Case Report

Embryonal Rhabdomyosarcoma of the Urinary Bladder: A Case Report

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Abstract

Embryonal rhabdomyosarcoma accounts for 4.5% of the childhood cancers. Rhabdomyosarcoma is the most common malignant soft tissue sarcoma in children and has the following subtypes: embryonal, alveolar, pleomorphic and spindle cell/sclerosing. About 17% of rhabdomyosarcomas occur in genito-urinary tract. A 2-year-old male child presented with complaints of excessive crying while passing urine and dribbling of urine for 1 day. Patient underwent cystoscopic biopsy and histopathological examination was done after taking written consent by the parents following which showed a cyst arising from bladder neck extending into right lateral wall of prostate. Partial excision of the cystic lesion was done. Gross examination of the biopsy revealed multiple grey-white to grey-brown soft tissue bits largest measuring 2cms in length and smallest measuring 0.2cm in length. Microscopy revealed a malignant neoplasm composed of poorly differentiated tumor cells arranged in sheets. Individual cells are primitive ovoid to spindle shaped with moderate pleomorphism and hyperchromatic nuclei with minimal cytoplasm admixed with myxoid areas. Few scattered Rhabdomyoblast like elongated cells with cytoplasmic eosinophilia and sparse atypical mitosis noted. Cambium layer was seen. Margin status was not assessed as the specimen was received in piece meal. Differential diagnosis of small round blue cell tumor was considered. Immunohistochemistry showed positivity for smooth muscle Actin, Desmin, Myogenin and MyoD1 suggesting Embryonal Rhabdomyosarcoma. Embryonal Rhabdomyosarcoma is the most common subtype of pediatric rhabdomyosarcoma. Age, Stage at diagnosis and treatment received are the predictors of the survival of Embryonal RMS.

Keywords: rhabdomyoblast; embryonal; genito-urinary tract.

Introduction

Embryonal rhabdomyosarcoma accounts for 4.5% of all childhood cancers. Rhabdomyosarcomas are the most common malignant soft tissue sarcomas in children and are divided into 4 subtypes: embryonal, alveolar, pleomorphic and spindle cell/sclerosing. About 17% of rhabdomyosarcomas occur in genito-urinary tract[1].

Case Report

A 2-year-old male presented with complaints of excessive crying while passing urine and dribbling of urine for 1 day.

Cystoscopy: Patient underwent cystoscopic biopsy after taking consent from the parents and histopathological examination was done which showed a cyst arising from bladder neck extending into right lateral wall of prostate. No other radiological reports were available. No past history of such complaints in the past.

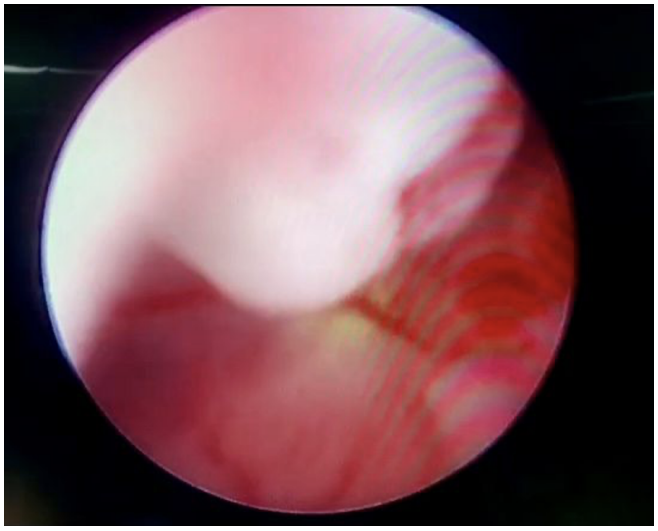

Figure 1: Cystoscopic view of the cyst.

Gross Examination: Partial excision of the cyst was done. Gross examination revealed multiple grey-white to grey-brown soft tissue bits largest measuring 2cms in length and smallest measuring 0.2cm in length.

Figure 2: Gross diagram of the cyst.

Microscopic Examination: Microscopy revealed a malignant neoplasm composed of poorly differentiated tumor cells arranged in sheets. Individual cells are primitive ovoid to spindle shaped with moderate pleomorphism and hyperchromatic nuclei with minimal cytoplasm admixed with myxoid areas. Few scattered Rhabdomyoblast like elongated cells with cytoplasmic eosinophilia and sparse atypical mitosis noted. Cambium layer was seen. No necrosis was seen. Margin status was not assessed as the specimen was received in piece meal. Differential diagnosis of small round blue cell tumor was considered.

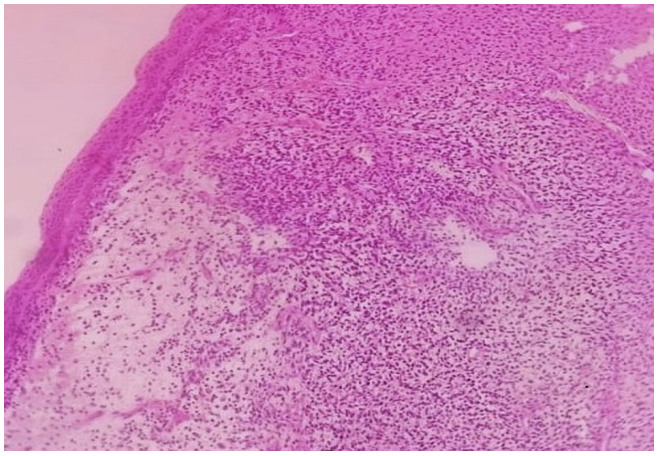

Figure 3: H & E stain – Cambium layer 10X.

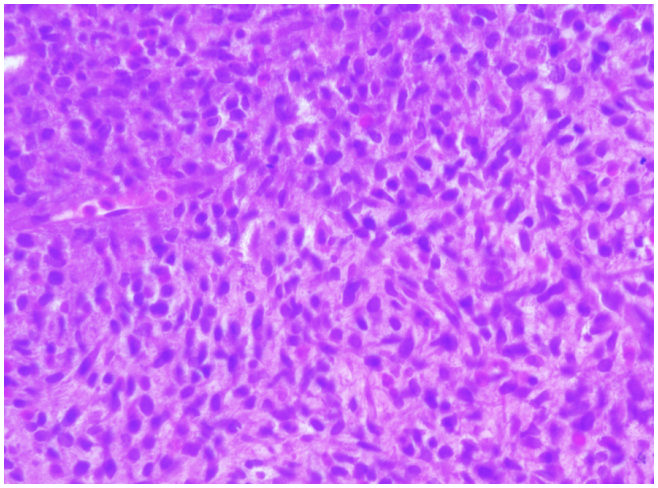

Figure 4: H & E stain of the cyst -40X.

Ancillary Studies: Immunohistochemistry was done which showed smooth muscle Actin, Desmin, Myogenin and MyoD1 positive tumour cells suggesting Embryonal Rhabdomyosarcoma. [Clones; Myogenin- F5D, Ki-67-SP 6, CD 45- PD7, Desmin-D33, CD34- QBEND/10]. As the patient had temporarily migrated from other state, follow up could not be done.

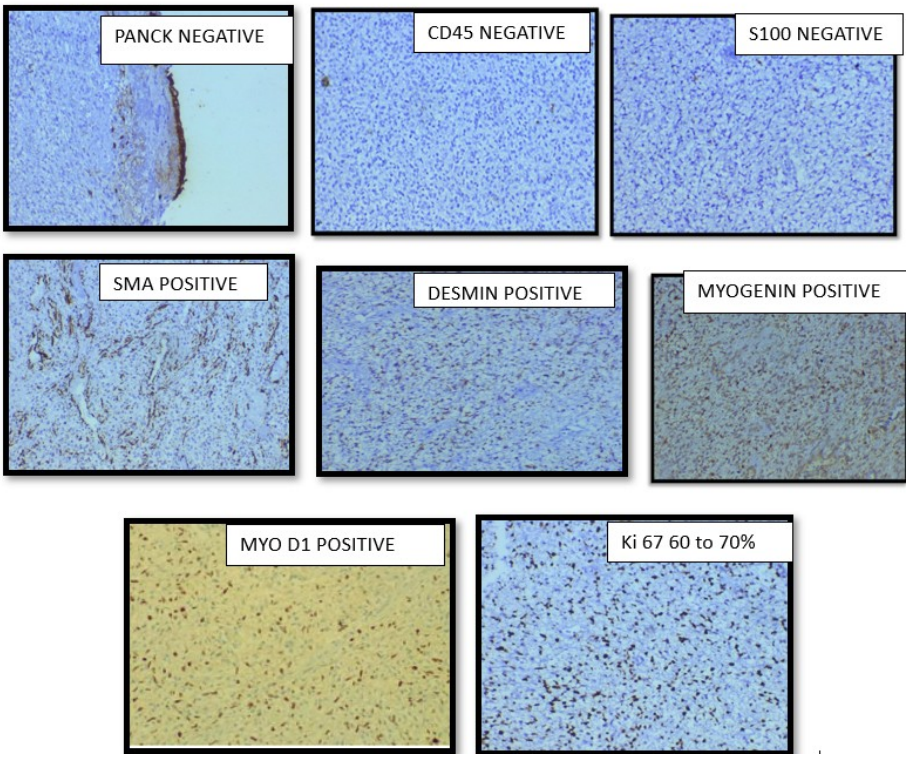

Figure 5: IHC work up.

Discussion

In infants, the most common soft tissue tumors are Rhabdomyosarcomas. Rhabdomyosarcomas share the property of myogenesis [1]. Embryonal rhabdomyosarcomas occur in sites such as urinary bladder, prostate, vagina and gall bladder which do not contain striated muscle fibres. The exact origin of rhabdomyosarcoma is not known but they can be induced by genetic manipulation and may be reproduced through tumorigenic influence [2].

There is bimodal age distribution of these tumors with peak occurrence in first two years of life and later in adolescence [3]. Increased prevalence of RMS is associated with Li-Fraumeni syndrome, Gardner syndrome, Beckwith-Wiedeman syndrome and Neurofibromatosis-1. These tumors show mutations like loss of heterozygosity at 11p15.5, whole/partial gain of chromosomes in 2, 8, 12, 13 with/or 20.2 mutations in FGFR4/RAS/AKT pathway and increased PTEN hypermethylation [4].

The rhabdomyoblasts seen in ERMS have heterogenous appearance. In low density areas, underdeveloped, round cells with

hyperchromatic nuclei with basophilic cytoplasm is seen in myxoid submucosa. Perivascular thickenings are seen in high cell concentration regions. Differentiated rhabdomyoblasts show acidophilic cytoplasm with cross striations [5].

On histology, cambium layer gives clue to diagnosis. Cambium layer shows densely cellular linear area tightly abutting the epithelial surface. Anaplasia in ERMS shows markedly enlarged hyperchromatic nuclei with or without atypical or bizarre mitotic figures [6].

Differential diagnosis of rhabdomyosarcomas include small round blue cell tumors in childhood namely neuroblastoma, Ewing sarcoma, synovial sarcoma, malignant melanoma, melanotic neuroectodermal tumor of infancy, lymphoma and malignant triton tumor [7].

Conclusion

Embryonal Rhabdomyosarcoma is the most common type of pediatric rhabdomyosarcoma. Age, stage at diagnosis and treatment are the predictors of survival.

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Competing Interests: No.

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