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A 19 Month's Old African Female with Gluteal Yolk Sac Tumour: A Case Report and A Review of Literature

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ABSTRACT
Primary Yolk sac tumour (YST) of extragonadal site is a very rare tumour, usually arising from malignant transformation of misdirected primordial germ cell on transit from the yolk sac to gonads. The present study reports a primary YST of the gluteus in a 19 years old African child presenting with acute urinary retention and constipation. Complete tumour excision was done resulting in relief of urinary and bowel symptoms. In the face of limited investigation tools, light microscope observation of Schiller-Duvall bodies in histopathology specimens, as seen in this case, is diagnostic. Complete excision with Platinium-based chemotherapy has been shown to gives excellent 5 year survival. Alpha fetoprotein (AFP) is useful marker in monitoring response to therapy. Poverty and unavailability of advanced diagnostic tools are however major challenge in diagnosis of atypical cases in Africa.
Key word: Yolk sac tumour, Gluteal mass, Histology

INTRODUCTION
Germ cell tumours are tumours derived primordial from germ cells. This heterogenic group of tumours can be either benign or malignant.[1] The malignant germ cell tumours include the geminomas(dsysgeminoma/seminomas) and the nongeminomas(endodermal sinus tumour/yolk sac tumour[YST], choriocarcinoma, embryonal carcinoma and immature teratoma).[1,2] These tumours account for 3% of all childhood cancers, out of which YST is the most common histologic type.[3] They most frequently arise within the gonads (testes and ovaries), but are rarely encountered elsewhere accounting for 10-15% of the extra-gonadal cases, in which there is no evidence of primary tumour in either the ovary or testes.[4,5] These extragonadal YST are usually seen along the body midline, including the urachus, spinal cord, vagina, abdominal wall, mediastinum, head and neck region, gluteus, brain, stomach, prostate gland, corpus uteri, ear and liver.[5] Here we present an YST of the gluteal region involving a 19 months female.

Case Report
A 19 months female, of African descent presented with a gluteal swelling of 3 months duration, acute urine retention and constipation of 3 days duration. The swelling was first noticed in the left gluteus, which gradually increased in size and extended to the right gluteus. It was initially painless but became progressively painful over time. The child had an unremarkable birth history and no significant past medical history. There was no history of vomiting or chronic weight loss. Physical features appeared otherwise normal and appropriate for the age. There was a gluteal mass predominantly on the left buttock with extension into the right, measuring 26cm by 18cm by 8cm. The mass was tender, firm to hard in consistency, fixed to underlying muscle but not to overlying skin. The abdomen was tense and distended with an umbilical hernia. Digital rectal examination revealed a patulous anus, loss of anal sphincter tone and a mass felt almost occluding the rectum.

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Ultrasound scan showed an irregular, non-homogenous, gluteal solid mass, impinging on the rectum and sigmoid colon, and mild bilateral hydronephrosis. No lesion was seen on chest x-ray. A preoperative clinical diagnosis of teratoma was made.

Surgery was performed with complete excision of the tumour and os coccygium. The tumour was submitted to the histopathology department in 10% neutral buffered formal saline. Urine and stool habit returned to normal on the 3rd day post-operation and the post-operative condition was stable. Gross examination of the surgical specimen showed gray-white, soft, hemorrhagic tissue. Histologic examination showed a tumour with papillary-cystic structure with numerous schiller-duval bodies. The resection margins were positive for tumour cells. The features were in keeping with diagnosis of YST. Patient's parents were informed of the diagnosis while the patient was worked-up for chemotherapy.

DISCUSSION

YSC or endodermal sinus tumour was first described in English literature by Telium in 1959,and was named because of its remembrance with yolk sac.[6] Since them more cases of varying histological types has been reported, mostly in the testes and ovary.[4,5] There is no general acceptance of the histogenesis of extragonadal YST. Two theories have been proposed by earlier researchers to explain this. In one of the theories, it has been proposed that these tumour arise from abnormal differentiation of some somatic cells. This theory may offer explanation to the occurrence of these tumours in the stomach, and endometrium. The second theory is that they arise from malignant transformation of misplaced primordial germ cells on migration from yolk sac to the gonads. This offers explanation to YST occurring in the midline structures such as sacrococcyx, peritoneum, abdominal wall, mediastinum and head and neck region.[7] The location of extragonadal YST tends to vary with the age and gender of the child. It is a general observation that gluteal YST almost always occur among children younger than 3 years, [8,9,10] while mediastinal tumours is commoner occur in young adults.[11] In the index case, the tumour was first noticed by the parents at the age of 16 months. It also grew to a massive size within 3 months, supporting the earlier report that it is a highly malignant, rapidly growing tumour.[12,13] We noted that there was relief of urinary and bowel obstruction on the third day after surgery. The underlying pathophysiology of the obstruction is therefore mechanical compression of the abdominopelvic viscera by the gluteal tumour. The bilateral hydronephrosis in this report is also the effect of chronic obstructive effect of the mass on the urinary system. Consequently, resection of the tumour relieved the pressure, with corresponding alleviation of the symptoms. It is also an indication that tumour metastasis was not the likely cause of the obstructive symptoms.

Grossly, YST usually appear as large solid, fat-free gray-white mass, with myxoid, mucoid or gelatinous external appearance. Cut section usually show areas of necrosis, hemorrhage and cystic changes are often seen within the tumour.[14] These features were also seen during surgical pathology evaluation of the tumour specimen. Imaging tools useful in diagnosis of YST include computed tomography (CT), magnetic resonance imaging (MRI), ultrasound scan (US), positron emission tomography (PET) scanning, and plain X-rays. These imaging devices are valuable in assessing the position, extent, invasiveness and secondary changes in adjacent structures, but are unable to demonstrate

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the tumour type.[15] In resource poor settings, CT and MRI are rarely available because of inherent poor funding of health sector by government. US and plain-X ray facilities are however easily affordable and were useful in this case.US scanning showed that the tumour was a fat-free, soft non-homogenous mass with areas of necrosis and cystic changes while the x-ray excluded lung metastasis.

As with other malignant germ cell tumours, histopathologic analysis and immunohistochemistry are important in demonstration of the tumour type and prognostic index. Eight decades after its initial description, different histological patterns has been described by other researcher including reticular, polyvesicular, hepatoid, glandular, papillary, myxomatoid, macrocystic, and solid patterns. Most times however, mixed histological patterns were observed these tumours especially in adult case. Typically the tumour cells have hyperchromic, irregular, pleomorphic nuclei, with frequent mitosis. Schiller-Duval bodies (SDB) is the diagnostic hallmark and is more common with the endodermal sinus sub-type. Morphologically, consists of glomeruloid, epithelial-lined space, containing a polypoid projection, covered by cuboidal to columnar cells with a single central vessel. [14,16] SDB is however only seen 30% of YST, and its absence does not in any case rule out YST as long as there are other histological features suggestive of it. [11] In our case, the presence of SDB was substantial for making diagnosis of YST. Other common interesting histologic feature is the presence of globular hyaline bodies. These hyaline bodies can be easily highlighted using Periodic Acid Schiff stain (PAS). In atypical cases however, other ancillary investigations such as immunohistochemistry are applied to confirm diagnosis. Useful immunohistochemistry markers in diagnosis of atypical YST include AFP, α-1 antitrypsin, glypican-3 and salt-like protein-4. Positive AFP staining is cytoplasmic and may be diffuse or focal while positive α-1 antitrypsin staining cytoplasmic staining is usually focal, and both are considered to be sensitive and specific for YST.[14,17] Sal-like protein 4 is a zinc finger transcription factor that has been identified to be highly specific and specific in differentiating YST from other non-yolk sac germ cell tumours.[17] Epithelial membrane antigen, which is specific and sensitive for carcinomas, is negative is YST.[14] Ki67 is a proliferative marker and is useful in a assessing the prognosis of the tumour.[14] Immunohistochemical services are still scarce in our environment and where available, only few antibodies are readily available , frustrating its use as a diagnostic tool.

Serum tumour markers particularly AFP, human chorionic gonadotrophin HCG and lactate dehydrogenase LDH are useful tool in timely diagnosis, staging, monitoring of tumour burden, treatment success and recurrence of these germ cell tumours. Of these, frequency of elevation correlates with tumour burden and histologic type. Studies has shown that LDH is elevated in 40-60% of all GCT, while AFP is elevated in 10-60% of non-seminomatous GCT.BHCG on the other hand is elevated in 15-20% of seminomatous GCT and in 10-40% of non-seminomatous GCT.[18] AFP is however found in fetal serum and in new born, the level drops to normal adult level at 8th month post-partum, so its presence after such age is invariably a sinister sign.[19] In the index case, these markers were not done not used in diagnosis because teratoma was the initial

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clinical consideration. Treatment is dependent of the extent of the tumour. Total excision may be appropriate in local tumour (stage 1) accompanied with serum AFP and beta-HCG serial monitoring. In high-risk cases, chemotherapy using a combination of bleomycin, etoposide and cisplatin is the treatment of choice. Because the tumour is highly chemosensitive, 5-year survival of up to 90% has been documented with this platinum-based therapy.[20] In our index-case, patient is being worked up for chemotherapy. However, the major anticipated fear is the risk of absconding from treatment because patient's parents are very poor and payment for cancer treatment in Nigeria is out of pocket. There is therefore need for more health insurance coverage to prevent such cases in the future. In conclusion, this paper presents a case of primary YST of the gluteus in a 19 year old African child. It is important to suspect such tumour in any gluteal mass in any child under the age of 3 years with rapidly growing midline mass. Histopathology is the gold standard for diagnosis for typical cases(with SDB). Atypical cases are confirmed using immunohistochemical markers. AFP is useful in monitoring therapeutic progress. With cisplatin-based chemptherapy, up to 90% 5 year survival is achievable.

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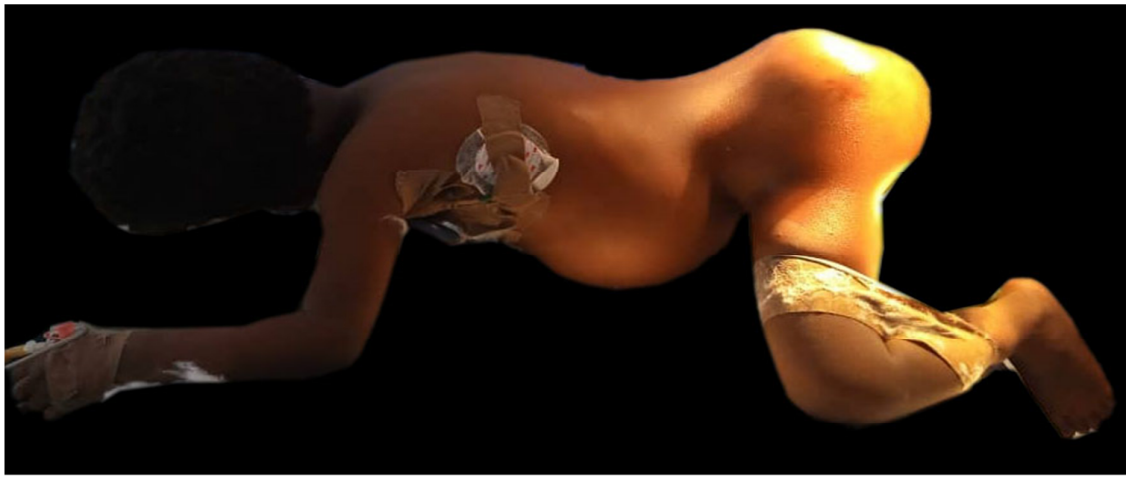

Figure I: 19 months old female with gluteal mass

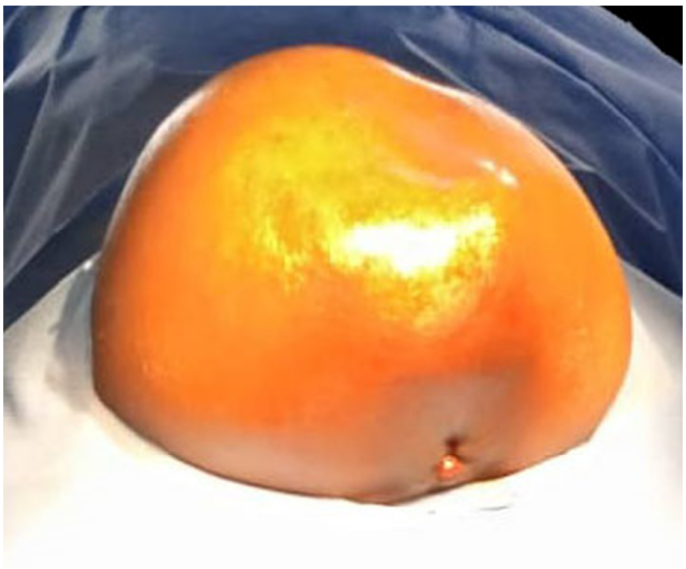

Figure: Dorsal view of gluteal mass

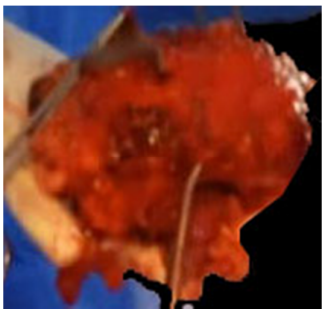

Figure III: Excised tumour mass

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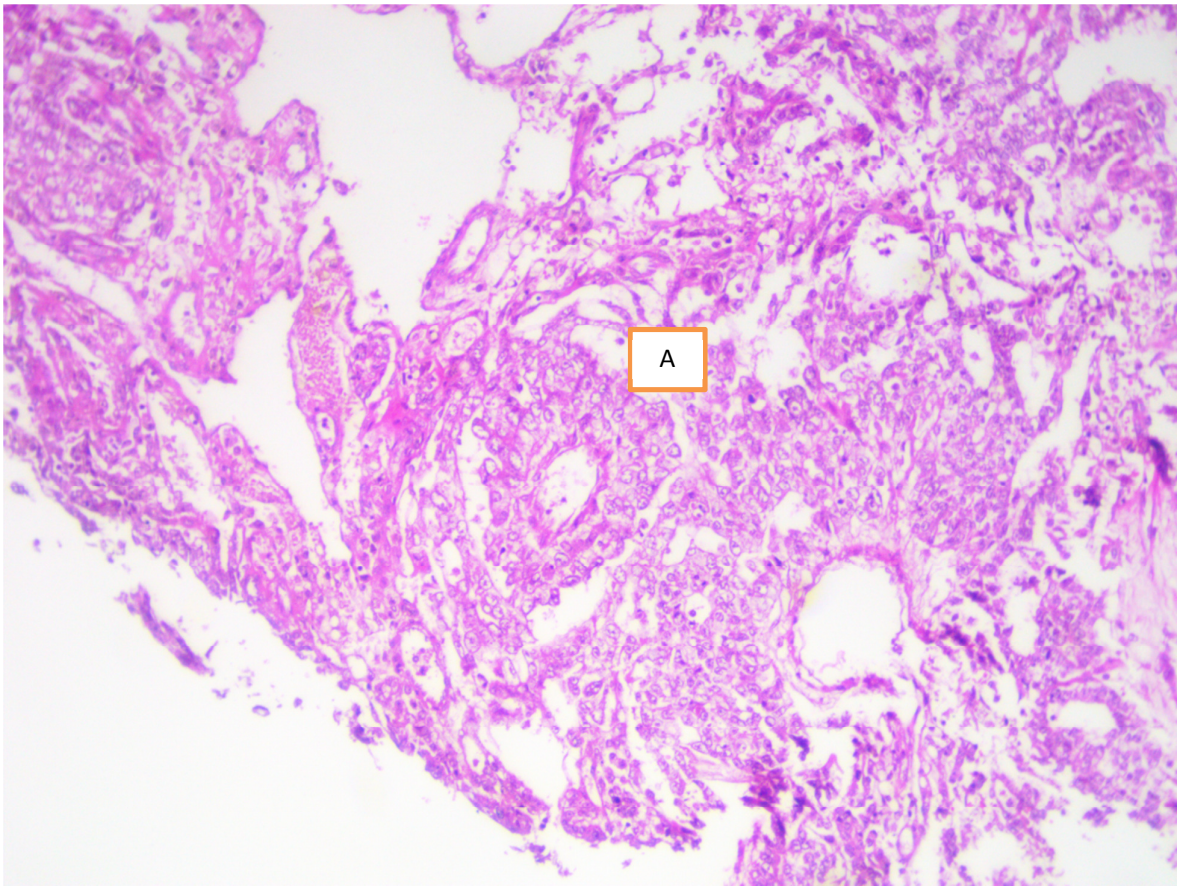

Figure IV: Histology of YST(x10) showing Schiller-Duval body (A)

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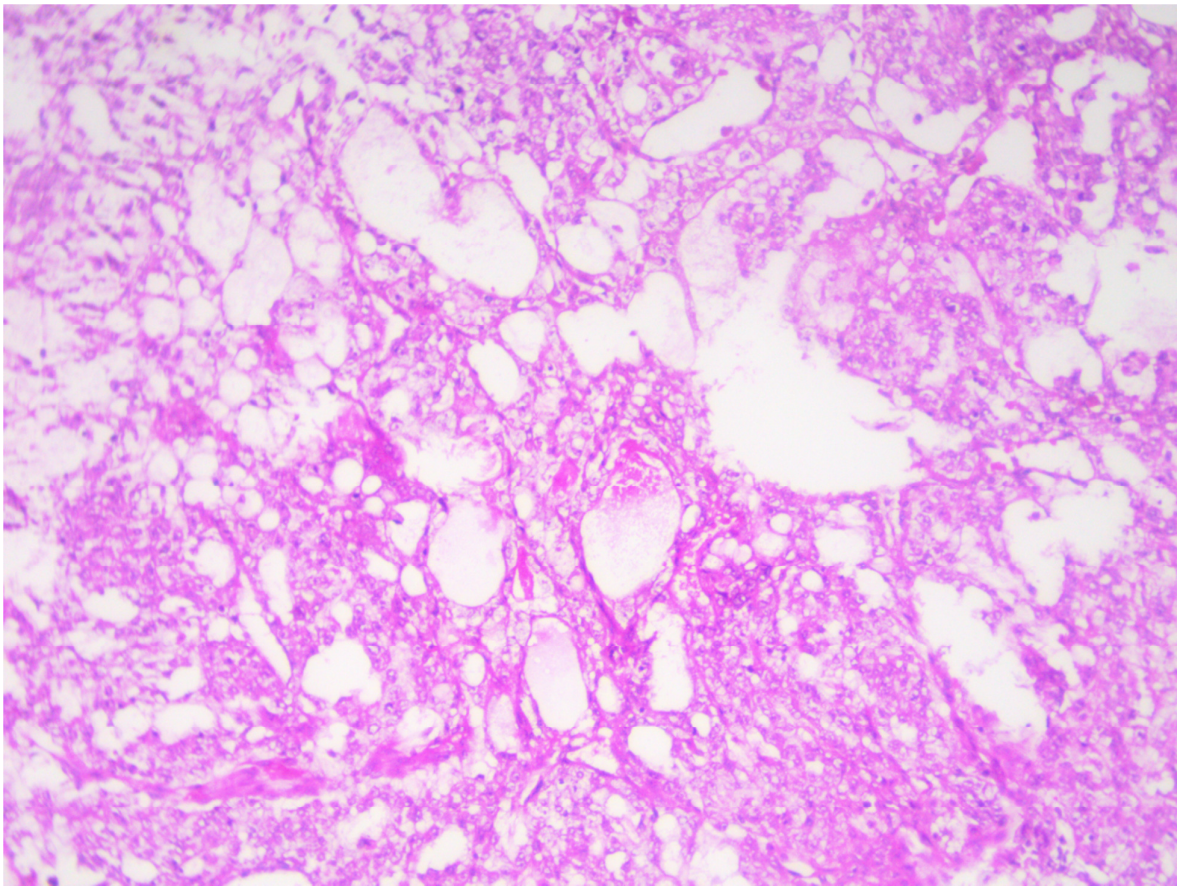

Figure V: Tumour mass showing Microcystic pattern