Vaginal Rhabdomyosarcoma in an Infant: A Case Report

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A girl of 5 months with vaginal bleeding and polypoid masses prolapsing through the vaginal opening was found to have sarcoma botroides. It had been noted 1 month prior to the time of the examination. Physical examination revealed soft, friable, and edematous polypoid, grape like, projections that almost fill up the vagina. Excisional biopsy was performed and histopathological examination revealed botroide rhabdomyosarcoma.

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Key words: Sarcoma botroides, Rhabdomyosarcoma

Introduction

Sarcoma botryoides, first described by Guersant in 1854, is an uncommon vaginal tumor composed of malignant embryonal rhabdomyoblasts usually arises from the anterior vaginal wall.[1] Approximately 90% of cases occur in girls under 5 years of age, with close to two-thirds appearing during the first 2 years.[2,3] These tumors tend to grow as polypoid, rounded, bulky masses that have the appearance and consistency of grapelike clusters (hence the designation botryoides, or grapelike). Such lesions can be mistaken for benign inflammatory polyps.

Case Report

A girl of 5 months from Pirgonj upazila was admitted into the Paediatric Surgery Department, Rangpur Medical College Hospital with complaints of vaginal bleeding and polypoid masses prolapsing through the vaginal opening for one month. On general examination, she was found to be ill looking. On local examination, soft, friable, and edematous polypoid, grape like, projections seen arising from anterior vaginal wall. Clinical presentation, the age of the girl and finding on examination of the lesion, go in favour of botryoid variant of embryonal rhabdomyosarcoma.

Interpretation of X-ray chest and all other investigation reports were insignificant except microcytic hypochromic anaemia.

The patient underwent excisional biopsy under general anesthesia. The postoperative course was uneventful.

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Pathological findings

On gross examination, three pieces of polypoid tissue were submitted. They were soft, friable gray white in color, measured about 3×3 cm each. On cut section, they were soft in consistency. On histopathologic examination, the tumor cells were small and had oval nuclei, with small protrusions of cytoplasm from one end. Beneath the vaginal epithelium, the tumor cells were crowded in a so-called cambium layer, but in the deep regions they lie within a loose fibromyxomatous stroma that was edematous and contain few inflammatory cells.

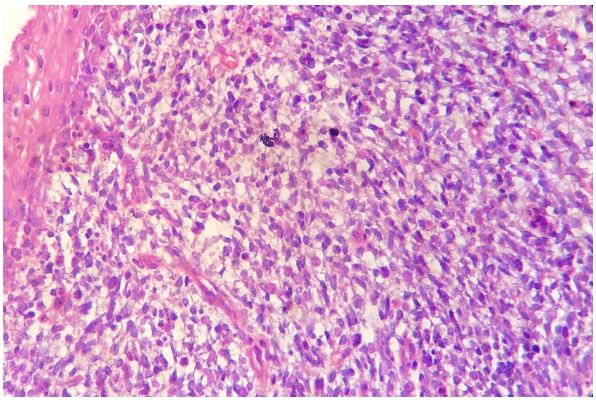

Fig 1. Histopathological section showing Cambium layer (H&E stain, ×400).

Discussion

Rhabdomyosarcoma (RMS) is a malignant mesenchymal tumor with skeletal muscle differentiation. Arthur Purdy Stout was the first to delineate it as a distinct entity.[4]

RMS is classified by the World Health Organization (2013) into embryonal RMS-49% (including botryoide, anaplastic), alveolar RMS-31% (including solid, anaplastic), pleomorphic RMS and spindle cell/sclerosing RMS-20%.[5,6]

RMS is the most common soft tissue tumor of childhood, and responsible for approximately one-half of all soft tissue sarcomas in this age group.[7,8] However, they are rare, representing only 3 to 4 percent of pediatric cancers overall. Approximately 350 new cases are diagnosed in the United States each year, and the annual incidence in children, adolescents, and young adults under the age of 20 is 4.3 cases per one million.[7] The incidence appears to be lower in Asians when compared with predominantly Caucasian populations.[9]

Embryonal RMS arises from unsegmented and undifferentiated mesoderm and is common in the head and neck region (particularly the orbit, nasopharynx, middle ear, and oral cavity), retroperitoneum, bile ducts, and urogenital tract. A smaller percentage occur in the extremities, within the thoracic cavity and in the skin are also on record. The large majority occurs in children between the ages of 3 and 12 years, but they can also be seen in younger patients and adults.[10, 11]

Sarcoma botryoides, is a variant of embryonal rhabdomyosarcoma accounts for 6% of all RMS.[6] Most botroides RMS develops in the walls of hollow, mucosal-lined structures, such as the nasopharynx, common bile duct, bladder, and vagina. Microscopically, a myxoid stroma is seen containing undifferentiated round or spindle cells. Some of these cells contain a bright eosinophilic granular cytoplasm suggestive of rhabdomyoblastic differentiation. Cross striations may or may not be present. An important diagnostic feature is the crowding of the tumor cells around blood vessels and a distinctive subepithelial dense zone (the 'cambium layer' of Nicholson). Foci of neoplastic cartilage may be found. The tumors tend to invade locally and cause death by penetration into the peritoneal cavity or by obstruction of the urinary tract. Of the 15 autopsied cases reviewed by Hilgers et al.,[12] the tumor was confined to the pelvis in about half.

The vast majority of RMS cases occur sporadically with no recognized predisposing factor or risk factors, although a small proportion is associated with genetic conditions. Li–Fraumeni cancer susceptibility syndrome, evident by a clustering of soft tissue malignancies (including sarcomas), has been discovered in a family to be caused by a heterozygous germline p53 mutation.[13]

According to review of the literature Miyamoto T. et al studied a case of botroid RMS.[14] In that study the tumor cells were immunoreactive for MyoD1 and myogenin. Due to limitation, in this case cytogenetic studies to detect any probable genetic factor and immunohistochemical examination could not be done.

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Conservative surgery coupled with chemotherapy offer the best hope, particularly in cases diagnosed sufficiently early.[15]

Conclusions

Polypoid mass protrude from the vagina in an infant or girl <5 years must necessarily be examined histopathologically because it might be a RMS. This is extremely important especially as an early disease stage at diagnosis is a highly favorable prognostic factor. Surgery and chemotherapy are the mainstays of treatment of RMS, and the prognosis of patients treated with multimodal therapy is very good.

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