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Sarcoma botryoides in a 14 month old girl: A rare case

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Abstract

Embryonal rhabdomyosarcoma (Sarcoma botryoides) of the urinary bladder is an uncommon entity. Because of extreme rarity its discussion has mainly been in the light of individual case reports. We report a case of a 14-month old female who presented with urinary retension and abdominal pain. Her biopsy specimen confirmed sarcoma botryoides and she underwent abdominal cystectomy after a multidisciplinary consultation.

Keywords: Embryonal rhabdomyosarcoma, Sarcoma botryoides

Introduction

Embryonal rhabdomyosarcoma (ERM) originates from embryonal mesoderm and arises under the mucosal surface of body orifices such as the vagina, bladder and cervix. ERM is the most common soft tissue sarcoma in children and young adults and accounts for 4% to 6% of all malignancies in this age group.(1,2)

Sporadic cases have been seen in association with wilms tumor: one such case reported in the child with dandy walker syndrome. (3) In the past prognosis was poor with isolated cure being achieved by radical cystectomy and radiation therapy. The addition of multidrug chemotherapy to these modalities has notably increased survival rates, even when surgical resection was incomplete.(4) The prognosis is better in the tumor present as classic polypoidal growth as compared to diffuse intramural spread. Specimen from the recuurances often show a marked maturation of rhabdomyosarcomatous elements, presambly induced by chemotherapy.(5)

Case Report

We report here a case of 14 month old girl child presented with complaints of urinary retention, abdominal distension and pain abdomen since 1month which was gradually progressive in nature.

There was no significant past medical or surgical history and no family history of any malignancy. On examination, patient had mild pallor and there was no lymphadenopathy. Parametrium was reported normal.

USG abdomen shows thickened irregular bladder walls with changes of bilateral obstructive uropathy. CT scan shows a solid mass measuring 28x24x26mm present in the bladder.

Patient underwent chemotherapy for 9 weeks than transurethral resection done for this under general anesthesia.

Gross examination shows multiple grey white to grey brown polypoidal soft tissue pieces collectively measuring 6.8x5.5cm.external surface smooth glistening surface grey brown polypidal .cut surface grey white soft glistening.

Microscopic examination shows moderately cellular tumor composed of primitive mesenchymal cells contain both hypo and hyper cellular areas with loose myxoid stroma. Tumor shows cambium layer a hyper cellular zone immediately beneath the epithelial surface. Perivascular condensation of tumor cells in the less cellular regions are seen. Sheets of small, stellate, spindled or round cells with scant cytoplasm and eccentric ,small oval nuclei with a light chromatin pattern and inconspicuous nucleoli seen. Occasiinally tumor cells that shows rhabdomyoblastic diffentiation (strap cells) also seen. Features are suggestive of embronal rhabdomyosarcoma botryoid variant (Sarcoma botryoides).

IHC shows positivity for Desmin and Myogenin. Negative for CD 45 and S100.(Confirmed by IHC).

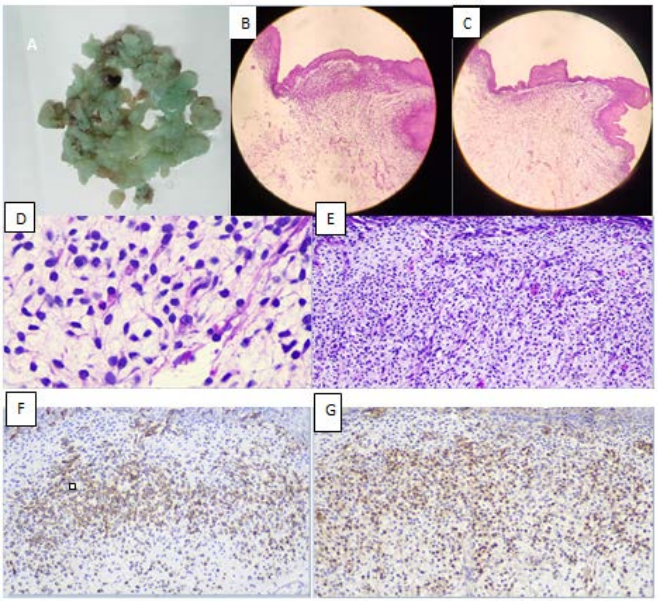

Fig A : Grapes like polypoidal tissue. Fig B & C – Shows Cambium layer & loose myxoid vascular stroma. Fig D & E –Shows hypercellular area,cells are round to oval with scant eosinophilic cytoplasm. Fig F & G –Desmin & Myogenin positivity respectively.

Discussion

Rhabdomyosarcoma is the most common pediatric soft tissue sarcoma. Approximately 10 to15% of all rhabdomyosarcoma cases arise from the pelvic organs [6]. The tumor is often large and involves adjacent organs and vessels. Histologically, bladder/prostate rhabdomyosarcoma is typically either embryonal or botryoid. After chemotherapy, radical surgery is required; this is usually urinary reconstruction or diversion, except for tumors arising at the top of the bladder [7]. Recently, multimodal treatment with chemo radio therapy and surgery has led to a good prognosis. Therefore, conservative surgery (subtotal/partial resection) is generally recommended [7]. The prognosis of localized embryonal bladder/prostate rhabdomyosarcoma is good [8], and conservative therapy improves patients' quality of life, as demonstrated by clinical trials [9]. cystoscopy for bladder rhabdomyosarcoma is usually performed only for biopsy or exploration, and not for definitive surgery, because the rhabdomyosarcoma arises from the muscle layer of the bladder. The long-term prognosis of conservative therapy requires consideration of clinical symptoms and objective evaluation of urinary function.

Conclusion

Histopathology is diagnostic but the treatment protocol is not codified due to its rarity. Surgery along with chemo radiation may results in prolonged survival, but requires more evidence.

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