Alveolar cell rhabdomyosarcoma of vulva in a HIV positive patient- a rare case report

Seethalakshmi Ganga Vellaisamy, Kumar Satagopan*, Shoba Dharmarajan**

Department of Skin & STD, Vinayaka Mission KirupanandhaVarayar Medical College & Hospital, Vinayaka Mission’s Research Foundation, Salem, Tamilnadu.
* Government Hospital Of Thoracic Medicine(GHTM), Tambaram, Chennai, Tamilnadu.
** Consultant Dermatologist, Porur, Chennai.

Abstract

A 21 year old female, HIV positive since 2008 presented with a multilobulated mass over external genitalia with ulceration of 3 months duration with bilateral inguinal lymphadenopathy. Histopathology was suggestive of alveolar cell rhabdomyosarcoma (ARMS). It was an uncommon sarcoma in a HIV infected individual which posed a diagnostic difficulty.

Key words
Alveolar cell rhabdomyosarcoma, HIV (Human Immunodeficiency virus), Multilobulated genital mass, Genital rhabdomyosarcoma.

Introduction

Alveolar cell rhabdomyosarcoma (ARMS) is an uncommon malignant soft tissue tumour rarely found in the female genital tract and carries a very poor prognosis especially in adults. Here, we report a case of genital ARMS in an immunosuppressed adult patient. To our knowledge, this is the first reported case of ARMS of the genitals in a HIV-infected patient.

Case report

A 21 year old unmarried female, HIV positive since 2008 presented in February 2015 with a multilobulated mass over the external genitalia with ulceration of 3 months duration. It started as a small nodule which gradually increased in size to about 16x12 cm and within one month the lesion ulcerated with foul smelling discharge. She was treated for pulmonary tuberculosis in 2008. She gave a history of blood transfusion at the age of 10 years for anaemias. She is currently on HAART (Highly Active Antiretroviral Therapy) (d4T/3TC/NVP) since June 2009 and denied any high risk behaviour.

On examination her body mass index (BMI) was 20 and she was found to be anaemic. Examination of abdomen revealed enlarged liver 5 cm below the right costal margin, firm in consistency, smooth surface, sharp borders and was not tender. No other mass was palpable per abdomen. Examination of external genitalia showed multi lobulated mass of about 16x12 cm in size, present over right labia majora, obstructing the vulval outlet (Figure 1) extending to perianal region (Figure 2) with ulceration and bleeding over the surface of the mass. Horizontal group of inguinal nodes were just palpable on both sides. Other systems were clinically normal. Per rectal and per vaginal examinations were not done because of the large size of the genital lesion.

Address for correspondence
Dr. G. V. Seethalakshmi M.D. (DVL)
11, Mullai Nagar, Near Chandra mahal,
Seelanaickenpatty, Salem- 636 201, Tamilnadu. India.
Ph: 91-98434 66486
Email: dr.seethalakshmiyadav@gmail.com
Figure 1  Multi lobulated mass present over right labia majora, obstructing the vulval outlet extending to perianal region with ulceration and bleeding over the surface of the mass

Figure 2  ARMS- extending to perianal region

Figure 3  Tumour cells distributed in alveolar pattern separated by thin fibrovascular septae (H and E, High-power magnification, x100)

Except for a hemoglobin value of 7g/dl all her other haematological, biochemical investigations were within normal limits. Her Chest X-ray was normal. Ultrasound examination of the abdomen revealed hepatomegaly. Her baseline CD4 count was 243 cells on initiation of HAART and her recent CD4 count was 320 cells. A skin biopsy was done from the lesion. On histopathological examination, the tumor consisted predominantly of cells distributed in alveolar pattern separated by a thin fibrovascular septae. The cells had moderate eosinophilic cytoplasm & vesicular nuclei with numerous vessels in between tumour cells suggestive of alveolar cell rhabdomyosarcoma (Figure 3).

The patient was referred to a tertiary care centre where she was put on combination chemotherapy (vincristin, adriamycin, cyclophosphamide). She underwent only one cycle of chemotherapy, after that she was lost to follow up.

Discussion

Rhabdomyosarcoma (RMS) has 4 histological types: alveolar, botryoid, embroynal and pleomorphic variants.\(^2\) The alveolar variant which is extremely rare in the female genital tract\(^2,3\) was first described by Riopelle and Thériault in 1956.\(^4\) ARMS tends to occur within skeletal muscle and is postulated to derive from precursor cells within the skeletal muscle. ARMS accounts for 20-30% of all RMS tumors. Therefore ARMS represents ~1% of all malignancies among children and adolescents, and has an annual incidence of ~1 per million.\(^5\)

It is an aggressive, rapidly progressing tumour.\(^3\) This tumor often presents as a painless mass, but in other cases, may be discovered from symptoms produced by compression of structures at the primary site.\(^5\) A substantial fraction of patients with ARMS (25-30%) will have metastatic disease at the time of diagnosis. ARMS most frequently spreads to bone marrow, distal nodes and bone.\(^5\)
Diagnosis of alveolar rhabdomyosarcoma is typically based on the cytological and architectural features of the tumour as seen on haematoxylin and eosin stain.\textsuperscript{3,6} Immunohistochemistry is essential in tumors that show poor cytological differentiation.\textsuperscript{7} Desmin is the best single marker for RMS cells as it is consistently present. Myoglobin, though a specific marker of skeletal muscle tumors, is restricted to more differentiated rhabdomyoblasts and generally absent in primitive round cells.\textsuperscript{7} Myo-D1, a nuclear antigen typical of cells of striated muscle lineage, apparent in primitive tumor cells is a sensitive and specific marker for RMS.\textsuperscript{8}

Due to the rarity of this tumour variety, there is no standard treatment established in the literature for adult patients and optimal management remains unknown.\textsuperscript{3} The treatment of rhabdomyosarcoma, including its alveolar subtype has markedly improved in children and adolescents over the past years mainly due to the use of a multimodality approach consisting of surgery, radiotherapy and chemotherapy.\textsuperscript{3} Ferrari et al. recommend similar treatment in adults.\textsuperscript{9} The Intergroup Rhabdomyosarcoma Study IV (IRS-IV) revealed that though VAC (Vincristine, Actinomycin and Cyclophosphamide) chemotherapy, with or without radiotherapy, remains the gold standard of treatment in children and adolescents, the alveolar subtype fared worse than other types.\textsuperscript{2,10}

Surgical management consisting of excision with the margins free of tumour and resection of the draining lymph nodes together with pre- and postoperative radiotherapy to the tumour site and involved nodal group offers the best chance of cure.\textsuperscript{3} While Ng et al. used surgery, aggressive adjuvant chemotherapy (three 6-weekly cycles of VAC, etoposide, ifosfamide and mesna) and radiotherapy to treat an adult case of alveolar rhabdomyosarcoma of the cervix\textsuperscript{10}, Case et al. used five cycles each of neoadjuvant and adjuvant VAC chemotherapy and surgery to treat another adult case of alveolar rhabdomyosarcoma of the uterus\textsuperscript{2}, with both cases achieving complete remission.\textsuperscript{2,10}

HIV-infected patients develop malignancies more frequently than the general population. The rates of AIDS-defining malignancies (ADMs) like Kaposi’s sarcoma, non-Hodgkin’s lymphoma and invasive cervical cancer have continued to fall since the advent of HAART, but the rates of non AIDS-defining malignancies (NADMs) such as Hodgkin’s disease, lung, liver, anal, head and neck cancers are rising and now account for the majority of cancer cases.\textsuperscript{11} The increasing rates of NADMs appear most related to the aging of the HIV population. Antiretroviral therapy appears protective for the development of ADMs, but had no significant impact on NADMs.\textsuperscript{11} In our patient the non AIDS –defining malignancy occurred inspite of HAART.

**Conclusion**

We are presenting this case for its rare occurence. To the best of our knowledge this happens to be the first case of alveolar rhabdomyosarcoma involving external genitalia in a HIV positive individual. Patient’s poor general condition and the resource limited setting were limitations for further investigations in this patient for academic evaluation.

**References**