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Case Report

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A case report on management of yolk sac tumour of uterine cervix in a 2-year-old child

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Abstract

Giant cell tumour (GCT) of malignant origin is a rare and heterogeneous group of tumours in the paediatric population. It could be seminomatous and non-seminomatous. Non-seminomatous group is less commonly seen. The most commonly seen type under the non-seminomatous group under the age of three years is the yolk sac tumour. Majority of yolk sac tumour is seen in the gonad while very few develop in extra gonadal sites such as the sacrococcygeal region, retroperitoneum, mediastinum, pelvis and stomach among others. Extral gonadal germ cell tumour is extremely rare in the cervix and when it occurs there may be challenges in the management. We presented a case of a 2-year-old female who presented with a 9-month history of persistent bleeding and discharge per vagina that was diagnosed as a yolk sac tumour and successfully treated with chemotherapy to preserve.

Keywords: yolk sac tumour, chemotherapy, uterine cervix

Introduction

Germ cell tumours (GCT) are rare malignant tumours with heterogeneous origins accounting for about 3% of all cancers in the paediatric population.[1] It could be seminomatous and non-seminomatous which is less commonly seen.[2] The most commonly seen type under the non-seminomatous group under the age of three years is the yolk sac tumour otherwise known as an endodermal sinus tumour..[3] Majority of yolk sac tumour is seen in the gonad while very few develop in extra gonadal sites such as the sacrococcygeal region, retroperitoneum, mediastinum, pelvis, pineal gland, liver, omentum and stomach among others.[4] The most common extragonadal site is the mediastinum.[5] GCTs predominate in female subjects (boy: girl ratio=0.8:1).[6] Commonly involved extragonadal site in the genital system in females is the uterus, vaginal and cervix in that order of sequence.[5] Similarly, yolk sac tumour accounts for 5-10% of all germ cell tumours in the USA.[7] The aetiology of the yolk sac tumour is largely unknown while the risk factors for germ cell tumours are undescended testes, radiation exposure, disorder of sex development, family history, infertility and Klinefelter syndrome among others the only consistent risk factor yolk sac tumour in the medical literature is Klinefelter syndrome.[8] The pathophysiology of extragonadal yolk sac tumour is unclear. Classic theory explained local transformation of misplaced primordial germ cells while the alternative theory suggests reverse migration of occult carcinoma in situ from the gonad.[9] The clinical presentation of patient with yolk sac tumour depends on the site of the tumour, the patients with ovarian yolk sac tumour may present with abdominal distension and pain and other constitutional symptoms of malignancy, the patients with yolk sac tumour of the cervix may present with persistent vaginal bleeding with or without attendant

symptoms of anaemia. Pelvic examination may reveal a palpable pelvic mass which may be obvious on either speculum examination or vagino-uteroscopy. The diagnosis of yolk sac tumour of the cervix is suspected following abnormal tumour markers such as Alpha feto Protein, Lactate dehydrogenase, and beta human chorionic gonadotrophin. The diagnosis is confirmed with biopsy for histology. Cross sectional imaging such as computed tomography, magnetic resonance imaging and positron emission topography scan of the chest, abdomen and pelvis are done to determine the extent of the disease for adequate staging.[10] The modality of treatment of yolk sac tumour of the uterine cervix can be chemotherapy, surgery and radiotherapy or combination of them depending on the stage of the disease.[11] Extragonadal yolk sac tumour staging is based on some prognostic factors such as the organ of involvement, involvement of the lung and the extent of tumour markers unlike the gonadal counterpart where tumour node and metastasis staging is used.[12] There may be treatment dilemma of patients with yolk sac tumour of the reproductive tract especially in patients that are yet to complete their family size or in female infants with great concern for future fertility. The options available for preservation of fertility as well as possibility of cure are primary chemotherapy and fertility sparing surgery as against radical surgeries. The chemotherapy regimen of choice are bleomycin, etoposide and cisplatin.[13] Follow-up is done to monitor relapse by periodic general physical examination, assay of tumour markers and imaging investigations. Extra gonadal germ cell tumour is extremely rare in the cervix and when it occurs there may be challenges in the management. We report this case due to its rarity and the possibility of cure by chemotherapy thus the potential for preserving fertility.

Case Presentation and Management

A 2-year-old female infant who presented at Federal Medical Centre Ebute Metta with a 9-month history of persistent bleeding and discharge per vagina. At first, the bleeding was once a month and progressively worsened on daily basis. There was associated passage of clots but no history of change in both urinary and bowel habit and no associated bleeding in any other part of the body. Furthermore, there was no history of trauma, abuse, weight loss, or difficulty in breathing.

On examination, she was healthy looking, not in distress, not pale, anicteric, afebrile, not cyanotic and well hydrated. The abdomen was soft with no palpable mass. A vaginal examination revealed pooling of blood in the vault. Every other system was clinically unremarkable through history and systemic examination

Pelvic ultrasound showed a bulky cervix containing a well-defined oval-shaped heterogeneously isoechoic mass measuring 46 x 35 x 44mm involving partly the uterus. A significant flow signal was seen within it on colour/power Doppler interrogation (Figure 1).

Magnetic resonance imaging (Figure 2) of the pelvis revealed a large (43 x 33 x38mm), well-circumscribed cervical mass displacing the uterus superiorly and rectum inferiorly. The mass was iso- and hyperintense on T1 and T2/STIR weighted sequences and demonstrated heterogeneous enhancement. No blooming was seen on gradient recalled echo. There was no fluid in the endometrial cavity but minimal fluid was seen in the vagina Both ovaries were normal. There was no pelvic lymphadenopathy. Chest radiogram did not reveal any suspicious findings. Laboratory investigations revealed the level of alpha-fetoprotein to be 3.92ng/ml. The haematological and biochemical results were essentially normal.

A core biopsy for histology during assessments under anaesthesia confirmed a yolk sac tumour (Figure 3). The diagnosis and options of treatment were explained to the patient relatives. There was a management dilemma as the patient was prepared for radical resection of the mass including total abdominal hysterectomy and bilateral salpingo-oophrectomy with or without adjuvant chemotherapy to guarantee cure. The patient relatives were not forthcoming with the procedure due to no chance of future reproduction. The patient was offered three cycles of cisplatin, etoposide and bleomycin. Post chemotherapy surveillance with ultrasound revealed no residual mass and alpha feto protein was negative as shown in Figure [illegible]e has been on regular follow up for 3 years with no clinical,laboratory and radiological evidence of relapse.

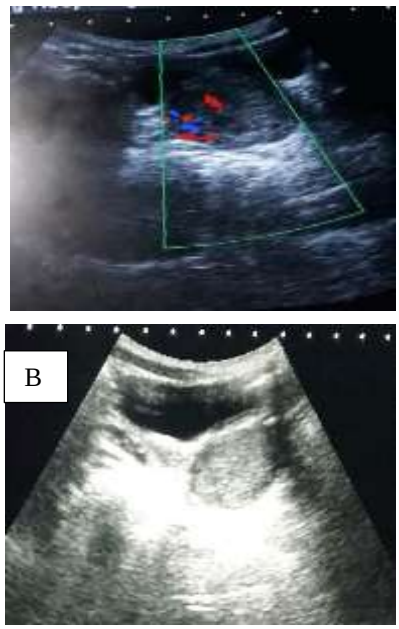

Figure 1: (A) Transabdominal pelvic ultrasound Longitudinal duplex sonogram, (B) transverse views how a heterogeneous solid isoechoic mass with well–defined regular margins (arrow) at the cervix, posterior to the urinary bladder. The significant flow was demonstrated within the mass on-power Doppler interrogation

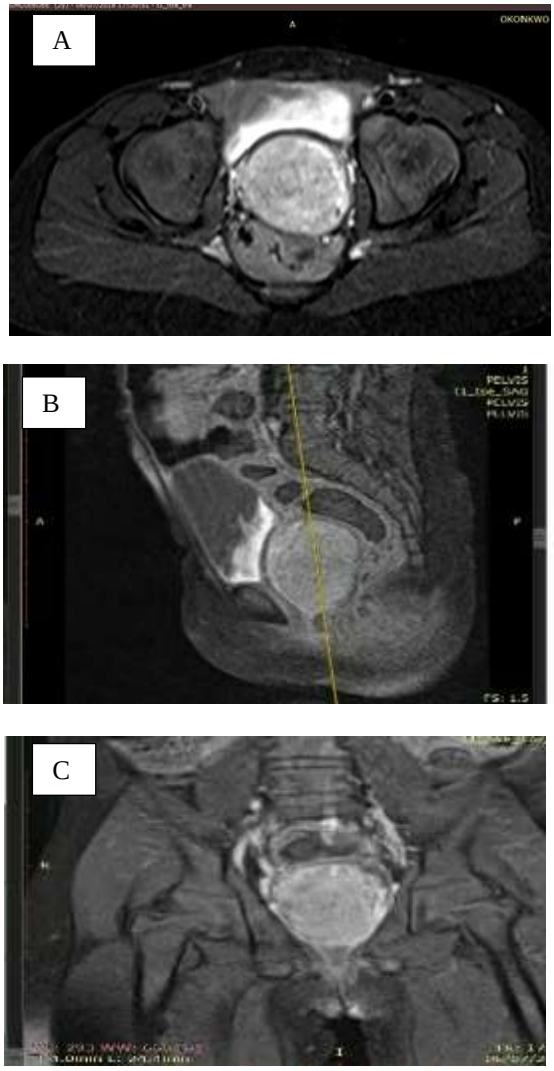

Figure 2: (A) Axial, (B) coronal and (C) sagittal contrast-enhanced fat-suppressed MRI images of the pelvis demonstrate an enhancing cervical mass, involving the uterine body superiorly and indenting the rectum. The fat planes around the cervix are preserved.

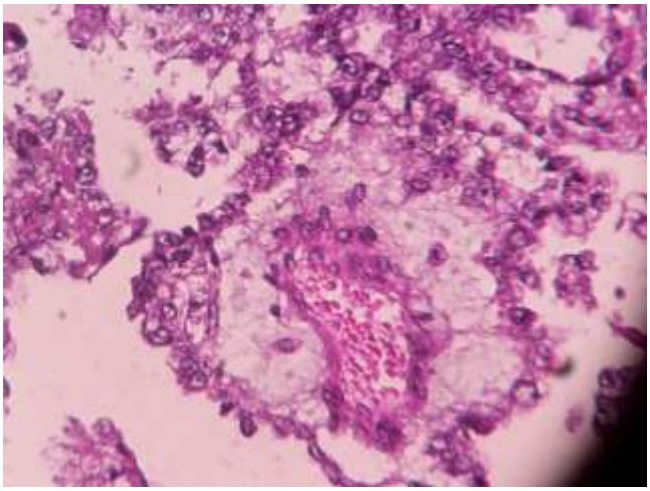

Figure 3: Section shows tumour tissue composed of malignant epithelial cells arranged in metacystic and nest patterns. The cells are round to oval with increased nuclear-cytoplasmic ratio, prominent nucleoli and scanty cytoplasm. A few of the cells are arranged radially around blood vessels reminiscent of Shiller Duval's body

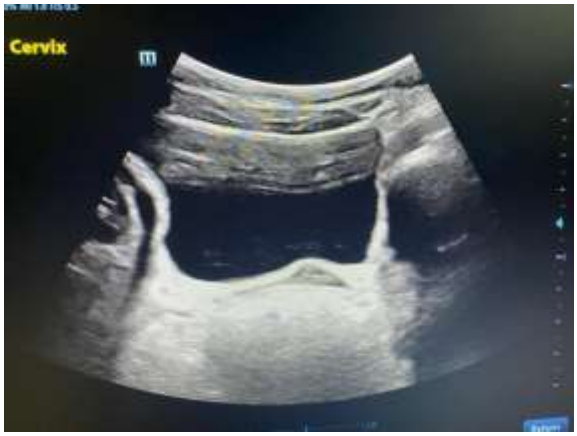

Figure 4: Transabdominal pelvic ultrasound with longitudinal and transverse view show complete resolution of the tumour

Discussion

We have illustrated our experience on the successful management of yolk sac tumour in a 2 years old female child. The management might be laden with challenges especially when fertility is at stake. We presented a case of yolk sac tumour of the cervix partly involving the uterus at Federal Medical Centre, Ebute Metta, Lagos State. Notably, uterine cervix is an uncommon site for a rare extra gonadal yolk sac tumour.[14] This case is one of the few that has been reported in the medical literature. A study in Zaria which examined 189 malignant childhood tumours only 15 children had germ cell tumours.[15] Williams also reported only 1 case of yolk sac tumour in a study spanning 13 years which examined 1325 cases childhood tumours in Ibadan, Nigeria.[14] Some of other genital sites that have been reported are vaginal, uterus and the penile shaft in male folks. More importantly, the most common germ cell tumour under the age of three years is yolk sac tumour. It has been reported that it has bi modal age distribution in which the first peak is seen before the age of one and the second peak at adolescence. This index case report did not differ from this age distribution.

The clinical features of extral gonadal yolk sac tumour depends on the site of the body that is involved hence the development of persistent vaginal bleeding in this case as it involved the cervix and partly the uterus. The diagnosis is usually suspected following raised tumour markers and advanced cross section imaging as done in this case. Positron emission topography-computed tomography scan is an ideal investigation of choice for metastatic work up.[16] This was not done in this case because information from abdominopelvic magnetic resonance imaging was sufficient. Elevation of alpha feto protein has been consistently linked with yolk sac tumour as against other tumour markers. This case was not an exception as the only detected tumour marker was alpha feto protein although it was normal. Others similar case series have reported raised alpha

feto prortein. This may be perhaps due to the degree of differentiation of the tumour. The well differentiated tumour may not elaborate high tumour marker. The diagnosis of yolk sac tumour of the uterine cervix is confirmed with histopathological examination of the mass resected following vagino-uteroscopy.

The primary treatment option for yolk sac tumour of the uterine cervix is controversial due to its rarity however, surgical resection of the mass with or without chemotherapy or radiotherapy is well known in the literature. The patient's relative declined due to desirous for future fertility. This was a big challenge considering the overall survival of the patient. A trial of chemotherapy not only potentially cured the disease but also enhanced the chance for future fertility. The follow-up of the patient has been unremarkable. This may not be unconnected with the good prognostic factors at presentation with normal alpha feto protein and no distant metastasis.

Conclusion

The 2-year-old female infant responded well to chemotherapy. This obviated the need for surgery. There was preservation of potential for future fertility. There was a big challenge considering the overall survival of the patient. A trial of chemotherapy not only potentially cured the disease but also enhanced the chance for future fertility. The follow-up of the patient has been unremarkable. This may not be unconnected with the good prognostic factors at presentation.

List of abbreviations

GCT – Germ cell tumours

Declarations

Ethical approval

None provided.

Availability of data and materials

The datasets used and/or analyzed during the current study are available from the corresponding author on reasonable request.

Competing interests

No conflict of interest associated with this work.

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Contribution of Authors

The authors declare that this work was done by the authors named in this article and all liabilities pertaining to claims relating to the content of this article will be borne by the authors. LOO (manuscript reviewing and editing; literature review, and editing and critical appraisal for intellectual content), BBO review of USS and CT Scan and critical reviewing, editing for intellectual content; AAS, review of CT San and critical reviewing, editing for intellectual content literature review and critical reviewing, editing for intellectual content. RMW (revising it critically for important intellectual content; manuscript preparation; final approval of the version to be published; and agreement to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved), OO; critical reviewing, editing for intellectual content literature review and critical reviewing, editing for intellectual content. OOF literature review and critical reviewing, editing for intellectual content.NAI; revising it critically for important intellectual content, literature review, abstract preparation, and discussion. All authors approved the final manuscript submitted for publication.

References

1. Kaatsch P, Häfner C, Calaminus G, Blettner M, Tulla M. Pediatric germ cell tumors from 1987 to 2011: incidence rates, time trends, and survival. Pediatrics. 2015;135(1):e136-e43.
2. Wefel JS, Vidrine DJ, Marani SK, Swartz RJ, Veramonti TL, Meyers CA, et al. A prospective study of cognitive function in men with non-seminomatous germ cell tumors. Psycho-oncology. 2014;23(6):626-33.
3. Rougemont A-L, Tille J-C. Role of HNF1β in the differential diagnosis of yolk sac tumor from other germ cell tumors. Human pathology. 2018;81:26-36.
4. Samaila MO, Maitama HY, Abdullahi K, Mbibu H, Waziri GD. Yolk sac tumour of the penile shaft: a rare primary extra-gonadal presentation. African Journal of Paediatric Surgery. 2011;8(2):241.
5. Dehner LP, editor Germ cell tumors of the mediastinum. Seminars in Diagnostic Pathology; 1990.
6. Schneider DT, Calaminus G, Koch S, Teske C, Schmidt P, Haas RJ, et al. Epidemiologic analysis of 1,442 children and adolescents registered in the German germ cell tumor protocols. Pediatric blood & cancer. 2004;42(2):169-75.
7. Stang A, Trabert B, Wentzensen N, Cook MB, Rusner C, Oosterhuis J, et al. Gonadal and extragonadal germ cell

tumours in the United States, 1973–2007. International journal of andrology. 2012;35(4):616-25.
8. Bonouvrie K, van der Werff ten Bosch J, van den Akker M. Klinefelter syndrome and germ cell tumors: review of the literature. International Journal of Pediatric Endocrinology. 2020;2020:1-7.
9. Pawar NP, Mahajan SV, Chaudhari RA, Chavan SD. Extragonadal GCT: A rare case report of sacrococcygeal pure yolk sac tumor. Indian Journal of Pathology and Microbiology. 2013;56(3):329.
10. Rescorla FJ, editor Pediatric germ cell tumors. Seminars in pediatric surgery; 2012:
11. Göbel U, Schneider D, Calaminus G, Haas R, Schmidt P, Harms D, et al. Germ-cell tumors in childhood and adolescence. Annals of oncology. 2000;11(3):263-72.
12. Rudaitis V, Mickys U, Katinaitė J, Dulko J. Successful treatment of advanced stage yolk sac tumour of extragonadal origin: a case report and review of literature. Acta medica Lituanica. 2016;23(2):110.
13. Rouge TdLM, Pautier P, Duvillard P, Rey A, Morice P, Haie-Meder C, et al. Survival and reproductive function of 52 women treated with surgery and bleomycin, etoposide, cisplatin (BEP) chemotherapy for ovarian yolk sac tumor. Annals of oncology. 2008;19(8):1435-41.
14. Clement PB, Young RH, Scully RE. Extraovarian pelvic yolk sac tumors. Cancer. 1988;62(3):620-6.
15. Samaila MO. Malignant tumours of childhood in Zaria. African Journal of Paediatric Surgery. 2009;6(1):19.
16. Ben-Haim S, Ell P. 18F-FDG PET and PET/CT in the evaluation of cancer treatment response. Journal of Nuclear Medicine. 2009;50(1):88-99.