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Case Report

Mature cystic teratoma in a 12-year-old with situs inversus totalis: a case report in a Nigerian teaching hospital

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ABSTRACT

Mature cystic teratomas are the most common benign ovarian tumors in children, adolescents, usually composed of mature tissues from all three germ layers. Situs inversus totalis (SIT) is a rare congenital condition characterized by complete mirror-image reversal of thoracic and abdominal viscera. Coexistence of these two entities is extremely rare, with only a handful of cases reported worldwide. We describe a 12-year-old girl who presented to the University of Medical Sciences Teaching Hospital, Ondo, Nigeria, with progressive abdominal swelling of eight months' duration, associated abdominal pain, weight loss, anorexia, and early satiety. Clinical and radiological evaluation revealed a huge heterogeneous ovarian mass with incidental findings of dextrocardia, left-sided liver and gallbladder, and right-sided spleen, consistent with SIT. She underwent exploratory laparotomy which demonstrated a lobulated left ovarian mass measuring 30×28 cm, moderate ascites, and complete visceral reversal. Left oophorectomy was performed. Histopathology confirmed a mature cystic teratoma. Her postoperative recovery was uneventful, and she remained well at follow-up. Mature cystic teratomas are common ovarian tumors in young females, but their association with SIT is exceptionally rare. The reversed anatomy poses diagnostic and surgical challenges, and careful preoperative evaluation is crucial. Complete excision remains the treatment of choice, with an excellent prognosis. This case appears to be the first documented report from Nigeria and contributes to the scarce global literature on this unusual association. This case highlights the importance of considering rare anatomic anomalies in surgical planning and underscores the need for thorough reporting of such cases in sub-Saharan Africa.

Keywords: Mature cystic teratoma, Situs inversus totalis, Ovarian tumor, Nigeria, Case report

INTRODUCTION

Teratomas are germ cell tumors that arise from pluripotent stem cells capable of differentiating into derivatives of the ectoderm, mesoderm, and endoderm.[1,2] They may develop in gonadal sites, such as the ovaries and testes, or in extragonadal locations including the sacrococcygeal region, mediastinum, and retroperitoneum. Ovarian teratomas are among the most frequently encountered tumors in children and adolescents, where mature cystic teratomas, also known as dermoid cysts, predominate.[3] These tumors are benign in the majority of cases, representing about 20-30% of ovarian neoplasms in this age group.[4,5] Histologically, they are characterized by mature tissues such as squamous epithelium, cartilage, bone, or adipose tissue, and although typically indolent, they may rarely undergo malignant transformation, reported in 1-2% of cases.[6]

SIT is an uncommon congenital anomaly in which the thoracoabdominal organs are arranged in a mirror-image reversal of the normal anatomic configuration.[7] It is

thought to result from abnormal embryologic development of left-right asymmetry during gastrulation, with an estimated incidence of between 1 in 8,000 and 25,000 live births.[8] Although usually asymptomatic, SIT is important to recognize as it complicates both clinical diagnosis and surgical interventions due to reversed anatomy.[9] It may occur as an isolated anomaly or be associated with syndromes such as Kartagener's, which includes chronic sinusitis and bronchiectasis.[10]

The association of SIT with ovarian teratomas is exceedingly rare, with only a few cases documented globally.[11-13] The co-occurrence raises important considerations for radiological interpretation and surgical management. To our knowledge, no such case has previously been reported in Nigeria. We present the case of a 12-year-old Nigerian girl with a huge ovarian mature cystic teratoma in the setting of SIT. This case emphasizes the diagnostic value of modern imaging, the importance of intraoperative awareness of anatomic variations, and the need to report rare pathologic associations to enrich regional and global literature.

CASE REPORT

A 12-year-old girl presented at Gynaecology Clinic of the University of Medical Sciences Teaching Hospital, Ondo with an 8-month history of progressive abdominal swelling, 6 6-month history of intermittent abdominal pain, and 4-month history of anorexia with associated weight loss. She also reported occasional early satiety but no vomiting, jaundice, bowel obstruction, or urinary symptoms. Her past medical history was unremarkable, with no family history of congenital anomalies.

On physical examination, she was mildly pale but afebrile and anicteric. The abdomen was distended, with a firm, lobulated, and slightly cystic abdominopelvic mass extending to the epigastrium. Bowel sounds were present. Cardiovascular examination revealed the apex beat on the right hemithorax, suggesting dextrocardia.

Investigations

Abdominopelvic ultrasound demonstrated a huge, thick-walled, predominantly cystic mass arising from the pelvis extending to the left hypochondrial region with septations and mild ascites. Multiple interspersed echogenic structures were seen within it. The cystic regions show low to medium level internal echotexture, the thick-walled internal septations show significant flow on Doppler interrogation. The uterus is normal in size, measuring 5.5×2.7cm (L×AP) with a regular outline and homogenous myometrial echotexture. The endometrial plate is preserved, measuring 6.4 mm. The liver is normal in size, measuring 12.9 cm in span with a regular outline and homogenous background parenchymal echotexture. Normal intrahepatic portobiliary and hepatic venous markings. No nodules or cavities seen. The common hepatic, cystic, common bile duct and main trunk of the portal vein appear preserved at the porta hepatis. The gall bladder is distended with clear bile and has a normal wall thickness, the pancreas appears normal in size, outline and echotexture. The pancreatic duct was not dilated; the spleen is normal in size (11.2 cm). Its outline and echotexture are also preserved. Both kidneys appear normal in shape, size and echotexture. The right kidney measures 9.7×4.7 cm (L×AP) while left kidney measures 11×3.3 cm (L×AP). Normal corticomedullary differentiation. The pelvicalyceal systems appear preserved. Normal sinus echoes MRI confirmed a huge, multiseptated heterogeneous mass with solid and cystic components extending from the pelvis to the upper abdomen, exerting pressure on bowel loops. Additionally, the liver and gallbladder were visualized on the left, the spleen on the right, and the heart on the right side of the chest, confirming situs inversus totalis. Routine blood tests revealed mild anemia (Hb 9.4 g/dL) and thrombocytopenia (platelet count $122\times10^3/\mu L$), with normal renal and liver function. Endoscopy and colonoscopy showed no abnormalities. tumour markers: beta hCG: 5.60 mIU/ml (<10), beta-hCG, AFP: 1.60 ng/ml (< or equal to 8.5) alpha fetoprotein, CEA: 18.2 (<or equal to 5.0) carcinoembryogenic antigen, CA 125: 89.7U/ml (<35) cancer antigen-125.

The magnetic resonance imaging

T2 coronal view of the lower abdomen and pelvis showing a huge left sided cystic mass with hypointense solid components arising from the pelvis (Figure 1A). Coronal view T1 + C MRI showing the cardiac apex on the right, left-sided liver and a huge cystic mass arising from the pelvis (Figure 1B). Sagittal T2-weighted MRI showing a predominantly cystic mass with heterogeneous hypointense solid component. Mild ascites is noted. Normal uterus and urinary bladder seen (Figure 1C). Axial T2 MRI showing left-sided Liver and gallbladder and an abdominal mass in Figure 1D.

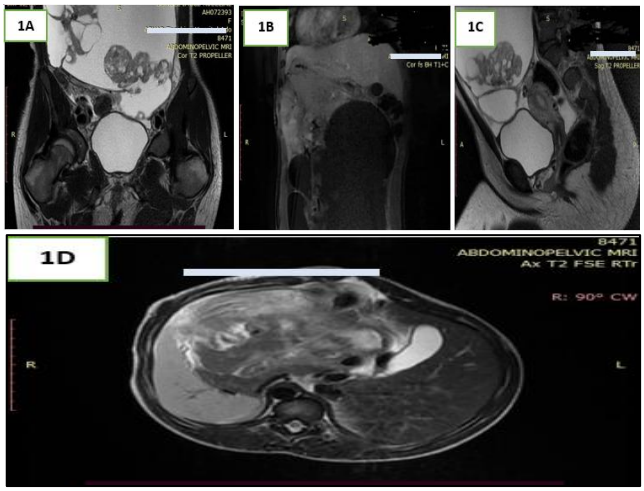

Figure 1 (A-D): (A) Coronal T2 MRI of the pelvis and lower abdomen showing the ovarian mass; (B) coronal fat suppressed T1 + C of the thoracic and abdominal cavity showing stus inversus totalis; (C) sagittal T2 MRI of the pelvis showing the ovarian mass and (D) axial T2 MRI of the abdomen showing the liver and gall bladder on the left.

Surgery

Following preoperative optimization, exploratory laparotomy was performed under general anesthesia. The abdomen was entered via a midline supraumbilical incision. Intraoperative findings included moderate clear ascites, which was aspirated for cytology. A huge lobulated cystic mass arising from the left ovary, measuring approximately 30×28 cm, was identified and carefully excised (Figure 2A). The ovary on the right side of the abdomen appeared grossly normal (Figure 2B).

Remarkably, the liver and gallbladder were located on the left side of the abdomen as against the right side (Figure 3A) also showing the liver lobe with the gall bladder on the left side (Figure 3B). The appendix was found in the left iliac fossa (Figure 3C), confirming SIT. No metastatic deposits or nodules were observed. Estimated blood loss was 150 mL, and the procedure was well tolerated. Figure 3D shows the abdominal postoperative condition.

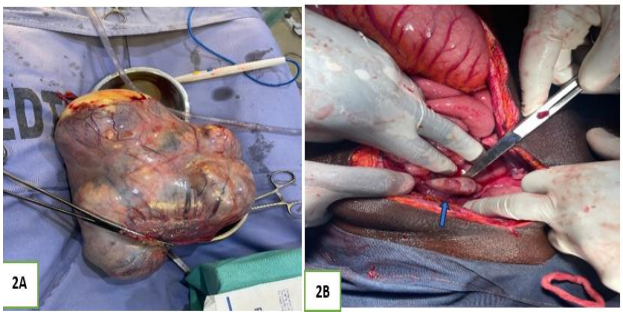

Figure 2: The intraoperative findings. (A) The huge ovarian mass and (B) the normal contralateral ovary.

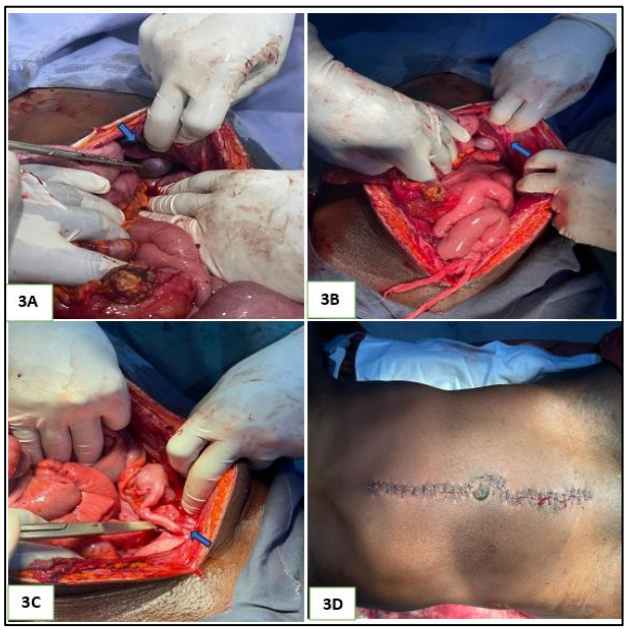

Figure 3: (A and B): The liver and the gall bladder on the left; (C) the appendix on the left; (D) abdominal post operative condition.

Photomicrographic view of the tumor post-operation

Macroscopic examination

Received multi-lobulated, huge cystic ovarian tissue weighing 4 kg, measured 30 cm in its widest diameter. Cut surface reveal presence of straw coloured, greasy yellow substance within the multi-lobulated cavity with some calcified areas admixed with tuft of hair.

Microscopic examination

Sections shows cystic tissue lined by stratified squamous epithelium and its adnexal structures such as sebaceous glands, sweat glands, and hair follicles. There are islands of mature cartilage, neural tissue, and intestinal tissue seen within the stroma (Figure 4).

Diagnosis was ovarian mass: mature cystic teratoma.

Peritoneal fluid cytology-acellular smear.

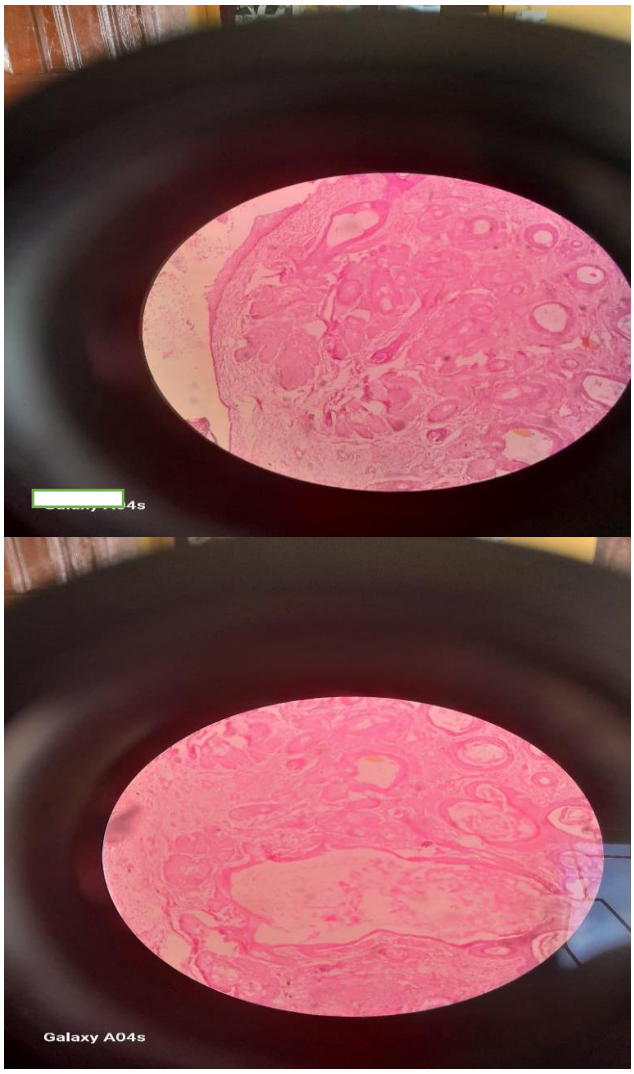

Figure 4: Histological photomicrographic view of the tumor post-operation.

Postoperative course

The patient was managed with intravenous antibiotics, analgesics, and fluid support. She resumed oral intake on the second postoperative day and ambulated early. The urethral catheter was removed on postoperative day one. Recovery was uneventful, and she was discharged home on the seventh day post-surgery in stable condition. At her one-month follow-up, she remained asymptomatic, with no recurrence.

DISCUSSION

Teratomas are among the most fascinating tumors in human pathology, given their origin from pluripotent germ cells and their capacity to differentiate into a wide range of tissue types.[14,15] Ovarian teratomas account for approximately 20-30% of all ovarian neoplasms and represent the most common germ cell tumor in children and adolescents.[16,17] They are typically benign, with mature cystic teratomas (dermoid cysts) constituting over 95% of cases.[18] Although common in gynecological practice, their presentation in childhood poses unique diagnostic and management challenges, particularly in resource-limited settings. The present case further highlights the additional complexity posed by the coexistence of SIT, an exceedingly rare congenital anomaly.

Mature cystic teratomas are predominantly seen in women of reproductive age, but approximately 10-15% occur in children and adolescents.[19] The median age at diagnosis in pediatric cases ranges from 10 to 14 years, consistent with our patient who was 12 years old. Clinically, patients may present with abdominal swelling, pain, menstrual irregularities, or incidental findings on imaging. Complications include torsion, rupture, infection, and, rarely, malignant transformation.[20,21] In our case, the patient presented with progressive abdominal swelling, pain, anorexia, and weight loss features that raised suspicion of malignancy, especially in the setting of anemia and cachexia (however, requested tumour markers- AFP, LDH, HcG, CA-125 and also CEA were essentially within normal for age). Large ovarian teratomas, such as the 30×28 cm mass excised in our patient, are unusual in children but have been reported in literature.[22] The size may reflect delayed presentation, limited access to healthcare, or misdiagnosis. The slow-growing nature of mature teratomas often allows them to reach enormous dimensions before detection. This contrasts with immature teratomas, which typically present earlier due to rapid growth and aggressive behavior.[23] SIT is a rare congenital anomaly characterized by complete mirror-image transposition of thoracic and abdominal viscera. The estimated incidence is 1 in 8,000 to 1 in 25,000 live births.[24] The condition results from disturbances in left-right axis development during embryogenesis, linked to defects in nodal cilia motility and signaling pathways such as NODAL and LEFTY.[25] Although usually asymptomatic, SIT is of clinical significance due to diagnostic pitfalls and surgical difficulties. For instance, appendicitis may present with left iliac fossa pain, and cholelithiasis may mimic left upper quadrant pathology.[26]

Our case demonstrated classical features of SIT: dextrocardia, left-sided liver and gallbladder, and right-sided spleen. Awareness of SIT was crucial intraoperatively, as misidentification of organ orientation could lead to surgical errors. Moreover, SIT is sometimes associated with congenital heart disease and Kartagener's syndrome; however, our patient had no such anomalies.

The concurrence of mature ovarian teratomas and SIT is extraordinarily rare. A limited number of cases have been reported globally.[27,28] The literature suggests no direct embryologic or genetic link between germ cell tumorigenesis and visceral laterality defects, and the association is likely coincidental. Nonetheless, the rarity of the co-occurrence warrants reporting to improve awareness among clinicians and enrich global data. To the best of our knowledge, this case represents the first published report from Nigeria and possibly sub-Saharan Africa.

Radiological consideration

Imaging played a pivotal role in the diagnosis of both the teratoma and SIT. Ultrasound remains the first-line modality for evaluating pelvic masses in children, capable of identifying cystic and solid components, septations, and calcifications typical of teratomas.[29] In this case, ultrasonography revealed a large complex cystic mass, but could not clearly delineate organ orientation. MRI provided superior anatomical detail, confirming the ovarian origin of the mass, detecting the heterogeneous nature of its contents, and incidentally revealing SIT. MRI is particularly valuable for surgical planning in SIT because it clarifies the reversed relationships of abdominal structures.[30]

The role of imaging extends beyond diagnosis to differentiation from other tumors such as mucinous cystadenocarcinoma or immature teratomas. Features such as fat-fluid levels, calcified teeth, and hair are highly suggestive of dermoid cysts.[31] In our case, MRI findings of cystic and solid elements were strongly suggestive of teratoma, but definitive diagnosis required histopathology.

Surgical challenges situs inversus

Surgery in SIT requires careful planning due to reversed anatomy. Surgeons must adapt to unfamiliar orientation, which may prolong operative time and increase risk of inadvertent injury. In this patient, intraoperative findings of left-sided liver, right-sided spleen, and left iliac fossa appendix confirmed SIT. Recognition of this anomaly allowed appropriate adaptation of surgical technique. The huge ovarian mass was excised without complications, and careful inspection revealed no evidence of metastasis.

The mainstay of treatment for mature cystic teratomas is complete excision, typically via oophorectomy or cystectomy depending on size, location, and ovarian preservation considerations.[32] In young patients, fertility-sparing surgery is ideal, but massive tumors often necessitate salpingo-oophorectomy. In this case, left oophorectomy was performed.

Histopathology and prognosis

Histology confirmed the diagnosis of mature cystic teratoma, showing squamous epithelium, sebaceous material, and cartilage without immature or malignant elements. Mature teratomas are benign, with an excellent prognosis after complete excision. Recurrence is rare, typically due to incomplete removal.[33] Malignant transformation occurs in 1-2% of cases, usually into squamous cell carcinoma, and is more common in older women.[34] Our patient remained well at three-month follow-up, with no evidence of recurrence. Long-term follow-up is important to detect recurrence or malignant change, though prognosis in pediatric benign teratomas is excellent.

Lessons from low-resource settings

This case also reflects the challenges of managing rare conditions in low- and middle-income countries (LMICs) like Nigeria. Delayed presentation is common due to financial constraints, limited access to diagnostic imaging, and reliance on traditional healers. Our patient presented with a very large tumor, likely due to these systemic barriers. Despite these limitations, successful surgical management was achieved, highlighting the capacity of regional teaching hospitals to handle complex cases when adequately resourced.

Additionally, there is a paucity of tumor registries in sub-Saharan Africa, limiting epidemiological data on rare tumors and congenital anomalies. Establishing multi-institutional registries would improve understanding, guide policy, and enhance collaborative research on such unusual cases.

Literature context and comparisons

Reports from Asia and Europe have described similar associations of SIT with ovarian teratomas.[35-37] Most patients were young women presenting with abdominal mass, and all were managed surgically with favorable outcomes. The rarity of the condition precludes large-scale studies, but accumulated case reports help document its existence and alert surgeons to anticipate reversed anatomy in patients with SIT. Our report adds to this limited pool and is, to our knowledge, first from Nigeria.

Limitations and future perspectives

The main limitation of this report is its single-patient nature. It cannot establish causal relationships or generalize outcomes. Moreover, follow-up duration was short, and long-term prognosis remains to be assessed. Nonetheless, the case underscores the need for vigilance in diagnosing rare anomalies and demonstrates successful management in a resource-constrained environment. Future directions include genetic studies to explore potential associations between germ cell tumors and laterality disorders, as well as broader efforts to strengthen surgical oncology capacity in LMICs.

CONCLUSION

Mature cystic teratomas are common benign ovarian tumors in children and adolescents, but their coexistence with situs inversus totalis is extremely rare. This case emphasizes the importance of thorough clinical and radiological evaluation, careful intraoperative assessment of organ orientation, and complete surgical excision. The excellent postoperative outcome demonstrates that even in resource-limited settings, successful management is possible when surgical teams remain vigilant. Reporting such rare associations contributes to global knowledge and highlights need for tumor registries in sub-Saharan Africa.

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