureter was not infiltrated with the tumor, but the right distal ureter was infiltrated about 2 cm from ureteral orifice. A large bladder tumor was found with size about 10 × 6,5 × 6 cm (Fig. 2).

Because of involvement of the ileum, the ileum was resected, from 40 cm proximal of Bauhin's valve to 110 cm proximal of Bauhin's valve, and then anastomotic of the ileum was performed.

The histopathological examination showed the same result as biopsy (Fig. 3). The border of ileum specimen was free from tumor.

Discussion

Vascular tumors of the bladder are rare. Hematuria is the most common presentation of both benign and malignant lesions.^{1,4} Hemangioendothelioma is a vascular neoplasm that show a borderline biological behavior, intermediate between entirely benign hemangiomas and conventional angiosarcomas.^{1,2} The tumor is mostly developed in soft tissue, lungs, heart, liver, and bones, but in very rare instances, it can be found in bladder.^{5,6}

Only four cases have been reported in English literature, with three case in adult and one case in a child.^{3,5,6} Our case was the second case of hemangioendothelioma of bladder in a child and

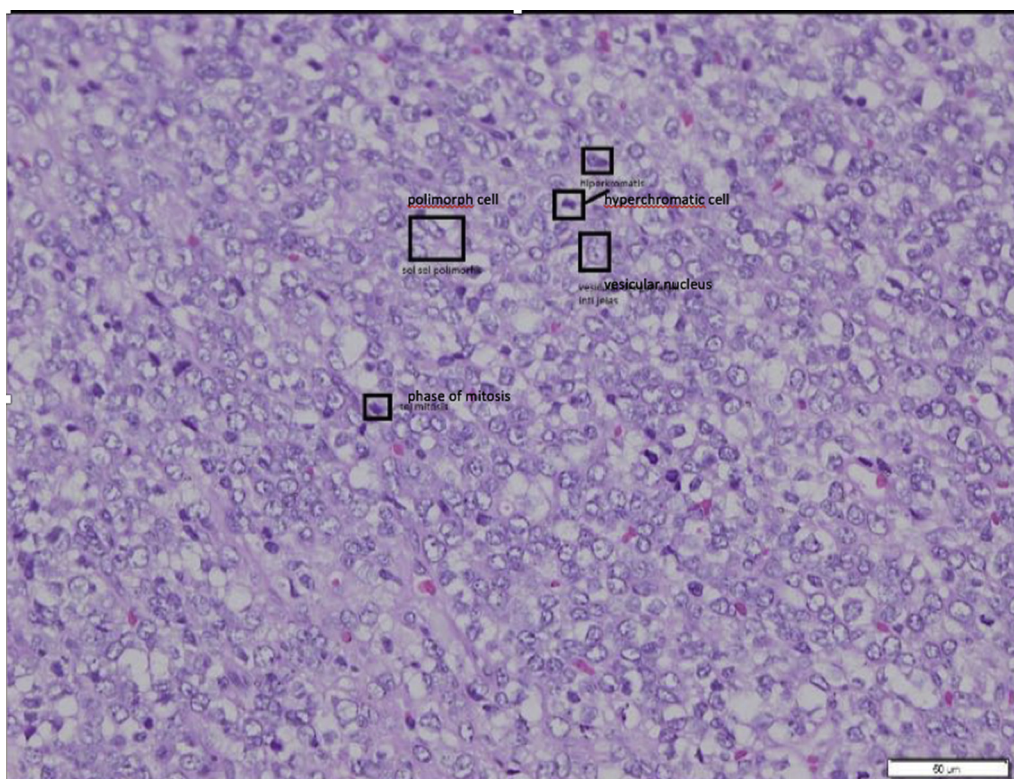


Figure 3. Pathology Anatomy Result with Hematoxylin-eosin stain (400x). The nuclei are polymorph, hyperchromatic, and there are mitotic figures.

was the youngest case. The first case in a child was reported in a 4 years old boy in English literature.⁵

The predisposition factor is still unknown.^{1,2,7} Our patient had no history of trauma and the prenatal history was good according to the family.

A majority of patients of soft tissue hemangioendothelioma can be cured following surgical excision of the tumor.

But the treatment of bladder hemangioendothelioma is still under consideration. In our patient, we performed total excision of the bladder with bilateral ureterocutaneostomy as urinary diversion. Bilateral nephrostomy was removed when we performed surgery.

Embolization is a therapeutic introduction of various substances into the circulation to occlude vessels, either to arrest or prevent hemorrhaging, to devitalize a structure, tumor, or organ by occluding its blood supply, or to reduce blood flow to an arteriovenous malformation.⁸ We can use embolization for hemangioendothelioma to get an adjuvant goal preoperatively, but because of intestine and bladder wall involvement, we decided to perform surgery without embolization.^{5,8}

The treatment of hemangioendothelioma in other sites may require a multimodality approach using steroid, cytotoxic agent, and interferon.^{2,9,10} Interferon alpha has been widely used in the treatment of human solid and hematologic malignancies. Interferon alpha appears to block cell proliferation, through the induction of apoptotic effects.^{10,11}

Diagnostic predictors is still unknown for this type of bladder tumor. At soft tissue, indicators of poor prognosis include nuclear atypia, increased mitotic figure, tumor cell splinding, necrosis, and extensive intravascular invasion or invasion into adjacent tissues or organs.^{2,5} In our patient mitotic figure and wide necrosis was found, so we can include this patient to poor prognosis.

The metastatic rate in soft tissue hemangioendothelioma is 21% and the mortality rate is 17%.^{1,2} Lungs and lymph nodes are the two most common metastatic sites.^{1,2,7}

From the others case, no recurrences of the tumor in 2 years follow-up, but in one case, there is slow progression of the tumor during 8 years follow-up.^{3,5,6} Our patient is doing well post operatively. But unfortunately interferon α -2b is not available in our hospital. at 3 months follow up, there was progressive progression of the tumor. Recurrent Abdominal mass was confirmed by Abdominal CT-Scan. The patient was died one week later. Interferon α -2b is might be an effective regimen on this tumor but further study is needed to confirm this statement.

Conflict of interest

There is no conflict of interest.

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