WEST AFRICAN JOURNAL OF MEDICINE

CASE REPORT

Urethral Fibrosis with Pyonephrosis in a Patient with Situs Inversus Totalis: A Case Report.

La Fibrose d'urethral Avec Pyonephrosis dans un Malade Avec Situs Inversus Totalis: Un Rapport de Cas

O. C. Osime*, U. Osime*, T. Njoku*, A. A. Adeyekun †, G. Obonna*, E. Okoro*, D. Obaseki‡, A. Igbe‡

ABSTRACT

BACKGROUND: **Cases of a case of situs inversus totalis are very rare and therefore when encountered, they are likely to be missed or poorly managed.**

OBJECTIVE: **To present a case of situs inversus totalis with a view to creating more awareness about this rare clinical entity.**

PATIENT AND METHOD: **The patient, a 22 year old boy was referred from a private clinic after a failed and complicated attempted appendicectomy carried out by a general medical practitioner. The patient was admitted, resuscitated and had investigations done. Some of the investigations done were abdominal ultrasound scan, computerized axial tomography scan, a chest x ray, complete blood count, urinalysis and electrolyte and urea.**

RESULTS: **Investigations showed features of situs inversus totalis. There was also bilateral hydronephrosis, worse on the right side. The patient also had features of renal failure. He died after three days on admission. Autopsy finding confirmed bilateral pyonephrosis which was worse on the right side. There was associated urethral fibrosis. Cause of death was urethral fibrosis resulting in obstructive uropathy, pyonephrosis and renal failure.**

CONCLUSION: **Situs inverses totalis is rare but can psented in diverse ways and thus the diagnosis may be missed and the patient wrongly managed. A high index of clinical suspicion is required if we the diagnosis is to be made early and wrong treatments avoided. WAJM 2007; 26(3): 246 – 249.**

Keywords: **Situs inverses totalis, urethral fibrosis, pyonephrosis, diagnosis, missed.**

RESUMÉ

Contexte: *Les cas d'un cas de situs inversus totalis sont très rares et donc quand rencontré, ils sont probables être manqué ou être géré mal.*

Objectif: *Pour présenter un cas de situs inversus totalis avec une vue à créant plus de conscience de cette entité clinique rare.*

Methodes: *Le malade, un 22- l'année vieux garçon a été référé d'une clinique privée après un d'appendicectomy raté et compliqué tenté a exécuté par un practicien médical général. Le malade a été admis, a été réanimé et a été eu des investigations faites. Certaines des investigations faites étaientultrasons abdominales que les scrutent, informatisé le balayage de tomographie axial, un rayon-x de poitrine, compléter le compte de sang, l'analyse d'urine et l'électrolyte et l'urée.*

Résultat. *des Investigations les caractéristiques montrées de situs inversus totalis. Il y avait hydronephrosis aussi bilatéral, pire sur le bon côté. Le malade a eu aussi des caractéristiques d'échec rénal. Il est mort après trois jours sur l'admission. L'autopsie trouvant pyonephrosis bilatéral confirmé qui était pire sur le bon côté. A été là-bas associé la fibrose d'urethral. La cause de mort était la fibrose d'urethral a pour résultat uropathy peu coopératif, pyonephrosis et l'échec rénal.*

Conclusion: *Totalis inverse est rare mais psented de boîte dans les façons diverses et ainsi le diagnostic pourrait être manqué et le malade a géré incorrectement. Un haut index de soupçon clinique est exigé si nous le diagnostic est être fait de traitements premiers et mauvais évités.**WAJM 2007; 26(3): 246 – 249.***

Mots clés: Situs totalis inverse, la fibrose d'urethral, pyonephrosis, misdiagnosis.

* Departments of Surgery, †Radiology, ‡Pathology University of Benin Teaching Hospital, Benin City, Edo State, Nigeria.

Correspondence: Dr. O. C. Osime, Department of Surgery,University of Benin Teaching Hospital, PMB 1111, Benin City, Edo State, Nigeria. Email: clementosime@yahoo.com

INTRODUCTION

Situs inversus (also called situs tranversus) is a rare congenital condition in which the major visceral organs are reversed from left to right, a mirror image of the normal condition (which is known as situs solutes). In other rare cases, in a condition known as situs ambigones or heterotaxy, situs cannot be determined. The term situs inversus is a short form of the Latin phrase "situs inversus viscerum" meaning inverted position of the internal organs. Dextrocardia (the heart being located to the right side of the thorax) was first recognized by Marco Severino in 1643. However situs inversus was first described more than a century later by Mathew Baillie.[1] Situs inversus is an autosomal recessive genetic condition. The prevalence of situs inversus varies among different populations, but is less than 1 in 10,000 people. However, it seems the figure may be more than this because many cases are often confused with a mislabeled chest x-ray, when in fact the patient has a dextrocardia.[2]

Some anomalies are associated with situs inversus viscerum.[3-5] About 25% of individuals with situs inversus have an underlying condition known as primary ciliary dyskinesia, also known as Kartagener's syndrome. It is characterized by the triad of situs inversus, chronic sinusitis and bronchiectasis. Patients with this condition are also prone to having various fibrotic changes. As a result of the change in location of the internal organs, there is often a confusing clinical presentation whenever they have symptoms and signs of certain clinical conditions that may lead to delay in diagnosis.[6,7]

Some patients with situs inversus viscerum may develop some complications and which are hardly detected except at post mortem.[3,4] Others may present with symptoms in early childhood, while some present in adulthood.[2,5] Considering the rarity of this condition, the Surgeon needs to be astute and detailed in clinical examination in order to avoid making a wrong diagnosis with the attendant morbidity and mortality.

In view of the ease with which an error in clinical diagnosis can occur in cases with situs inversus viscerum, we present a case of situs inverses totalis who presented with pains over the right iliac fossa and right lumber regions. The aim of this presentation is to present a case of situs inversus totalis we managed at the University of Benin Teaching Hospital with a view to creating more awareness about this rare clinical entity.

CASE REPORT

The patient was a 22-year old man. He was the first of six siblings (2 males, 4 females) in a monogamous setting. He presented to a private general medical practitioner with complaints of abdominal pain and abdominal distension of about three weeks duration. The patient's complaints started with pain over the right iliac fossa and lumber regions. There was associated fever, and nausea and vomiting. An assessment of acute appendicitis was made. Attempts at locating the vermiform appendix were not.

The patient was thereafter managed with antibiotics on the ward. The condition of the patient started deteriorating as evidenced by the persistent fever, abdominal distension and peripheral oedema. He was subsequently referred to the University of Benin Teaching Hospital (UBTH). On examination at the UBTH the patient was acutely ill-looking febrile, dehydrated and jaundiced. He had bipedal pitting oedema up to the knee and also had abdominal distension. There was an infected Lanz incision wound discharging purulent exudate. There was generalised abdominal tenderness with guarding and boarding rigidity. The percussion notes were hyper-resonant and bowel sounds were absent. There was bogginess of anterior rectal wall with associated tenderness. The rectum was empty. The respiratory rate on presentation was 36/min. Clinical examination of the chest revealed bilateral basal crepitations. The pulse rate was 112 beats/min and regular with a supine blood pressure of 90/40mmHg. The apex beat was not palpable. An assessment of peritonitis was made. An abdominal ultrasound scan and CT scan (figs. 1 & 2) showed transposition of the intra-abdominal organs. There was free fluid within the peritoneal cavity. The ultrasound also showed bilateral hydronephrosis, worse on the right side. Plain abdominal X-Ray showed dilated loops of bowel with multiple air-fluid levels. The chest X-ray showed dextrocardia. There was also bilateral pleural effusion, worse on the right side. Serum biochemistry showed a urea level of 293mg/dl, potassium of 5.8 mmol/l, chloride of 98mmol/l, sodium of 135 mmol/l, bicarbonate of 10mmol/l and creatinine of 2mg/dl. The total white cell count was 19,000 cells/mm^3 with a neutrophilia of 83%. The packed cell volume was 21%. The considerations at this point were peritonitis with paralytic ileus and pyoperitoneum,, bilateral hydronephrosis with imminent renal failure and situs inversus totalis

He was commenced on broad spectrum antibiotics and intravenous fluid administration. A nasogastric tube was passed. A urethral catheter was also passed. In the first 24 hours on admission,

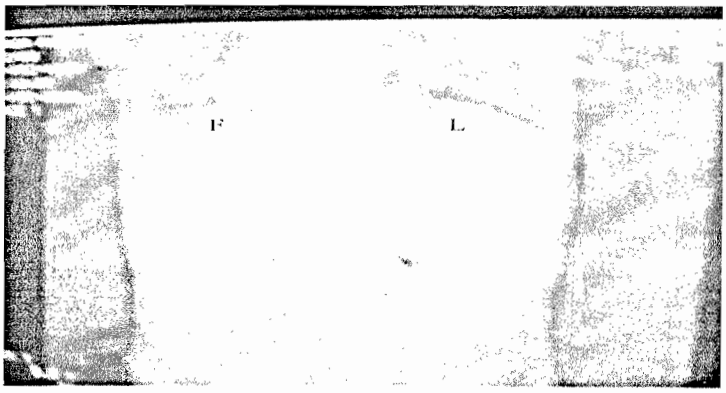

Fig 1: **Scanogram shows cardiac apex white arrow (A) on the right; gastric fundus (F), sited to the right of the midline. The liver (L) is to the left. (Note marker for right [R] on scanogram.**

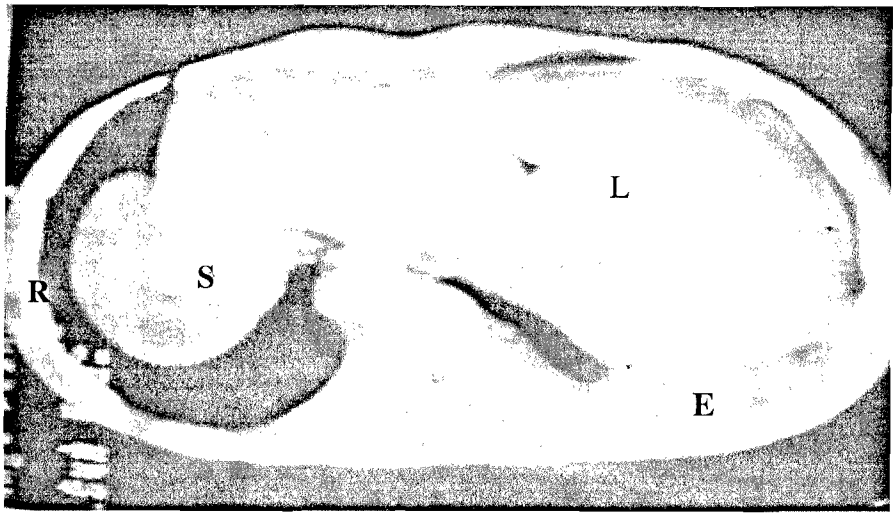

Fig. 2: **Axial CT of the upper abdomen following oral contrast ingestion confirming the left sided location of the liver (L) and a right sided stomach (S) containing contrast. Note marker (R) on scanogram and label (E) for pleural effusion.**

the urine output was 200mls, despite having received 4 litres of intravenous fluid. The patient's condition continued to deteriorate and on the third day on admission the patient went into shock and efforts at resuscitation failed. .

Essential findings at autopsy were severe anemia, bilateral pitting pedal oedema, septic right iliac fossa wound, transposition of abdominal organs, ascites, peritonitis, a left sided vermiform appendix with no signs of inflammation, dilated posterior urethra secondary to urethral fibrosis, dilated ureters, bilateral pyonephrosis-gross pyonephrosis on the right side, dextrocardia and bilateral pleural effusion. The cause of death at autopsy was concluded to be urethral obstruction leading to pyonephrosis, septicaemia and renal failure.

DISCUSSION

Situs inversus affects all major structures within the thorax and abdomen. In the absence of congenital heart defects, individuals with situs inversus can lead normal healthy lives, without any complications relating to their medical condition. Douard et al[8] reported a case of situs inversus totalis in an 87 year old lady at autopsy. Most people with the condition are unaware of it. The reversal of the organs may then lead to some confusion as many signs and symptoms will be on the 'wrong' side. There have been several reports of cases of missed diagnosis in situations were the vermiform appendix was located on the left side.[6,7,9] However, the case presented here is that of right iliac fossa and right lumber region tenderness which was thought to be due to appendicitis. But at operation, the vermiform appendix was not found on the right side, instead autopsy findings reveals a left sided vermiform appendix with no signs of inflammation. Our patient had urethral fibrosis and pyonephrosis.

Some studies have reported the association of situs inversus with some renal complication. Podkor[4] reported a case of acute pyelonephritis and calculous cholecystitis in a patient with complete situs inversus. Pinar et al[3] reported a case of renal dysplasia, situs inversus totalis and multisystem fibrosis. Similarly, reported a case of renal dysplasia and situs inversus totalis. Hiraoka et al[10] described a rather complex case of bilateral renal dysplasia, pancreatic fibrosis, intrahepatic biliary dysgenesis and situs inversus totalis in a young boy. Some patients with situs inversus may also have congenital renal anomaly.[11] Domasaki et al[12] reported a case of nephritic syndrome in a patient with situs inversus totalis.

There have been other reports of fibrosis or atresia of some organs in patients with situs inversus viscerum. Aziz et al[13] reported a case of biliary atresia with situs inversus. Njoh[14] et al reported pancreatic fibrosis and calcification in a Liberian female that had situs inversus viscerum. Cases of duodenal obstruction secondary to atresia in patients with situs inversus viscerum were reported by Nawaz et al.[15] There are some other situs inversus. Madharan et al[16] reported a case of retinitis pigmentosa in a sickle cell patient that also had situs inversus viscerum. Khalil et al[17] reported a case of a 54 year old man with situs inversus viscerum who presented with acute myocardial infarction. Other modes of presentation have similarly been reported by other studies.[18]

The condition has been reported to run in some families. Bala et al[11] reported a case of three sibs diagnosed prenatally with situs inversus totalis, renal and pancreatic dysplasia. In the case presented in our report, we were not able to screen the family to detect if there were other members that had situs inversus totalis. After the death of our patient, the father of the patient was most unwilling to come with the members of his family for clinical examination.

In conclusion, situs inversus totalis, a rare clinical entity is often associated with a number of conditions and complications and often present a diagnostic puzzle that will task the diagnostic acumen of the physician. It is recommended that patients who are aware that they have situs inversus should inform their physician early enough in the course of clinical examination and investigations to reduce the chances of wrong diagnosis. The case presented here further emphasizes the obvious need for detailed clinical examination of patients and to investigate such patients fully wherever there is a doubt about the clinical findings. It is relevant for doctors to ensure they carry out complete physical examination while examining patients. If a thorough physical examination was carried out in this patient we have presented, he would have been found to have his apex beat

on the right hand side and this would have raised a suspicion of a left sided vermiform appendix. If this was the case, a diagnosis of appendicitis would not have been a consideration. It is very likely that if the correct diagnosis was made in our patient, the obstructive uropathy could have been detected and taken care of much earlier before the onset of more severe complications that culminated in renal failure. While not disputing the fact that general medical practitioners can perform appendicectomy in our setting in Nigeria, it is submitted that whenever there is an 'unusual' finding during such operations, the Surgeon should be involved in the management of that patient. This will go a long way in minimizing the cases of litigations vis-à-vis the tort of negligence.

REFERENCES

1. Cleveland M. Situs inversus viscerum: anatomic study. *Arch Surg* 1926; **13:** 343 –348.
2. Hynes KM, Gau GT, Titus JC. Coronary heart diseases in situs inversus totalis. *Am J Cardio* 1973; **31:** 666 – 669.
3. Podkur PE. Acute pyelonephritis and calculous cholecystitis in a patient with complete situs inversus viscerum. *Khirurgia* 1991; **11:** 126 –127.
4. Pinar H, Rogers BB. Renal dysplasia, situs inversus totalis and multisystem fibrosis: a new syndrome. *Pediatr pathol* 1992; **12:** 215 – 221
5. Chaturvedi P, Thomas K. Congenital renal anomaly in a patient with situs inversus. *Postgrad Med J* 2003; **79:** 359 – 361.
6. Carmichael KA, Gayle WE Jr. Situs inversus and appendicitis. *South Med J* 1979; **72:** 1147 – 1150.
7. Kamiyam T, Fujiyoshi F, Hamade H, Nakojo M, Harado O, Horaguchi Y. Left sided acute appendicitis with intestinal malrotation. *Radiat Med* 2005; **23:** 125 –127.
8. Donard R, Feldman A, Bargy F, Loric S, Delmas V. Anomalies of lateralization in man: a case of total situs inversus. *Surg Radiol Anat* 2000; **22:** 293 – 297.
9. Nelson MJ, Pesola GR. Left lower quadrant pain of unusual cause. *J Emerg Med* 2001; **20:** 241 –245.
10. Hiraoka K, Haratake J, Horie A, Miyagawa T. Bilateral renal dysplasia, pancreatic fibrosis, intrahepatic biliary dysgenesis and situs inversus totalis in a boy. *Hum Pathol* 1998; **19:** 871 – 873.
11. Bala S, Bostanglu S, Altinok G, Ozaltin F. New syndrome?: Three sibs diagnosed prenatally with situs inversus totalis, renal and pancreatic dysplasia and cysts. *Am J Med Genet*; 2000; **90:** 185 – 187.
12. Domanski M, Domanski L, Ciechanowski K. Nephrotic syndrome and situs inversus viscerum: correlation or coincidence. *J Nephrol* 2005; **18:** 623 – 625.
13. Aziz S, Soomru GB, Leuk NH, Hussaid SM, Mirza R, Naqui SA, Rizvi AH. Biliary atresia with situs inversus: an experience shared. *J Pak Med Assoc* 2005; **55:** 350 –35 2.
14. Njoh J, Findley G, Dharmavrotha S. Pancreatic fibrosis, calcification and situs inversus in a Liberian female: a case report. *Trop Georgr Med* 1986; **38:** 188 –189.
15. Nawaz A, Matta H, Hamchon M, Jachobez A, Trad O, Al Salem AH. Situs inversus abdominus in association with congenital duodenal obstruction: a report of two cases and review of the literature. *Pediatr Surg Int* 2005; **21:** 589 – 592.
16. Madheran C, Bhende P, Gopal L, Vasanthi SB, Kumaramanickavel G. Retinitis pigmentosa patients with sickle cell disease and dextrocardia and situs inversus syndrome. *Indian J Ophthamol* 2001; **49:** 193 – 195.
17. Khahl S, Kamal A. Acute myocardial infarction in a patient with dextrocardia and situs inversus. *J Ecectrocardiol* 2005; **38:** 412 – 413.
18. Danbauchi SS, Ahassan MA. Case report: dextrocardia with situs inversus; two cases presenting differently. *Niger Postgrad Med J* 2002; **9:** 248 – 252.