Case Report

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Uterine Alveolar Rhabdomyosarcoma in a Young Adult: Case Report of Rare Uterine Tumour

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ABSTRACT

Alveolar rhabdomyosarcoma (ARMS) of the corpus uteri is a very uncommon malignant tumour that has rapid progression and a poor prognosis particularly in young adults. Guidelines and data regarding treatment are limited and usually applied by extrapolating from protocols for treating sarcomas from non-uterine sites and from guidelines for paediatric sarcomas.

Case: A 21-year-old nulliparous female was evaluated for an abdominopelvic mass. Examination findings were a large abdominopelvic mass. An exploratory laparotomy was done. Massive intraoperative bleeding from a friable uterine mass with adhesions necessitated a subtotal hysterectomy. Histology confirmed a uterine alveolar rhabdomyosarcoma. The patient was planned for adjuvant chemotherapy postoperatively with the VAC regimen but follow up was a challenge due to financial challenges.

Conclusion: This case report describes a rare cause of an abdominopelvic mass in a young adult of reproductive age due to an alveolar rhabdomyosarcoma and discusses therapeutic options available.

Keywords: Alveolar rhabdomyosarcoma, Uterus, Adult

INTRODUCTION

Alveolar rhabdomyosarcoma (ARMS) are rare aggressive malignant mesenchymal subtypes of rhabdomyosarcomas (RMS) usually seen in the paediatric age groups in children younger than 10 years and arising from embryonic muscle cells. [1]When they do occur in adults, they tend to be located in deep soft tissue of the limbs.[1] Primary RMS of the corpus uteri are even more uncommon compared to other gynaecological sarcomas and where they do occur, may be preceded by leiomyoma of the corpus uteri.[1] For some uncertain reasons, the prognosis is worse in adults than in children[1] This tumour is thought to arise from cells that have skeletal tissue lineage and constitutes 2-5% of soft tissue sarcomas that may be differentiated into embryonal,

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pleomorphic or the alveolar type.[2] The presented case is especially uncommon because of its alveolar tissue type and the location on the corpus uteri. It is presented to further our understanding about the presentation and management options for rare uterine masses.

CASE REPORT

Miss. GB was a 21-year-old nulliparous Senior Secondary 2 student who presented to the gynaecological emergency unit of the Jos University Teaching Hospital (JUTH) following a referral from St. Louis Hospital, Zonkwa Kaduna State with a 12-month history of recurrent abdominal swelling and a 2-month history of abdominal pain. About 10 months earlier she had undergone abdominal surgery at General Hospital, Zonkwa for similar complaints and a right oophorectomy was done but the sample was not sent for histology. No uterine biopsy was performed.

On examination, significant clinical findings were an asymmetrically distended abdomen with a Pfannenstiel scar, generalized tenderness and a huge abdominopelvic mass of about 30weeks pregnancy size. The uterus could not be appreciated bimanually due to the size of the mass. Packed cell volume (PCV) was 30%, white blood cell count (WBC) was essentially normal, serum chemistry revealed an elevated creatinine of 238 µmol and alkaline phosphatase of 100IU.Tumour markers showed an increased CA-125 of 48 IU and a normal AFP level of 6.42 IU. HIV and Hepatitis B surface antigen tests were non-reactive. Ultrasonography showed a huge heterogeneous, predominantly hyperechoic mass with lobulated outline measuring more than 22 by 15 cm, harboring an anechoic central area surrounded by a thick echogenic rim and demonstrating colour flow on doppler interrogation. The mass extended from the pelvis to the epigastrium and was reported to have been arisen from the right ovary, separate from the uterus. The left ovary was not visualized. There was enlargement of the liver and both kidneys, while the spleen measured 10.3 cm and was noted to harbor multiple echogenic foci. Mild ascites was reported. A provisional diagnosis of an ovarian tumour was made She was counselled on the examination and investigation findings and the need for an exploratory laparotomy with the possibility of a hysterectomy, for which a written informed consent was obtained.

Exploratory laparotomy revealed a bulky, friable, and highly vascular uterus of 28-week pregnancy size with extensive greyish white tissues and a single nodular friable mass. The left ovary and uterine tubes were not visualized but the right ovary and tubes were grossly normal. There were bowel loops adherent to the posterior uterine wall, the estimated blood loss of 3500 milliliters. She was transfused with a total of 5 units of blood in theatre. She was admitted into the intensive care unit (ICU) for the initial 24 hours post-operative during which she received an additional 3 units of whole blood. Her pulse rate on admission in the ICU was 110 beats per minute, the blood pressure was 80/60mmHg.The pulse rate was 90 beats per minute and the blood pressure increased to 110/70mmHg after 6 hours of admission in the ICU. She was discharged to the gynaecological outpatient clinic on the seventh post operative day after referral to the radio- oncology unit for chemotherapy. Microscopic examination showed ramifying fibrous septae with monomorphic small round blue cells attached with minimal cytoplasm and hyperchromatic vacuolated nuclei in foci. There was focal eosinophilic tapering with plasmacytoid features

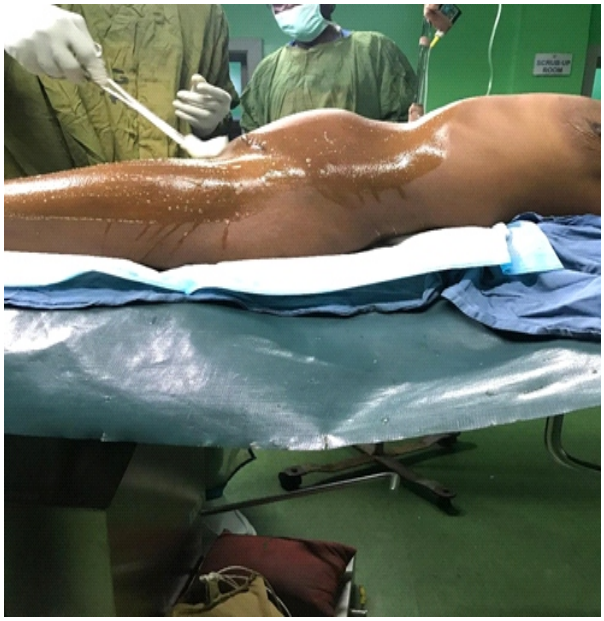

Figure 1: Abdominopelvic mass before surgery

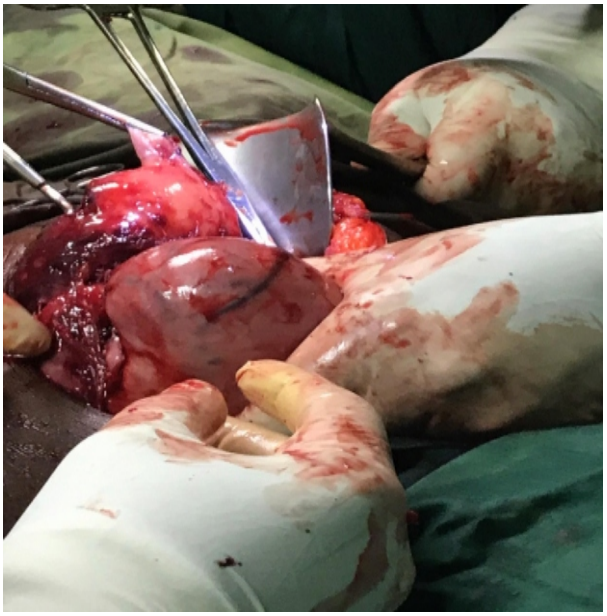

Figure 2: friable uterine tumour intraoperation

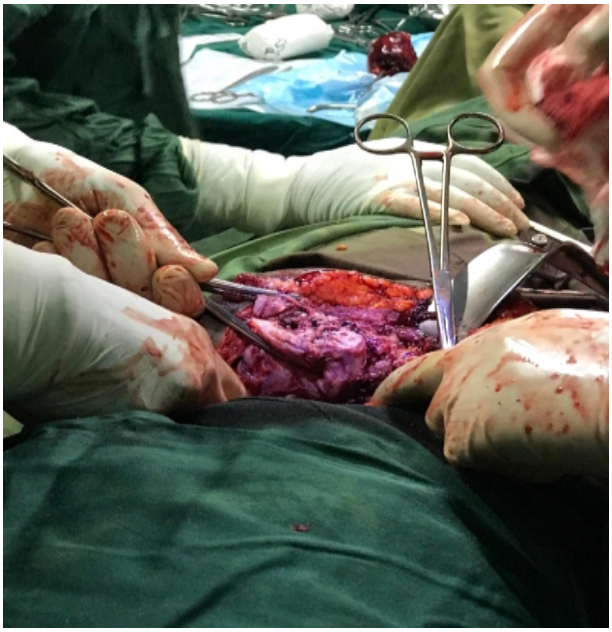

Figure 3: Residual stump after subtotal hysterectomy

with focal necrotic and mitotic areas. A histological diagnosis of alveolar rhabdomyosarcoma was made.

DISCUSSION

There are four histological varieties of rhabdomyosarcomas alveolar (which has a botryoid subtype), embryonal and pleomorphic subtypes.[3,4] Embryonal rhabdomyosarcomas are commonly called sarcoma botyroides and are the most common soft tissue sarcoma in childhood and young adults accounting for 4-6% of malignant tumours in this age group typically located in the vagina and rarely the uterine cervix or the urinary bladder.[5] The presented case in a young adult is unusual therefore being alveolar in nature and occurring in a young adult. Reviews of RMS of gynaecological origin in young adults show that ARMS are second in prevalence to embryonal sarcomas with an age range of 16-48 years with 79% of patients presenting with locoregional tumours that require surgery as a primary intervention but have a more aggressive progression and worse prognosis.[2 6,7]

The mean Progression Free Survival (PFS) in some reviews was as low as 9 months, and a 5-year disease specific survival was 29%.[8] PFS was 19 months in embryonal sarcomas compared to just 3 months in cases with non-embryonal histology.[8] Gerber in a 10-year review found the age at presentation in adults to be 28 years with the embryonal types being the commonest presentation with 54% of cases. This was followed by the alveolar types (33%), and the pleomorphic tumours accounting for 13% of cases. The 5year survival rates for non-metastatic disease was 45% while the 5 year survival rate for metastatic disease was 26%.[9] There is no consensus on the treatment modalities for ARMS. Multimodal therapy with debulking surgery, chemotherapy and radiation has improved outcomes particularly in paediatric age groups. Combination therapy with Vincristine, dactinomycin and cyclophosphamides, chemotherapy treatment regimen for adults are options that have been used but these are yet to be standardized as protocol options.[9] The patient had debulking surgery with preservation of the solitary ovary seen at surgery because of her young age, and the significant blood loss occurring during the procedure. While being planned for adjuvant chemotherapy that she defaulted on due to financial constraints. Patients with alveolar RMS have poorer outcomes even without metastatic spread than any of the other subtypes.[10] It has been suggested that outcomes of RMS are better when the treatment guidelines for children are applied to adults who present with ARMS, with the rate of response to chemotherapy in adults similar to children at 85%.[1113] Adjuvant chemotherapy before exploratory laparotomy could be considered a non-surgical option but, in this case, we were unaware of the likely tumour type before surgery and sought to relieve the symptoms of abdominal pain and discomfort. The initial surgery at the referral hospital provided an opportunity for maximal debulking surgery that was missed particularly because no specimen was sent for histology. The uncommon nature of this tumour has made it difficult to find standardized treatment protocols in the literature.[14]

CONCLUSION

ARMS are rare aggressive tumours of the uterus with poor prognosis and the worst disease-free survival rates of gynaecological sarcomas with unclear protocols for chemotherapy. Local reports on its prevalence in adults are scarce. Further studies are required to offer patients the best chances of disease survival, but early detection is likely to improve survival in patients.

Disclosure Of Conflict Of Interest

No conflict of interests in this report.

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