

SARCOMA BOTYROIDES OF THE CERVIX IN A HIV POSITIVE 45 YEAR OLD WOMAN: A CASE REPORT

*DCD Anyiam, *CO Ukah, *IV Onyiaorah, **N Okafor

*Departments of *Histopathology, NAUTH, Nnewi and **Kanayo Specialist Hospital, Onitsha.*

ABSTRACT

Embryonal rhabdomyosarcomas (Sarcoma botyroides) are malignancies of connective tissue with abnormal cells which are thought to arise from skeletal muscle progenitors. It is a rare childhood malignant solid tumour and occurs in children mostly less than 10 years. It can occur at any age but two commonest peaks are 1-5years (when approximately 66.6% occur) and 15-19 years accounting for the rest. Sarcoma botyroides which usually occurs in the vagina in childhood is rare in the female reproductive tract of adult women especially those over the age of 40. We report here a case of a 45 year old HIV-positive woman who presented with profuse bleeding from the vagina with severe anaemia. Macroscopically, a cauliflower polypoid growth was seen protruding from the cervix into the vagina. Histopathological studies confirmed a diagnosis of sarcoma botyroides.

Conclusion: The need for a high index of suspicion of sarcoma botyroides of the cervix must be borne in mind when making a diagnosis on all cervical polypoidal masses. Also the importance of looking out for lesions and malignancies on all HIV positive patients cannot be over-emphasized.

Key Words: Sarcoma botyroides, cervix, rhabdomyosarcoma

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INTRODUCTION

Rhabdomyosarcoma is a rare tumour thought to arise from rhabdomyoblasts- a primitive muscle cell. There are five histological subtypes namely: Embryonal, alveolar, spindle cell, pleomorphic and sarcoma botyroides.

Embryonal rhabdomyosarcoma is the commonest type with cells which have a similar appearance to embryo cells aged 6-8weeks. Alveolar rhabdomyosarcoma with cells similar to embryo cells aged 10-12 weeks and occurs more in older children and teenagers and is more aggressive. Spindle cell type in which cells are spindled and appear to have a better prognosis. Pleomorphic type in which the cells are pleomorphic, they are more common in adults and has a tendency to affect muscles of the extremities and appears to have a worse prognosis. Sarcoma botyroides is a variant of the embryonal type and presents as grape-like lesions in the vagina, bladder or rarely cervix. Almost all cases are seen in infants.^{1,2}

Embryonal rhabdomyosarcoma (sarcoma botyroides) of the cervix, in contrast with the corresponding tumour in the vagina, usually occurs in women in their late teens and early twenties.³⁻⁸ Sarcoma botyroides of the cervix macroscopically is characterized by a polypoid, grape-like appearance. The polypoid masses have a glistening translucent surface and a soft consistency and may be

pedunculated or sessile. Their size ranges from 2-10cm. Cut sections are smooth and myxoid with small haemorrhagic areas.

Microscopically, it is composed of cells with round, oval or spindle-shaped nuclei and eosinophilic cytoplasm that may show differentiation towards striated muscle cells (rhabdomyoblasts). The subepithelial cambium layer is prominent.^{1,2}

CASE REPORT

Mrs. A.O. is a 45 year old female rural dweller who doubles as a petty trader and farmer. She is a widow. She presented on 13th January, 2008 in a private hospital at Onitsha having collapsed in her village due to bleeding per vagina. She was rushed to the hospital and resuscitated with two pints of blood and was later interviewed.

Patient was found to be a PLWA on anti-retroviral drugs for over two years. She said she had apparently been well and on her drugs till last year when she noticed a mass protruding from her vagina. The protrusion was said to have started insidiously on easing herself and never went back unless she pushes it back. The mass was red in colour and at first was not associated with bleeding but much later associated with daily bleeding. It was non-tender. There was no history of persistent chronic cough, trauma to the genitals, constipation or previous abdominal surgeries. There was no history of post-coital bleeding. She said she had been operated twice in different hospitals for this same problem. The first was in the last one year with no histology report but a few months later it recurred. She had her second

Correspondence: Dr DCD Anyiam
E-mail: dananyiam@yahoo.com

operation and a histopathological diagnosis of cervical polyp was made about four months prior to this current episode.

Past obstetric history showed she had three children none of which was alive. The first was born in January 1993 and died at 10 months. The second was born in 1998 and died at 4 years while the third was born in the year 2000 and died at 6 weeks. The cause of death of any of the children was unknown as no autopsies were done. There was no history of prolonged labour or instrumentation in all her deliveries.

Clinical examination revealed a cachectic patient with very severe palor. Vaginal examination revealed a mass protruding into the vagina and to the exterior. Gross examination with the speculum revealed bulky haemorrhagic multiple polypoidal fleshy masses extruding from the cervical area with focal areas of bleeding.

Findings at operation showed a cauliflower soft tissue mass that was quite large and extruding through the vagina. An incision biopsy was done to obtain tissue for histology.

Pathologically, gross examination revealed friable pieces of grayish white to tan tissues with a glistening translucent surface and a soft consistency admixed with some blood clots together measuring 6.8x5.8x0.7cm. Cut sections show a grayish white tissue admixed with clotted blood.

Microscopic examination showed the surface of the tumour to be covered by normal endocervical epithelium. Subepithelially, there was a dense cambium layer composed of closely packed cells with small hyperchromatic nuclei with an inconspicuous nucleoli. The central portion was typically hypocellular and myxomatous. Also seen were a lot of mitotic figures. The tumour cells were round or spindled with scanty eosinophilic cytoplasm. The nuclei were ellipsoid and some spindled with some atypia. There were also extensive areas of necrosis and haemorrhages. A diagnosis of sarcoma botyroides of the cervix was made.

Patient recovered and was advised to go to Ibadan or Lagos for combined modality therapy which would include chemotherapy, radiation therapy and surgery involving complete tumour resection, but nothing has been heard about the patient since then.

DISCUSSION

Embryonal rhabdomyosarcoma of the cervix (sarcoma botyroides) is a malignant neoplasm showing skeletal muscle differentiation arising from primitive mesenchymal cells.^{1,2} It is a rare tumour, but most commonly occurring in the late teens and early twenties and usually presenting as a cervical polyp.³⁻⁸ The polyps could be multiple.

The neoplasm in this case report has a multiple polypoidal large cauliflower shape and was protruding through the vagina. The patient in this case was older than most cases previously reported with embryonal rhabdomyosarcoma as it is very rare in patients forty

Figure 1: **Observe the pleomorphic hyperchromatic spindle to round body shaped cells and myoblasts with the ellipsoid and spindle shaped nuclei with some mitotic figures.**

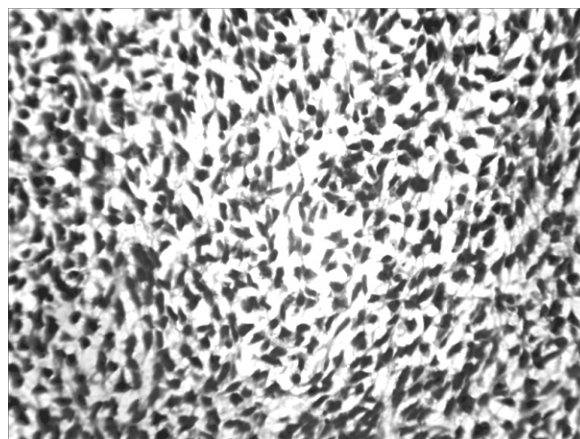
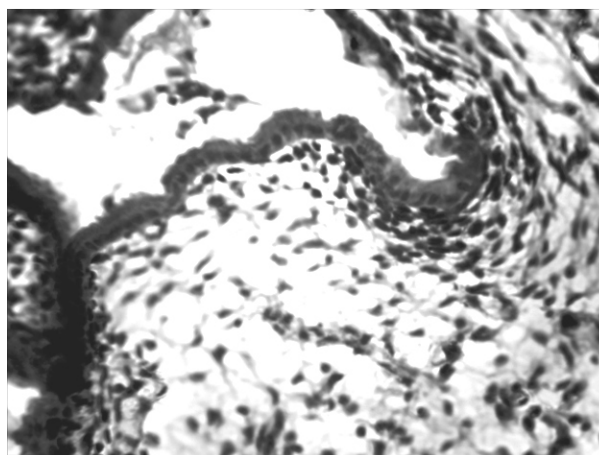


Figure 2: **Note the normal endocervical epithelium and the dense cambial layer with bizarre looking malignant cells.**



Five years and above.

Daya et al. reviewed nine cases of cervical rhabdomyosarcoma in patients 45 years of age and above between 1867 and 1963 (a period of almost one century) but there were no detailed clinicopathologic information on these cases.³ Ober reported this lesion in a 75 year old woman.⁵ A medline search for reports of cervical sarcoma botyroides since 1966 yielded only four well documented cases in patients 45 years of age and older.^{4,7}

The typical presentation is a polypoidal mass associated with vaginal bleeding. As exemplified by this case presented and those in the literatures, the tumours are often initially underdiagnosed or misdiagnosed as a benign endocervical or endometrial polyp, leading to often multiple recurrences after local excision.^{4,7} The corresponding tumour in the vagina usually occurs in infants and young children and the pathologist has to keep this possibility in mind when looking at sarcomatous lesions in the vagina of this age group.

This diagnosis must be borne in mind by both gynaecologists and pathologists in older age groups without obvious clinical suspicion of malignancy and in whom cervical polyps are common. A high index of suspicion by both the gynaecologist and pathologist would be of great assistance in all cases of cervical polyps to avoid misdiagnosis of this lesion as the combination of epithelial elements, squamous and glandular, in an oedematous stroma may result in misdiagnosis as a benign polyp.

The initial clue to the diagnosis is often the gross appearance, in that the polyp may be unusually large and myxoid or may be multiple. Other important clues to the diagnosis include the characteristic cambium layer, with increased cellularity around the subepithelial region. This cellularity diminishing towards the central myxoid area. Others include the nuclear atypia and mitotic activity seen in these areas. Presence of rhabdomyoblasts confirms the diagnosis though they may be sparse and in some cases absent. Positive staining with skeletal muscle antibodies helps in confirming the diagnosis, although some cases are negative and embryonal rhabdomyosarcoma can be diagnosed in the absence of staining with skeletal muscle markers.^{1,2}

Most useful immunochemical markers have been desmin, muscle-specific actin (HHF35) and myoglobin.^{1,9}

Apart from endocervical polyps, other differentials of interest would include prolapsed endometrial polyp, fibroepithelial polyp, endometriosis, leiomyoma, endometrial stromal neoplasm, adenofibroma and adenosarcoma. The later is most typical with same features as presence of a polypoid lesion with a cambium layer and associated nuclear atypia and mitotic activity may strongly suggest it also. However, adenosarcomas may contain heterologous elements in the form of rhabdomyoblasts and cartilages.¹

Embryonal rhabdomyosarcoma of the cervix usually have a good prognosis with an aggressive behaviour in a few cases.¹ The only known adverse prognostic indicator is deep cervical invasion. The worry about this case is its pleomorphic nature, pleomorphic cells having been observed and described and documented only in two previous cases of cervical rhabdomyosarcoma.^{4,10} The implications of the pleomorphic cells are not clear but it is believed that this may impart a worse prognosis as the presence of such cells was associated with a less favourable outcome.¹¹ Also worrisome in this patient is the presence of HIV which had been diagnosed over two years ago before the onset of this lesion. No literature was seen where HIV co-existed with sarcoma botryoides and this tumour is yet to be associated with HIV patients unlike HIV-associated tumours like Kaposi's sarcoma, B cell Non-Hodgkin's lymphoma, Primary brain lymphoma and squamous cell carcinoma of the cervix.

The advanced age at presentation and diagnosis in this patient may as well be due to her HIV positive status and her poor state of well-being. There is a reason to suspect this if we consider that Daya et al in their study

Of almost a century had only nine cases and had no clinicopathologic information on the cases. HIV status may as well be a clinical condition aiding and abetting rare childhood tumours to manifest and present at adulthood due to the lowered immune system.

CONCLUSION AND RECOMMENDATION

A strong advocacy must be encouraged to carry out and build a strong research capacity to detect and diagnose the patterns of HIV-Associated Malignancies and Associated Viruses as seen in our various environments in Nigeria.

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