Case Report

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Embryonal rhabdomyosarcoma of the cervix

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ABSTRACT

Embryonal rhabdomyosarcoma (sarcoma botyroides) of the cervix, which is rare, is described in a 16-year-old. The combined use of chemotherapy, radiotherapy and surgery has markedly improved survival in those with this condition. However, our patient did not benefit from this treatment modality due to late presentation and loss to follow-up.

Key words: Cervix, rhabdomyosarcoma, teenager

INTRODUCTION

Embryonal rhabdomyosarcomas (sarcoma botyroides) of the female genital tract are rare and usually occur during infancy in the vagina.[1-3] Only in rare cases can they be found in the cervix, where they are most commonly seen in adolescence and appear to have a better prognosis than sarcoma botryoides of the vagina and uterus.[3-5] Embryonal rhabdomyosarcoma of the cervix usually presents as a cervical polyp and sometimes there are multiple polyps.[5,6] A combined modality approach to treating this condition using multidrug chemotherapy, radiotherapy, and surgery has markedly improved survival.[7,8]

This case report illustrates this uncommon genital tract cancer and the problems associated with its management in our environment.

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CASE REPORT

A 16-year-old schoolgirl presented with a six-month history of a mass protruding from her vagina which progressively increased in size, associated with foul-smelling vaginal discharge and bleeding.

Significant clinical findings were fever (temperature 38.6^{0}C), and severe pallor with tachycardia. Pelvic examination revealed a prolapsed foul-smelling, oedematous, ulcerated, partly gangrenous mass which measured 20 cm x 20 cm with purulent discharge, bled on contact and was not reducible [Figures 1 and 2].



Figure 1: Photograph of the lesion with patient in dorsal position.

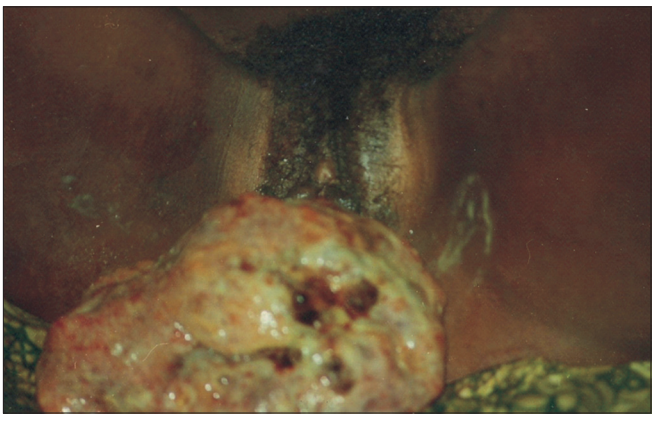


Figure 2: Prolapsed lesion with areas of necrosis and haemorrhage.

Her full blood count was within normal range except for her packed cell volume which was 19%. Urinalysis, blood chemistry and liver function tests were normal. Swab culture, microscopy and sensitivity of the mass yielded *Pseudomonas* spp sensitive to streptomycin, genticin and ofloxacin. Ultrasound scan revealed no renal or pelvic abnormalities except an absent uterus.

She was commenced on antibiotics and received blood transfusion. She had examination under a general anaesthesia and excision of the mass. The vagina was normal. The patient did not have full staging evaluation for distant metastasis and no pelvic lymph node sampling was done. The prolapsed mass was simply excised and sent for histology.

Pathological findings: The surgical specimen was a soft encapsulated greyish brown mass which weighed 200 g, measuring 14 x 14 x 8 cm and attached to the cervix. Cut sections showed variegated multiloculated haemorrhagic surfaces. The histological section of the tumour revealed an anaplastic sarcoma consisting of oval or spindle-shaped cells and showed marked pleomorphism and brightly eosinophilic cytoplasm. There were occasional strap cells and frequent racquet cells were observed in many areas. Islands of attendant necrosis and haemorrhage were present. Histochemistry demonstrated cross-striations in the cytoplasm of most of the strap-like cells. [Figure 3) Immunohistochemistry was not done as the facilities for this were not available in our centre. Embryonal rhabdomyosarcoma arising from the cervix was diagnosed. Hence the tumour stage according to the international Rhabdomyosarcoma Study Group clinical grouping classification was Class II (intermediate prognosis). She did well postoperatively and was discharged after six days. She however failed to turn up for follow-up. Efforts made to trace her later revealed that she died six months after surgery following recurrence of the mass with torrential vaginal bleeding.

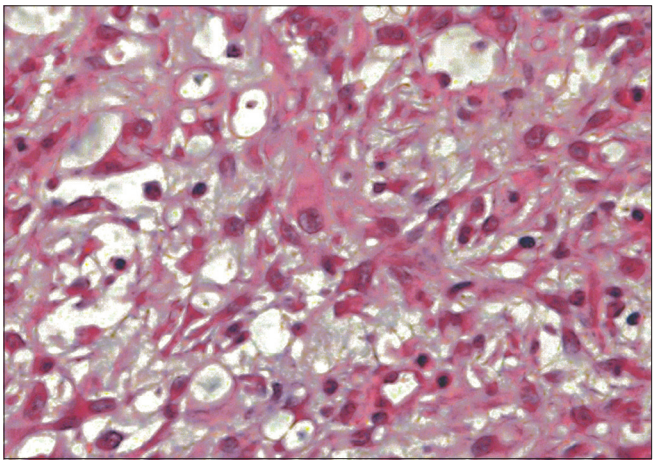


Figure 3: Histological section of the tumour. (H & E, x40)

DISCUSSION

Rhabdomyosarcoma (RMS) of the lower genital tract is a rare tumour. It usually occurs in childhood in the vagina and in rare cases originates from the uterine cervix, which occurs in women in their late teens and early 20s.[1-5] It could also occur in adults where it is usually encountered as a heterologous component of a malignant mixed mesodermal tumour.[1] Embryonal RMS of the cervix usually presents as a cervical polyp and sometimes as multiple polyps and may be associated with vaginal bleeding.[2,4,5] Our patient falls within the age group of those previously reported with cervical embryonal RMS.[2-5] She presented with vaginal bleeding and a fungating mass protruding from the vagina associated with foul-smelling discharge. This is not in keeping with most reported cases of embryonal RMS of the cervix. Though this was due to the advanced nature of the disease which is a common problem found in developing countries, it also pointed immediately to the malignant nature of the disease. Many patients from developing countries like Nigeria present late as they may first seek alternative treatment before presenting to the specialist[9] thereby presenting with advanced stages of the disease as this patient did.

Anaemia likely due to haemorrhage from the lesion was present in the patient for which she received blood transfusion. Recurrence of vaginal bleeding, probably resulting in shock led to the final demise of the patient. The lesion was also found to be infected for which she received antibiotics. Anaemia and sepsis are common findings in advanced genital cancers and result in patient demise as was seen in our patient.

Diagnosis is usually made by clinical findings and confirmed by histology[2] as was the case in this patient who had examination under anaesthesia, excision of the mass and diagnosis confirmed by histology

RMS of the genital tract is recognised as one of the most curable forms of RMS.[7] A multidisciplinary approach with more efficacious multidrug chemotherapy, conservative surgery, and limited radiotherapy, such as brachytherapy, has been used to help improve success rates.[3,5,7] Surgery has inclined towards a more conservative approach including radical abdominal trachelectomy with a view to preserve the patient's future fertility potential.[4,8] Unfortunately, our patient did not have fertility-sparing surgery due to the

advanced nature of the disease and did not receive chemotherapy and radiotherapy because she did not return for follow-up. Patient follow-up is a big problem in our environment due to poverty and ignorance.[9]

Embryonal RMS of the cervix usually has a good prognosis, especially in the early stages, with an aggressive behaviour in a minority of cases and the only known prognostic indicator is deep cervical invasion.[2,3] The patient presented with advanced disease and thereafter was lost to follow-up and these were factors in the poor outcome in this patient. Early presentation and treatment would have prevented the early demise of the patient. Public enlightenment, alleviation of poverty and accessible healthcare would encourage early presentation and proper follow-up of patients to avert unfavourable outcomes as was seen in this patient with advanced rhabdomyosarcoma.

REFERENCES

1. Borka K, Patai K, Rendek A, Sobel G, Paulin F. Pleomorphic rhabdomyosarcoma of the uterus in a postmenopausal patient. Path Oncol Research 2006;12:102-4.
2. Houghton JP, McCluggage WG. Embryonal rhabdomyosarcoma of the cervix swith focal pleomorphic areas. J Clin Pathol 2007;60:88-9.
3. Zeisler H, Mayerhofer K, Joura EA, Bancher-Todesca D, Kainz C, Breitenecker G, Reinthaller A. Embryonal rhabdomyosarcoma of the uterine cervix: Case report and review of the literature. Gynecol Oncol 1998;69:78-83.
4. Bernal KL, Fahmy L, Remmenga S, Bridge J, Baker J. Embryonal rhabdomyosarcoma (sarcoma botryoides) of the cervix presenting as a cervical polyp treated with fertility-sparing surgery and adjuvant chemotherapy. Gynecol Oncol 2004;95:243-6.
5. Ghaemmaghami F, Karimi Zarchi M, Ghasemi M. Lower genital tract rhabdomyosarcoma: Case series and literature review. Arch Gynecol Obstet 2008;278:65-9.
6. Bhandare D, Madiwale C, Kothari K, Pandit A, Kane S. Mullerian adenosarcoma of the uterine cervix. Indian J Pathol Microbiol 2001;44:371-2.
7. Magne N, Haie-Meder C. Brachytherapy for genital-tract rhabdomyosarcoma in girls: Technical aspects, reports and perspectives. Lancet Oncol 2007;8:725-9.
8. Kayton ML, Wexler LH, Lewin SN, Park KJ, La Quaglia MP, Abu-Rustum NR. Pediatric radical abdominal trachelectomy for anaplastic embryonal rhabdomyosarcoma of the uterine cervix: An alternative to radical hysterectomy. J Pediatr Surg 2009;44:862-7.
9. Chukwuanukwu TO, Anyanwu SN. Giant fibrosarcoma prostuberans of the anterior abdominal wall: Management problems in resources-constrained country. Nig J Clin Pract 2009;12:338-40.

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