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CASE REPORT

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Sacrococcygeal teratoma in a newborn : A case report

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Abstract: *Background:* Sacrococcygeal teratoma is an extragonadal teratoma arising from multipotent stem cells from the Hansen node located around the coccyx. In our setting, the neonatal and pediatric surgery units each have two documented cases of sacrococcygeal teratoma per year. This gives an estimated annual incidence of 1 case per 100,000 live births in Dares Salaam. Given the rarity of the condition and because early diagnosis and surgical intervention improve the overall outcome
Case Presentation: We present a 2 -day-old female baby who was referred to our facility with a gluteal mass. After the initial workup, surgical resection and biopsy confirmed a teratoma. The surgery was done successfully and the child fully recovered with an excellent outcome.
Conclusion: Early recognition, referral to the appropriate center, and intervention averts morbidity and mortality and improves the quality of life in neonates with sacrococcygeal teratoma. Serial Alfa Fetoprotein measurements are a good indicator to monitor progress.

Key Words: Sacrococcygeal Teratoma, Resource limited settings, Early intervention.

Résumé: *Contexte:* Le tératome sacrococcygien est un tératome extragonadique provenant de cellules souches multipotentes du ganglion de Hansen situé autour du coccyx. Dans notre établissement, les unités de chirurgie néonatale et pédiatrique comptent chacune deux cas documentés de tératome sacro-coccygien par an. Cela donne une incidence annuelle estimée à 1 cas pour 100 000 naissances vivantes à Dar es Salaam. Compte tenu de la rareté de l'affection et du fait qu'un diagnostic et une intervention chirurgicale précoces améliorent le résultat global, nous présentons le cas d'un bébé de sexe féminin âgé de 2 jours qui a été adressé à notre établissement pour une masse fessière. Après le bilan initial, la résection chirurgicale et la biopsie ont confirmé la présence d'un tératome. L'opération a été réalisée avec succès et l'enfant s'est complètement rétabli avec un excellent résultat.
Conclusion: Le dépistage précoce, l'orientation vers le centre approprié et l'intervention permettent d'éviter la morbidité et la mortalité et d'améliorer la qualité de vie des nouveau-nés atteints de tératome sacro-coccygien. Les mesures en série de l'alpha-fœtoprotéine sont un bon indicateur pour suivre l'évolution.

Mots clés: Tératome sacro-coccygien, ressources limitées, intervention précoce.

Introduction

Sacrococcygeal teratoma is a congenital germ cell tumor originating from multipotent stem cells concentrated around the cervix. It is the most common newborn tumor occurring in 1 for every 40,000 births.[1] SCT is more common in a female with a female-to-male ratio of 3:1.[2] Histologically SCT can be mature containing well-differentiated tissues, immature with incomplete differentiation, or may contain malignant elements that are usually associated with an elevated level of tumor markers such as Alpha-fetoprotein (AFP).

Sacrococcygeal teratomas occurring in infancy rarely contain malignant elements when compared to those of adult-onset. The likelihood of developing SCT with malignant elements increases with an increase in post-natal age. Nevertheless, early diagnosis is crucial for the best outcomes. This includes imaging to know tumor extent that guides surgical intervention and measure of tumor markers such as Alpha-fetoprotein (AFP).
We report a case of a newborn female with SCT as it is rarely encountered in our center, more so to emphasize early referral to centers with surgical expertise for early intervention such as the one done to our patient in preventing morbidity and mortality and improving the quality of life. SCT has the potential for malignant transformation and death from metastasis,[3,4]the tumor contributes to mass effect on surrounding structures causing constipation, urine retention, and hydronephrosis.[5]

Case presentation

A two-day-old female baby was referred to our facility from a primary health facility in Dar es Salaam due to a mass on the gluteal region noted at birth.
The baby was born by spontaneous vertex delivery at 38 weeks of gestation with 2.6 kg and had an Apgar score of 8 and 10 in the 1st and 5thminute, respectively.
The mother is Para 2, 31 years of age while the father is in his mid 30's. There was no consanguinity. Mother started the antenatal clinic late in the second trimester and this is when she started using folic acid and antimalaria prophylaxis. A routine second-trimesterobstetric ultrasound was normal. There was no history of alcohol use, cigarette smoking, or substance abuse by the mother during pregnancy. Had yellowish per vaginal discharge in the second trimester with positive serology for VDRL. She was treated and the vaginal discharge resolved. HIV serology was negative, she was normotensive and euglycemic throughout the pregnancy.

On admission, the baby was alert with a respiratory rate of 44 breaths/min, oxygen saturation of 99% on room air, Pulse rate of 144b/min, and temperature of 36.8^{o}C.
She had a mass extending from the sacral area to involve the gluteal region measuring 15 cm by 10 cm. (Fig 1a, 1b, 1c). Had mixed consistency where some parts of the mass were firm while other regions were soft, non-fluctuating, and mobile, with the normal surrounding skin, no ulceration with a negative transillumination test. Had a patent anal opening with normal female genitalia. There were no other malformations or dysmorphic features noted on examination.
The occipital circumference was 32.8 cm and the length was 49 cm.
A provisional diagnosis of sacrococcygeal teratoma was made.

Fig1a

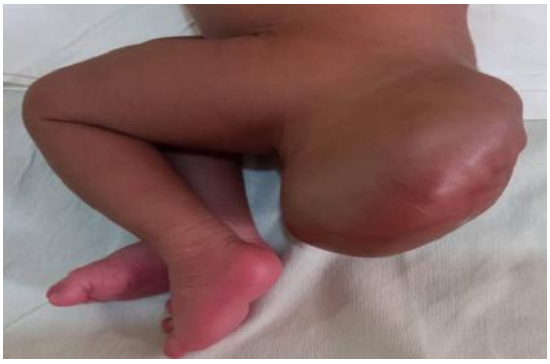


Fig 1b

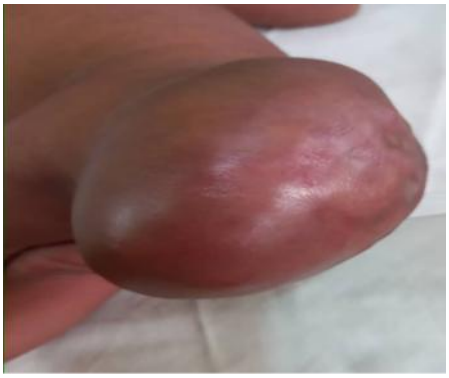
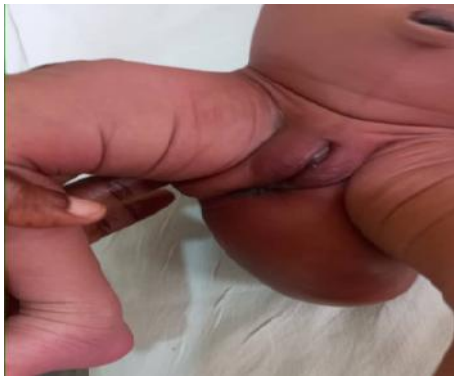


Fig 1c



Investigations

Magnetic Resonance Imaging of the mass was performedwhich revealed a hanging sacral mass measuring (8.5x5.02x8.88) cm. The lesion had mixed intensity and on T1W, fatty components appeared with a high signal and calcific/bony components with a low signal. On T2W, fluid (cystic) components appeared with high signal,and calcific bony components with low signal. The mass had no colonic displacement or ureteric dilatation. It was not associated with hip displacement, intraspinal

extension, or vaginal dilatation. There was no involvement of the right and left sacral iliac joints and muscle planes were maintained. Rectum had unremarkable findings. This is tallied with the diagnosis of sacrococcygeal teratoma. (Fig 2a and 2b)
Alpha-fetoprotein (AFP) on day four of life was 3430.88 and decreased to 1662.73 on day nineteen and was 114.1 at 4 months of age. Beta human chorionic gonadotrophin (beta HCG) was 1.44 on day four of life.
Echocardiogram revealed a structurally normal heart.

Fig 2a: T1W MRI coronal section. A well-defined mass of heterogeneous signal intensity ranging from low signal, intermediate to high signal in keeping with fluid, calcific and fat components respectively.

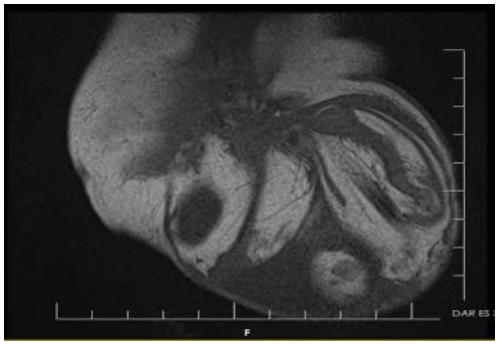
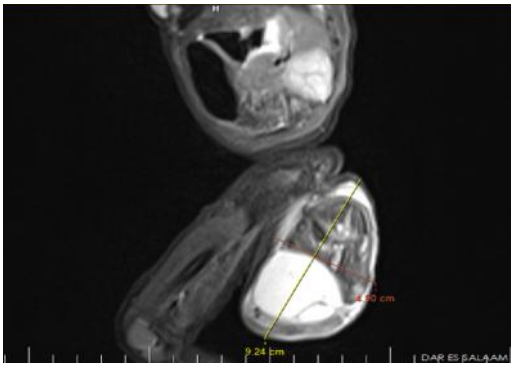


Fig 2b: T2W MRI sagittal plane. A well-defined mass in the sacrococcygeal region containing predominantly high signal intensity consistent with a fluid containing mass and intermediate and low signal intensity component consistent with the calcific component.



Differential diagnosis

A differential diagnosis of distal neuro tube defect (Sacral myelomeningocele) was made.

Treatment

The patient was reviewed by the pediatric surgery team and the excision of the mass was done successfully. Intraoperative findings revealed sacrococcygeal mass Altmann class II measuring 12 by 7 cm extending to the pelvis more prominent on the left side with the right side spared. The tissue was sent for histology.

Histology results

Macroscopic findings: Piece of tissue covered by skin measuring 12 by 10 by 5 cm. O/s solid areas, cystic mass, skin, and cartilaginous areas were noted.
Microscopic findings: A tissue section from sacrococcygeal areas showed abundant lobules of adipose tissue and a cystic lesion lined by stratified squamous epithelium with abundant laminated keratin pearls. There were focal areas that showed smooth muscles, skin adnexa, gastrointestinal lining with all the layers, and cartilaginous matrix. No features of atypia. The margins were free of tumors.

Conclusion: Sacrococcygeal mass, mature teratoma.

Fig 3: Image taken post-tumor excision

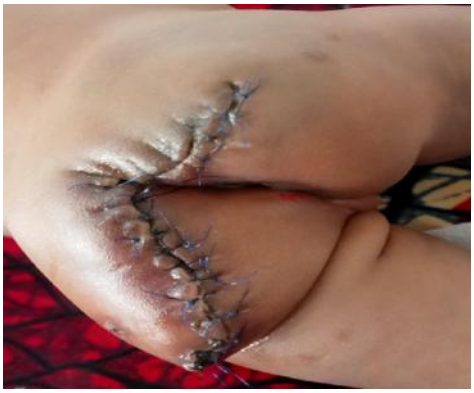


Fig 4a: Low power view showing lobules of mature adipocytes (black arrows) which are separated by fibro-cellular strands (Hematoxylin and eosin stain, 40X magnification).

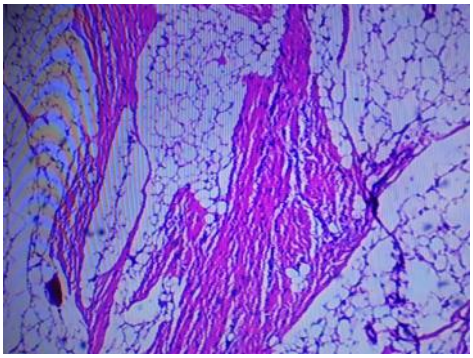


Fig 4b: Low power view showing benign intestinal glands (black arrow) with reactive submucosal lymphoid aggregates (red arrow). The lamina propria is expanded by lymphocytic infiltrates. Other areas show edema and vascular ectasia (Hematoxylin and eosin stain, 40X magnification).

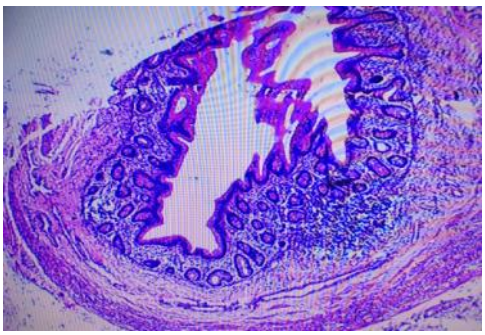
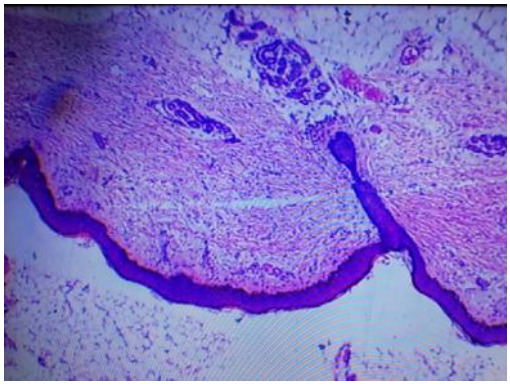


Fig 4c: Low power view showing a cystic lesion(black arrow) lined by epidermis with adnexal structures and the adipose tissue seen in the deeper layer (red arrow). The cystic lumen is filled with abundant amorphous materials.(Hematoxylin and eosin stain, 40X magnification).



Outcome and follow-up

The patient will be followed up monthly initially for the first year. Monthly alpha-fetoprotein will be measured. Magnetic resonance imaging at the primary site of the tumor will also be repeated at 3- and 12-monthspost-excision.

Discussion

Sacrococcygeal teratoma is a germ cell tumor arising from multipotent cells from the Hansen node that is located on the coccyx. It is the most frequently encountered tumor in newborns.[6]
It is caused by the failure of regression of totipotent cells in the primitive streak after the formation of the intraembryonic mesoderm. However, there are no consistent genetic alterations that have been linked with SCT.[4]
Our patient was female and it has been shown sacrococcygeal teratoma to be more common in a femalewith a female-to-male ratio of 3 to 4:1.[2]

SCT contains all three germ layers (Ectoderm, mesoderm, and endoderm). Histologically they can be classified into mature, immature, or malignant SCT.
Mature sacrococcygeal teratoma contains fully differentiated tissue which can be bone, glandular tissue, teeth, and hair. This account for at least half of all SCT.[8]Immature SCT contains tissues that are not fully differentiated.
Histological results in this patient revealed mature teratoma in keeping with other studies that have shown a majority of sacrococcygeal teratoma in neonates to be mature.[9,10]

Malignant SCT contains malignant elements with the yolk sac being the most common malignant element and associate delevation of alpha-fetoprotein.[2]Other malignant elements can be from the chorion.
SCT can be cystic or mixed cystic and solid with calcifications. This was also seen in this patient.
According to the American Academy of Pediatric Surgery, SCT can be further classified based on the extent to which the tumor is internal or external(Altman classification). With only a minor presacral component, Type I tumors are primarily external (sacrococcygeal); Type II tumors present externally but have a sizable intrapelvic extension; Type III tumors are visible externally but the majority of the mass lies in the pelvis and extends into the abdomen, while Type IV tumors only develop in the pelvis.[11]Type 4 usually has a late diagnosis when compared to other types as they lack an external component.[8]This can cause compression of organs such as the urinary bladder and rectum. This patient had Altman class II tumor classification.

Differential diagnosis includes distal neuro tube defects such as myelomeningocele. Clinically they can present with a sacral mass such as that seen in SCT.
Magnetic resonance imaging is the modality for confirming the diagnosis. It aids in identifying the extent of the tumor hence giving the Altman type. It also aids in distinguishing SCT from distal neuro tube defects.
Serum alpha-fetoprotein(AFP) and beta human chorionic gonotrophic (B-HCG) are usually elevated in malignant SCT. Their interpretation however in SCT should be done with caution because this tumor presents at birth where AFP and B-HCG are high. The levels of AFP in this patient based on age were within the normal range, this is consistent with a prospective study done by Yadav DK et al where all the patients aged less than one month had AFP levels appropriate for age.[9]
The level of alpha-fetoproteinis typically high at birth and gradually declines to reach normal levels between 8 and 12 months of age.[12] This was observed in our patient where there was a serial drop in AFP levels. At the age of four months, AFP was within the normal range (114.1ng/ml).

In the majority of SCT in newborns, treatment is by surgical resection. The surgery usually extends to remove the coccyx, and this was done in this patient.
Malignant SCT is staged after surgery using Children's Oncology Group (COG) staging guideline, neoadjuvant and adjuvant chemotherapy is included in treatment depending on the stage of the malignant tumor.[13,14]
A systematic review has shown the following to be associated with higher in-uterosurvival of SCT; tumor size with tumors less than 10 cm, cystic tumors, mature tumors, and absence of hydrops fetalis.[15]

Post-surgical complications include bowel or urinary incontinence and constipation.

Follow-uppost-surgery includes monthly measurement of AFP and lactate dehydrogenase for the first year. MRI at the site of resection is done at 3 months and 1-yearpost-surgery, thereafter imaging is performed as necessary to assess new symptoms.
The parents of the infant perspective: The fact that my baby was born with this abnormality caught me off guard, not only because it was missed during my pregnancy's ultrasound but also because, despite seeing and

hearing about children born with malformations on television and the radio, I had never dreamed it would happen to me. But now that I have heard the physicians describe the condition and learned that my baby will need surgery, I'm calmer than I was at first.

Conclusion

This is among the few reported cases of Sacrococcygeal teratoma from LMIC highlighting the importance early referral. Serial measurements of Alfa Fetoprotein post-operatively is a good marker to indicate that there is no residual mass and the interventional has been successful.

Authors' contributions

All authors contributed in the writing of Manuscript.
EE : Initiated the first draft and obtained statistics from the hospital.
AN : Performed the Histopathological examination and reviewed the manuscript .
ZB: Was the Pediatrics Surgeon.
KM : Initiated the first draft, supervised, reviewed .
All authors reviewed and approved the final manuscript.

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