Sarcoma botryoides a management dilemma: A review of two cases

Aliyu Labaran Dayyabu, Adogu IO, Makama BS

ABSTRACT

Introduction: Sarcoma botryoides is a rare and rapidly growing tumor affecting primarily the female genital tract of children. It presents as bleeding per vaginum or as a polypoid fleshy mass protruding through the vagina. Treatment is surgery combined with multidrug chemotherapy. In developing environment, one has to contend with late presentation, availability and affordability of cytotoxics and the surgical options available at advanced stage, their acceptability, their management postoperative and their impact on quality of life.

Case Series: The first case was a four-year-old child who presented with a rapidly growing mass protruding per vaginum, intermittent vaginal bleeding and difficulty in passing stool and urine. She had examination under anesthesia and biopsy. Histology confirmed sarcoma botryoides. Her parents opted for multidrug chemotherapy and was treated with a combination of cyclophosphamide, vincristine and doxorubicin. She had six courses of the drugs with remarkable improvement in symptoms and the tumor regressed almost completely. Patient was subsequently lost to follow-up. The second case was a six-month-old child who presented with vaginal bleeding, vaginal growth and abdominal swelling. Histology confirmed sarcoma botryoides and the parents were counselled on management options which they declined and took the child home. The child was brought back with obstructive uropathy and septicemia. She had suprapubic cystostomy to relieve the obstruction. However, her condition deteriorated and she died a week after admission.

Conclusion: Sarcoma botryoides is a rapidly growing malignancy, early presentation and prompt and aggressive surgery combine with multidrug chemotherapy may be the answer if outcome is to be improved. To ensure this in low resource countries free health care for children and all individuals with malignancies and chronic debilitating diseases should be provided. In addition education and poverty alleviation will further save the situation.
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Keywords: Sarcoma botryoides, Rhabdomyosarcoma, Multidrug chemotherapy

INTRODUCTION

Sarcoma botryoides is a malignancy that arises from embryonal rhabdomyoblasts. It is also called embryonal rhabdomyosarcoma. The word botryoid in Greek means a bunch of grapes which characteristically describes the clinical appearance of the tumor. It is the most common soft tissue sarcoma in childhood and young adulthood, and account for 4–6% of all malignancies in this age group [1]. Sarcoma botryoides is usually reported as a
vaginal tumor in female reproductive tract of infants [2]. However, it also occurs rarely in the cervix or uterine fundus [3]. Unlike its counterpart in the vagina, which has poor prognosis, sarcoma botryoides of the cervix in young women has a favorable outlook [4]. Two other studies also reported better prognosis when the lesion affect the cervix [5, 6]. The survival rate of vaginal and cervical lesions has been reported to be 60–96%, respectively [7]. Vaginal bleeding is the most common presenting feature even though non-specific [8]. It may also present as a polypoid or fleshy mass in the vagina, or more classically projecting from the introitus. Other forms of presentations include urinary symptoms especially when the tumor is anteriorly situated or tenesmus where there is posterior extension [9].

There are different approaches in the management of this tumor, from simple excision to extensive radical mutilating procedures. These procedures may be combined with radiotherapy. However radiotherapy has been abandoned as it is now generally agreed that these tumors are not radiosensitive [9]. New multidrug chemotherapy regimens with or without radiotherapy are also used in combination with less radical surgery with good results, although outcome data are not yet available [10]. Radical surgery takes the centre stage in treatment as the disease is uniformly fatal with a five-year survival rate of between 10–35% [11]. Sarcoma botryoides has marked tendency for recurrence locally after excision and to invade adjacent organs [9].

We present two cases of Sarcoma botryoides to illustrate the difficulties encountered by Gynecologists in the management of this malignant and aggressive tumor (Sarcoma botryoides) in children in low resource environment and the benefit of multidrug chemotherapy when the disease is advanced.

CASE SERIES

Case 1: A four-year-old child was presented with a four-month history of serous vaginal discharge, three-month history of intermittent slight vaginal bleeding and two-month history of a mass protruding per vagina. The mass started increasing in size rapidly and occasionally bleeds on contact. Later, the mother noticed that the abdomen was progressively becoming distended with associated difficulty in passing stool and urine. There was also progressive weight loss. The mother started applying some herbal preparations on the mass without improvement. The child had five other siblings none of them had similar problem. Her pregnancy was carried to term and was uncomplicated. Mother had no antenatal care while carrying the child’s pregnancy.

On examination the patient (child) was found to be slightly emaciated, mildly pale, afebrile and anicteric. Her chest was clinically clear with 18 respiratory cycles per minute. Her pulse rate was 74 beats per minute and had no abnormal heart sounds. Her abdomen was distended firm and there was a suprapubic mass about 16 cm in size slightly mobile, one can get above but not below it. There was no demonstrable ascites and liver, kidney and spleen cannot be palpated. Pelvic examination revealed a polypoid mass, pink in colour, covering the whole of vaginal introitus (Figure 1). Further examination of the vagina was not possible because it was filled with the mass. The mass bleeds on touching. Rectal examination revealed a smooth anterior rectal wall with a mass pushing into the rectum.

An abdomino-pelvic ultrasound revealed a distended bladder and a mass measuring 6.1×5.2 cm lying posterior to the bladder. Both kidneys, liver, gallbladder and pancreas were grossly normal. There was no ascites. She had some investigations such as full blood count and differential, electrolytes, urea and creatinine, liver function tests and chest X-ray. All the test results were within normal ranges.

The parents were counselled on need for biopsy to confirm diagnosis before further management.

An examination under anesthesia and biopsy was done. Histology result revealed embryonal rhabdomyosarcoma (Figure 2).

The parents were informed of the result and options of management were discussed. The parents opted for cytotoxic chemotherapy.

The child was prepared for chemotherapy and the plan was to give a combination of cyclophosphamide (300 mg iv), vincristine (0.9 mg iv) and doxorubicin (22 mg iv) which will be given at two weekly interval. Before each course of treatment, the following investigations were done: full blood count and differentials, liver function test and electrolytes, urea and creatinine and any abnormality corrected before the next course is given. After four courses, the mass was noted to have regressed significantly in size (Figure 3), and the abdominal swelling disappeared. The difficulty of passing urine and stool also improved. Before the last course, the vaginal mass had disappeared completely and five days after the last course, the patient was discharged to come for follow-up in two weeks.

On follow-up the mother complained that the child’s underwear were stained with clear malodorous discharge. The patient was examined and vulva was found to be stained with serous discharge and after a course of antibiotics the discharge stopped. She had two more uncomplicated follow-up visits and was subsequently lost to follow-up.

Case 2: A six-month-old child was brought to the hospital with a complaint of a vaginal growth that appeared a month prior to presentation and vaginal bleeding. The mass bled intermittently especially on contact. There was also abdominal swelling which appeared about two weeks following the appearance of the vaginal mass. The vulva appeared normal at birth and pregnancy was uncomplicated and was carried to term. No history of similar problem in her siblings.
On examination the infant appeared stable, not pale and not febrile, no jaundice. Respiratory rate was 20 cycles per minute and her chest was clinically clear. Her pulse rate was 80 beats per minute and had no added heart sounds. Abdomen was slightly distended with a suprapubic mass of about 14 weeks size, firm, mobile and non-tender. Other organs were not palpably enlarged. Pelvic examination revealed a mass protruding through the vaginal introitus completely covering the urethral opening (Figure 4).

The parents were counseled on the need for biopsy before further management. The patient was admitted and the plan was to do a full blood count, liver function test, electrolyte, urea and creatinine and abdomino-pelvic ultrasound. Biopsy was taken and the result confirmed embryonal rhabdomyosarcoma (Figures 5 and 6) and the parent were informed of the result and counselled on the line of management. They signed against medical advice and left. After four weeks, the patient was brought back with complaints of inability to pass urine and constipation for three days.

On examination she was ill looking, dehydrated, febrile to touch (temp 38°C), pale but anicteric. Respiratory rate was 24 cycles per minute and there were fine basal crepitations in the lower lung fields. The pulse rate was 110 beats per minute and there were no added heart sounds. The abdomen was grossly distended and tender and organs could not be palpated. Pelvic examination revealed a large infected mass, with irregular
surface covering the whole of the vulva. Urethral opening could not be identified (Figure 7). Rectal examination revealed a mass bulging into the rectum with irregular surface and mobile anterior rectal wall mucosa.

An assessment of sarcoma botryoides, obstructive uropathy and septicemia was made.

Packed cell volume of the infant was 18%, urea level was elevated and electrolytes were derrange. Liver function was normal and urine microscopy and culture revealed a growth of *Escherichia coli*. She was rehydrated with intravenous fluid and was also transfused with packed cells and placed on parenteral antibiotics. She was also empirically treated for malaria parenterally. A Urologist was invited who suggested that suprapubic cystostomy was needed to relieve the urinary obstruction which was done. Patient’s condition continued to deteriorate despite all the treatment and died after a week of admission.

**DISCUSSION**

Rhabdomyosarcoma is a malignant tumor which arises from embryonic muscle cells [12]. It presents as a submucosal lesion giving the typical ‘grape like’ appearance and is usually seen in female infants and young children [13, 14]. Our two patients presented with grape-like, pinkish oedematous polyps filling the whole vagina. Other ways of presentation include recurrent vaginal bleeding, urinary symptoms and tenesmus [9]. The second patient presented with all these features while the first presented in a similar manner but with no tenesmus. A gynecologist managing such a case has to contend with a lot of issues which ultimately determines the outcome of treatment and the prognosis of the patient. One important issue is late presentation, this impact significantly on outcome. Some of the reasons for this is going to see traditional healers in the first instance and seeking modern medical care as the last resort. Both patients were initially treated by local traditional healers who gave them some herbal preparations to apply in the vagina. When the lesions continue to grow the parents brought the children to hospital. Financial constraints and lack of easily accessible health facilities also lead to late presentation. The parents of both children were peasant farmers living in remote villages very far from the nearest health facility. When eventually the patients presented the tumor had advanced to a stage where only palliative care is possible. The second problem encountered is refusal to accept the counselling given on the type of treatment which is likely to give a better result. In fact, after proper evaluation and counselling the mother of the second child signed against medical advice.
and left only to come back when the child had developed obstructive uropathy, anemia and septicemia.

Our intention in the treatment of the first case was to do surgery and follow it up with multidrug chemotherapy, however, when the parents refused surgery we offered the patient multidrug chemotherapy. The child had remarkable improvement after six courses given at two weekly intervals and the tumor grossly disappeared. She had three symptom-free follow-up visits and was subsequently lost to further follow-up.

All these problems arise because of illiteracy and poverty which create a vicious circle which compound the management of patients not only for a malignant and aggressive disease like sarcoma botryoides but also in the treatment of simple diseases. The solution lies in elimination of poverty and illiteracy.

CONCLUSION

Sarcoma botryoides is a rapidly growing malignancy, early presentation and prompt and aggressive surgery combine with multidrug chemotherapy may be the answer if outcome is to be improved. To ensure this in low resource countries free health care for children and all individuals with malignancies and chronic debilitating diseases should be provided. In addition, education and poverty alleviation will further save the situation.

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Author Contributions

Aliyu Labaran Dayyabu – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Adogu IO – Acquisition of data, Revising it critically for important intellectual content, Final approval of the version to be published

Makama BS – Acquisition of data, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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