Case Report

Challenges of Managing Childhood Cancers in Resource-Constrained Settings: A Case of Sarcoma Botryoides in a 3-Year-Old Girl

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Abstract

Sarcoma botryoides is a rare histological variant of rhabdomyosarcoma (RMS) found in the mucosal lining of body cavities of the bladder, vagina, nasopharynx, and biliary tract. Vaginal RMS typically affects young girls below 2 years of age but has also been reported in adolescents. Most patients present late when the tumor has already caused significant regional effects and complications. The management of such children usually poses numerous challenges in a resource-poor setting. A 3-year-old girl presented with a 1-year history of a progressive mass protruding through the vaginal introitus, associated with bleeding, progressive weight loss of 6 months, and abdominal swelling of 3 months duration. After 6 weeks of hospital stay, a diagnosis of sarcoma botryoides was made, and chemotherapy was commenced 2 weeks following the diagnosis. Delays were encountered due to financial constraints, laboratory stock-outs, and hesitance in accepting treatment by the parents. She completed one cycle of chemotherapy, comprising vincristine, actinomycin, and cyclophosphamide, with demonstrable reduction in the tumor sizes. Unfortunately, her management was complicated by severe measles infection acquired during admission; and she succumbed to the illness. Treatment of childhood cancers is bedevilled by the multiple levels of delays, and a host of co-morbidities that combine to produce an undesirable outcome. There is a need for a highly coordinated multidisciplinary approach that is hinged on a strong support for infection prevention and treatment access, in addition to good nutrition, adequate supply of blood, and blood products.

Keywords: Childhood cancer, embryonal rhabdomyosarcoma, sarcoma botryoides

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Introduction

Rhabdomyosarcoma (RMS) is a highly malignant skeletal muscle tumor that arises from embryonal mesenchyme. It is the most common soft-tissue sarcoma in childhood and young adults. RMS botryoides or sarcoma botryoides is a polypoid variant of embryonal RMS found in the mucosal lining of body cavities of the bladder, female genital tract, nasopharynx, and biliary tract.[1] In the female genital tract, vaginal tumors are five times more common than the cervical type and the latter appears to have a better prognosis than the former.[2] Vaginal RMS is usually of the botryoid variety, and typically affects young girls below 2 years of age but has also been reported in adolescents.[3,4] The tumor is thought to occur due to certain genetic mutations affecting chromosomes 2, 8, and 11 and may be associated with genetic disorders such as Beckwith–Wiedemann syndrome and Costelo syndrome among others. The clinical features may include protruding or infiltrative vaginal masses, vaginal bleeding, urinary symptoms, and pelvic or abdominal masses.[3]

Sarcoma botryoides arises from embryonal rhabdomyoblasts and constitutes approximately 3% of all RMS.[5] The appearance of botryoides RMS results when the tumor arises under the mucosal surface of the organs, which forces the growth to assume a typical grape-like structure. A distinct

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cambium layer beneath the epithelium is characteristic of botryoides RMS.[6] Treatment ranges from radical surgery to conservative surgery, in addition to chemotherapy.[7]

Case Report

A 3-year-old girl presented with a 1-year history of a progressively increasing mass, protruding from the vaginal introitus. There was associated intermittent vaginal bleeding of 5–10 ml per episode, with spontaneous resolution, and bleeding intervals of 3–4 weeks. No bleeding from other body orifices. She developed intermittent painful micturition 6 months into the illness with no other urinary symptoms. She also had progressive weight loss of 6 months and abdominal swelling of 3 months duration. She was treated at home with several oral and topical traditional medications, and later visited several hospitals without relief of symptoms.

Examination revealed a chronically ill-looking child, with marked wasting and stunting. She was febrile (38.1°C), and pale, and showed the features of malnutrition: Mild peri-orbital puffiness, brownish silky hair, pedal edema, and a baggy pant sign. Her abdomen was symmetrically distended with prominent anterior abdominal wall veins draining upward; the abdominal girth was 56 cm (10 cm distal to xiphisternum). There was an umbilical hernia with ring diameter of 2 cm. She had a suprapubic mass which measured 8 cm above the pubis symphysis and 5 cm in its horizontal axis. It had a firm consistency, smooth surface, and was tender to touch. On inspection of the perineum, there were irregular grape-like masses protruding from the vaginal introitus. The masses appeared smooth with glistening surfaces, with a focal necrotic spot inferiorly [Figure 1]. A digital rectal examination revealed a palpable mobile mass pushing on the rectum. Other systemic examination findings were normal.

Abdominopelvic ultrasound scan showed a huge mixed echogenic mass in the pelvis measuring 7 cm × 5 cm in dimension. The uterus was not clearly delineated from the mass. There was an oval shaped multiseptated cystic mass seen in the left adnexa which measured 4 cm × 3.5 cm in dimension. The urinary bladder was unremarkable.

Figure 1: Protruding grape-like mass from the introitus

Biopsy of the vaginal mass was performed after 3 weeks on admission due to the lack of funds by the parents. A histology report was obtained 6 weeks after admission, owing to stock-out of reagents in the laboratory. The results showed a focally ulcerated stratified squamous epithelium exhibiting papillomatoses and pseudoepitheliomatous hyperplasia overlying a tumor arranged in the diffuse pattern with varying cellularity. The tumor cells were noted to condense beneath the epithelium forming a cambium layer. They were round to oval and spindly cells having hyperchromatic nuclei with inconspicuous nucleoli placed eccentrically in the bright eosinophilic cytoplasm. Tadpole cells and strap cells (rhabdomyoblasts) were also noted. The stroma was loose and myxoid [Figure 2].

A diagnosis of advanced sarcoma botryoides was made, and the child was commenced on cytotoxic chemotherapy with a combination of cyclophosphamide, adriamycin, and vincristine. Treatment was started 2 weeks after the diagnosis, as parents initially were hesitant owing to the financial implications of the drugs. She had a cycle of the chemotherapy with palpable evidence of tumor shrinkage by approximately 25%. Unfortunately, she developed severe measles infection 5 days after the completion of the cycle and died from severe pneumonia.

Discussion

The average age at the diagnosis of embryonal RMS of the vagina is 2 years.[7] The index patient was within this age bracket and presented with a palpable pelvic mass up to the umbilicus and a grape-like mass protruding through the vaginal introitus.

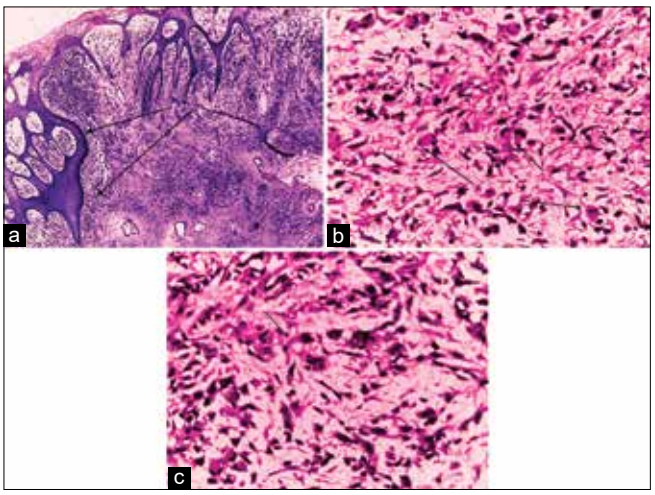

Figure 2: (a) Histologic section showing a polypoid stratified squamous epithelium overlying a tumour with sub-epithelial cambium layer (arrows). H and E, ×40 (b) Histologic sections showing a tumour with numerous rhabdomyoblasts having abundant, eccentric eosinophilic cytoplasm (arrows). H and E, ×200 (c) Histologic sections showing a "tadpole cell" (rhabdomyoblast)

She also had vaginal bleeding, urinary and bowel symptoms which are usual features of the disease and are similar to the cases reported by Dayyabu *et al*.[8] The histological findings of rhabdomyoblasts of varying differentiation dispersed within a loose myxoid stroma, and a distinct "cambium layer" beneath the epithelium further confirmed the diagnosis of sarcoma botryoides. As such this patient fulfilled the criteria essential for the diagnosis of sarcoma botryoides, which are: A polypoid appearance, origin below a mucous membrane-covered surface and the presence of a cambium layer.

Sarcoma botyroides is mostly treated with surgery alone or with adjuvant chemotherapy and/or radiotherapy.[3] The tumor is chemosensitive and could regress with only chemotherapy as reported by Dayyabu *et al*.[8] However, late presentation posed a threat to the success of treatment due to huge tumor bulk, with regional pressure effects on the gastrointestinal and urinary systems, and the possibility of metastasis.

This patient presented to our facility more than 1 year after the onset of her symptoms. Moreover, the time taken to make her diagnosis after admission was 6 weeks, followed by another 2 weeks before chemotherapy was commenced. Thus, there were multiple levels of delays encountered with this patient. The delay in the diagnosis had to do with procurement and logistics associated with laboratory, while financial constraints and delay in accepting treatment options by the parents delayed commencement of treatment. Sadly, she contracted measles on the open ward where she was being managed, from measles patients on admission at the same time. She developed a complicated measles course and succumbed to the illness. Even though the patient died of an illness not related to the primary disease, there were several factors that impacted negatively on the outcome. Some of these factors were late presentation due to poor health-seeking behavior, the use of traditional medications, untimely acceptance of treatment options, and financial constraints hampering timely interventions. Among the health care provider factors were delays in making a diagnosis due to stock-out of histological stains. The late presentation usually leads to advanced stage of the disease at presentation that impacts negatively on the treatment outcomes.

Infection control is crucial in the management of cancers since infections play a key role in the eventual outcomes. The immunodeficiency due to the cancer itself coupled with the immunosuppression of chemotherapy and or radiation necessitates prophylaxis and vigorous search for bacterial, viral, and fungal infections. This patient was nursed in the open ward due to the absence of a dedicated ward for children with cancer. Therefore, such a setting will always have the potential of infectious complications for immunosuppressed patients.

Conclusion

The treatment of childhood cancers is bedevilled by multiple levels of delays, and a host of co-morbidities that combine to produce an undesirable outcome. There is a need for a highly coordinated multidisciplinary approach that is hinged on a strong support for infection prevention and treatment access, in addition to good nutrition, adequate supply of blood, and blood products.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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