Case Report
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A Ten-Month Infant with Bladder Botryoid Sarcoma

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Abstract
Bladder tumors resulting in voiding difficulty in a child are rarely encountered in clinical practice. There are various investigations that help with preoperative diagnosis of bladder tumors and a systematic approach is needed to draw an early diagnosis. We report a case of the botryoid variant of rhabdomyosarcoma of the bladder in an infant that presented with complaints of a lower abdominal mass, poor urinary stream for three months and hematuria for one month.

Keywords: Botryoid Sarcoma; Bladder; Infant.

Conflict of interest: The authors declare no conflict of interest.
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Introduction
Bladder masses are rare in children and can be due to non-neoplastic or neoplastic causes. Neoplastic lesions may arise from the urothelium and underlying mesenchymal tissue. Papillary urothelial neoplasm of low malignant potential (PUNLMP) is the most common benign bladder neoplasm and rhabdomyosarcoma is the most common malignant bladder neoplasm in children. Other bladder tumors in children are leiomyoma, neurofibroma, inflammatory myofibroblastic tumor, hemangioma, and leiomyosarcoma (1). We present a rare case of the botryoid variant of embryonal rhabdomyosarcoma of the bladder in an infant.

Case report
A 10-month-old male infant presented with abdominal distension, weak urinary stream for three months, and occasional episodes of hematuria for one month. Examination revealed a soft, well defined, palpable mass in the suprapubic area. Computed tomography scan of the abdomen revealed a heterogeneously enhancing multiloculated polypoidal mass of 6x6x4cm with thick internal septa arising from the posterior bladder wall with no metastasis. Cystoscopy showed a patent urethra with a pinkish polypoidal mass arising from the posterolateral bladder wall. Exploratory laparotomy and excision of the bladder mass were done. Grossly, the tumor resembled multiple grape-like bunches with a gelatinous pearly white cut surface (Fig-1A & 1B).

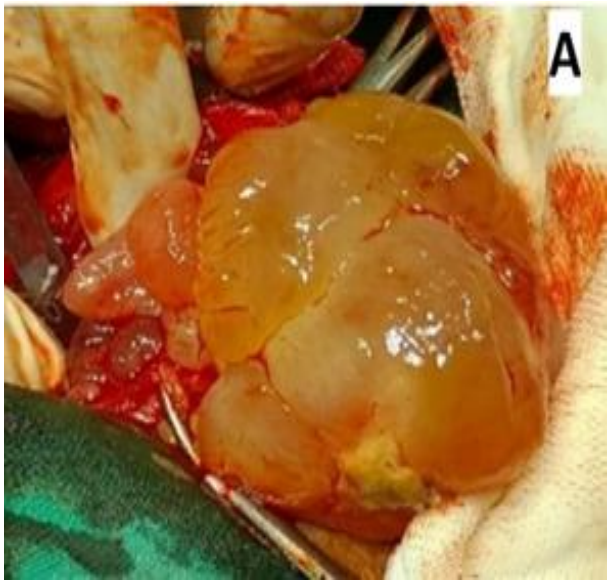

Figure 1A. Intraoperative photograph showing polypoidal translucent bunch of grape-like mass excised from bladder.

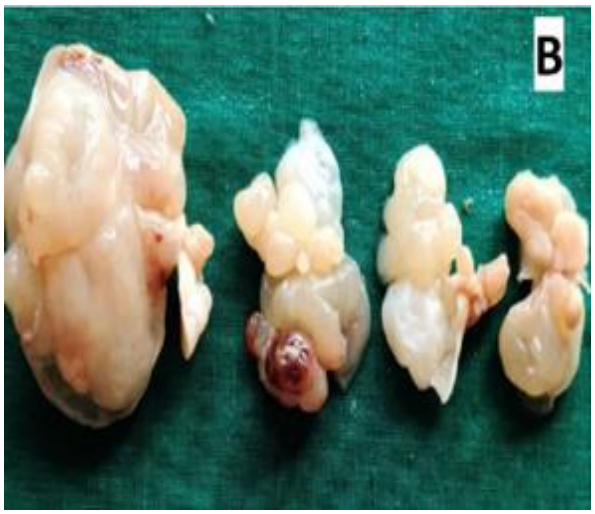

Figure 1B. Gross photograph showing gelatinous pearly white cut surface of mass.

Histopathological examination showed that the tumor was lined by transitional epithelium with a subepithelial collection of small, stellate, spindle cells with scant cytoplasm eccentrically placed oval nuclei known as the cambium layer of Nicholson (Fig-1C).

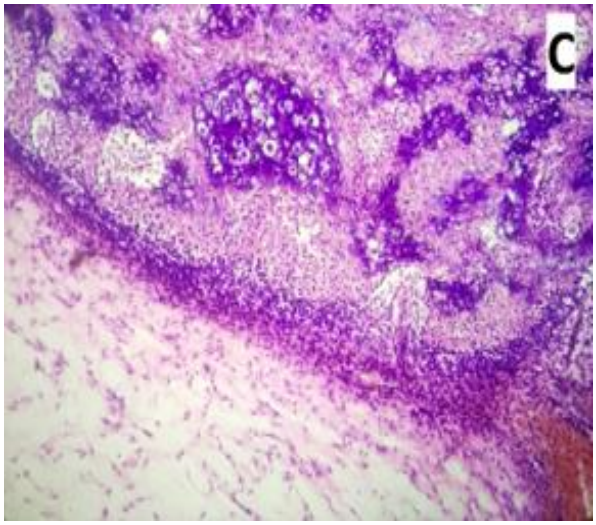

Figure 1C. Photomicrograph showing transitional epithelium with subepithelial collection of stellate to spindle shaped tumor cells known as cambium layer, which is characteristic of botryoid variant of embryonal rhabdomyosarcoma (H&E stain,40x)

The tumor cells were loosely arranged in a background of myxoid stroma (Fig-1D) with focal areas of immature cartilage.

Tumor cells showed nuclear positivity for desmin, myogenin and myoD1, confirming a diagnosis of the botryoid variant of embryonal rhabdomyosarcoma.

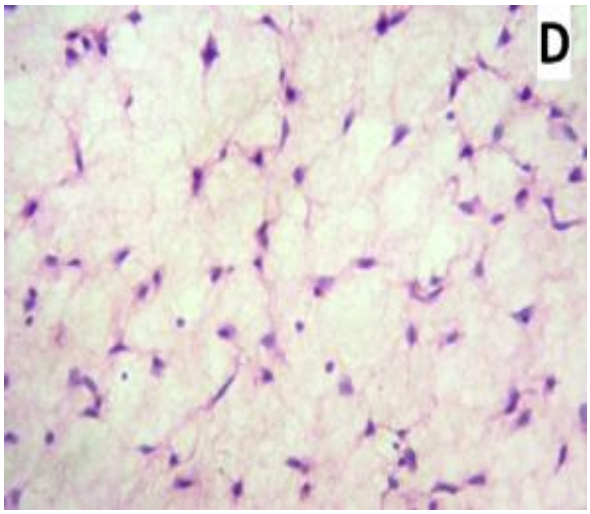

Figure 1D. Photomicrogrpah showing loosely cohesive stellate to spindle shaped cells with scant eosinophilic cytoplasm and round to oval nuclei in the background of myxoid stroma. (H&E stain, 400x)

Discussion

Rhabdomyosarcoma is a malignant embryonic tumor that arises from embryonic mesenchymal cells. It is the most common cause of soft tissue sarcomas in children accounting for 5% of childhood solid tumors. The tumor cells show differentiation towards mature striated skeletal muscle (2). Embryonal, alveolar, and pleomorphic rhabdomyosarcoma are three types of rhabdomyosarcoma. The embryonal type is the most common and occurs in the head and neck region, genitourinary tract, and retroperitoneum.

The botryoid variant accounts for 5-10% of all rhabdomyosarcomas and is a variant of embryonal rhabdomyosarcoma. It typically occurs in mucosa lined cavities like the vagina and bladder. The most common presenting symptom of rhabdomyosarcoma is difficulty in voiding due to a weak stream of the urine, as in our case (2). Gross appearance is typically described as a polypoidal bunch of grapes with a gelatinous cut surface (3). A characteristic microscopic feature is the presence of the cambium layer of Nicholson with aggregation of subepithelial malignant cells, which is necessary for diagnosis. The presence of the immature cartilage tissue in embryonal rhabdomyosarcoma is extremely rare (4). Ultrasound or CT scan of the pelvis is recommended to diagnose the nature of the bladder mass before cystoscopic examination as it is associated with bladder neck injury (1). Myogenin and myoD1 are the markers of skeletal muscle differentiation and are the most specific immunohistochemical markers to diagnose rhabdomyosarcoma (5). The botryoid variant of

embryonal rhabdomyosarcoma has an excellent prognosis compared to other types.

Conclusion

We report a case of the bladder botryoid sarcoma in a 10-month-old male infant that presented with abdominal distension, weak urinary stream, and hematuria. As botryoid sarcoma is rare, systematic and thorough evaluation with CT scan, cystoscopy, histopathological examination, and immunohistochemistry are needed to make an accurate diagnosis.

Conflict of Interest

The authors declare no conflicts of interest.

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