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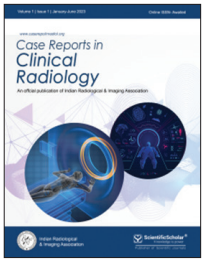

*Case Report*

# Situs ambiguous-polysplenia syndrome: Torsion splenunculus – A rare cause of acute abdomen pain

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## ABSTRACT

Situs ambiguous, also known as heterotaxy syndrome, is a rare congenital condition in which there is an abnormal positioning of internal organs of the chest and abdomen. Polysplenia (left isomerism) is a common association with it and hence called as "Situs Ambiguous-Polysplenia" syndrome. Torsion splenunculi is a rare complication associated with it. Herein, we report a case of splenunculi torsion in an adult with situs ambiguous associated with polysplenia.

**Keywords:** Heterotaxy syndrome, Situs ambiguous, Torsion splenunculi

## INTRODUCTION

"Heterotaxy syndrome" also known as "situs ambiguous" is derived from two Greek words; heteros – meaning other and taxis implies arrangement. This rare congenital condition can either present with the left isomerism (polysplenia syndrome) or right isomerism (asplenia syndrome). The prognosis of former better is due to less severe cardiac abnormalities. Polysplenia is generally asymptomatic, but can present with various acute complications like torsion in rare instances.

## CASE REPORT

A 3-year-old boy presented to pediatric outpatient department with a complaint of acute onset of abdomen pain localized to the right hypochondrium associated with fever. There was no complaint of cough, vomiting, loose stools, or constipation. Peripheral blood smear of the patient showed thrombocytosis (platelet count 719,000 cells/cc), lymphocytosis (54%), and microcytic hypochromic anemia with mild anisocytosis (Hemoglobin 8.3 g%).

The patient was referred to ultrasound (USG). USG showed a well-defined hypoechoic lesion with internal low level septations [Figure 1] with possibilities of necrotic node or omental torsion. Contrast computed tomography (CT) of the abdomen and chest was suggested for further evaluation. Contrast-enhanced CT revealed multiple splenunculi in the right hypochondrium with absent parent spleen. One of the splenunculi was relatively hypodense with only marginal and peripheral enhancement, measured (4 × 3.8 × 3.7 cm). Situs ambiguous with the right-sided liver, dorsal pancreatic agenesis, aorta on the right side, inferior vena cava (IVC) on the left side, altered superior mesenteric vein superior mesenteric artery – axis, and left side ileocecal junction

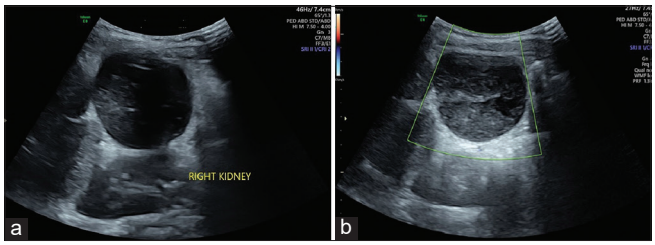


**Figure 1:** (a) Ultrasound (USG) B mode shows a well-defined hypoechoic lesion with internal low-level septations with possibilities of necrotic node or omental torsion (b) USG color Doppler show no uptake of color flow.

were also noted [Figure 2]. However, the right side lung was trilobed and the left lung was bilobed with corresponding number of bronchi [Figure 2]. The diagnosis of situs ambiguous-polysplenia syndrome with torsion splenunculi was made.

Laparoscopic excision of the infarcted splenunculus done with prophylactic pneumococcal vaccination. Pre-operative cardiac evaluation, however, did not show any congenital heart disease in this case.

## DISCUSSION

Heterotaxy syndrome is characterized by an abnormality in the distribution of the thoracic and intra-abdominal contents. It differs from situs inversus as the former does not entirely correspond to the mirror image seen in latter.[1] It occurs from the early embryological developmental disturbance with most cases being sporadic in distribution.[2] The incidence of heterotaxy syndrome is rare, which is around 1/8000–25,000 live births.[1]

Radiological features of heterotaxy syndrome are extremely complex. There may be duplication of the left- or right-sided intra-thoracic contents with associated changes below the diaphragm. Classically, there is malposition of liver, stomach, and spleen. In addition, the vascular anatomy can be altered. Congenital heart diseases are often associated with this condition.[1,3] Polysplenia can be seen in 2.5/100,000 of live births which is approximately seen in 55% of cases with the left isomerism.[1] Interrupted IVC is the second most consistent associated finding next to polysplenia, although extremely rare (1%).[3]

Embryologically, spleen arises from mesenchymal buds in the dorsal mesogastrium during the 5$^{th}$ week of gestation. Incomplete fusion results in splenunculi and when associated with situs ambiguous, they are seen on the right side of the abdomen. They however are still vascularized by the branches of splenic artery.[4] The most common sites of accessory spleen are splenic hilum (75%), followed by pancreatic tail (20%).[4] Usually asymptomatic, they can rarely get complicated with torsion, infarction, and ruptured with bleeding.[4]

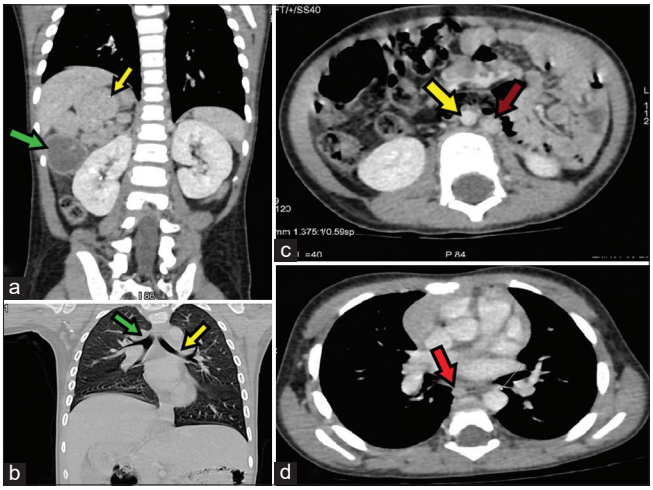


**Figure 2:** (a) Contrast-enhanced CT (CECT) of abdomen and pelvis coronal section shows multiple splenunculi (yellow arrow) in the right hypochondrium with absent parent spleen. One of the splenunculi was relatively hypodense with only marginal and peripheral enhancement, measured (4 × 3.8 × 3.7 cm) (green arrow). (b) CT lung window shows, trilobed right side lung (green arrow) and bilobed left lung (yellow arrow) with corresponding number of bronchi. (c) CECT axial section of abdomen shows aorta on the right side (yellow arrow) and inferior vena cava (IVC) (brown arrow) on the left side. (d) Contrast-enhanced CT axial section of the chest shows azygos continuation of IVC (red arrow).

The first case of splenunculi torsion was reported by Alexander and Romanes[5] and since then only very few cases are reported in the literature.[4] Imaging plays a very important role in accurate diagnosis of this complication.[4,6] Twisting of the vascular pedicle when present can be diagnostic.[7] Venous congestion can result in enlargement of the torsed spleen, with reduced enhancement of the surrounding fat. There is usually an intensely enhancing capsule supplied by the patent capsular arteries and characterized by the peripheral enhancement of the torsed spleen.[7] The differential diagnosis includes wandering spleen, necrotic nodes, omental infarction, and chronic encapsulated fat necrosis. Awareness of this syndromic association is essential to suspect this rare cause of acute abdomen pain.

Necrotic nodes, commonly seen in tuberculosis (TB), are often conglomerate due to periadenitis which is classical of TB. They are often seen in multiple sites. Encapsulated fat necrosis is characterized by fat attenuation in center (Hounsfield units-10–20) with peripheral rim of calcification, unlike the central necrosis in splenunculus torsion.[8] Omental infarction is evidenced by inflammatory fat stranding and fat inflammation characterized by altered normal homogeneous fat signals of omentum.[9] Wandering spleen is a condition in which spleen is normally fused and formed but located at lower abdomen which can rarely undergo torsion due to long course of artery supplying it.

**Differential diagnosis**

| | |
|---|---|
| 1. Situs inversus totalis | • There is a total transposition of abdominal and thoracic viscera (mirror image of internal organs normal positioning)<br>• Locations of the heart apex, aortic arch, stomach bubble and liver are important to differentiate situs in-versus from situs ambiguous. |
| 2. Omental infarct | • Focal area of inflammatory fat stranding<br>• Swirling of omental vessels adjacent to site of torsion<br>• Adjacent bowel may not show significant wall thickening differentiating this from diverticulitis. |
| 3. Wandering spleen | • Absence of the spleen in the left upper quadrant<br>• Ovoid or comma-shaped abdominal mass, most commonly in the left lower quadrant<br>• Whirl sign: a whirled appearance of splenic vessels in case of torsion. |
| 4. Encapsulated fat necrosis | • Encapsulated fat density mass lesion with peripheral calcification. No adjacent signs of inflammation. |
| 5. Necrotic nodes | • Most common cause is tuberculosis, they are usually multiple and appear conglomerated<br>• Most common site is para-aortic and mesenteric regions<br>• Metastatic nodes and treated lymphoma are other causes of necrotic nodes but are often multiple. |

It can also be associated with heterotaxy syndrome but splenunculi are not seen.[10]

## CONCLUSION

It is important to be aware of types of isomerism and complications associated with it as they are very rarely seen in clinical practice. Differentiation from situs inversus is important and imaging the chest can be helpful in the diagnosis. Cardiac anomalies are very common with heterotaxy syndromes, and hence, screening echocardiography is mandatory before any surgical interventions.

## TEACHING POINTS

1. Polysplenia is characteristic of situs ambiguous (left isomerism).
2. Torsion of splenunculi should be kept as a differential diagnosis in cases of polysplenia presenting with acute abdominal pain.
3. Congenital heart diseases should be ruled out as they are closely associated with heterotaxy syndrome.

**MCQs**

1. Which of the patients have a better prognosis among heterotaxy syndrome?
   a. Left isomerism
   b. Right isomerism
   c. Both
   d. None

Answer Key: a

2. All are associations of heterotaxy syndrome (left isomerism) except
   a) Polysplenia
   b) Asplenia
   c) Intestine malposition
   d) Azygos continuation of IVC

Answer Key: b

3. All are differential diagnosis for torsion of splenunculi except
   a) Omental infarction
   b) Necrotic nodes
   c) Encapsulated fat necrosis
   d) Pseudo-aneurysm

Answer Key: d

**Declaration of patient consent**

Patient's consent not required as patients identity is not disclosed or compromised.

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Nil.

**Conflicts of interest**

There are no conflicts of interest.

## REFERENCES

1. Dash MR, Upasani AV, Chandna SB, Rathod PB, Prajapati KK, Patel DN. Splenic torsion in a child with polysplenia and situs inversus: A very rare presentation. Indian J Surg 2013;75(Suppl 1):236-7.
2. Garófano-Jerez JM, Benedicto-Hernández E, Gila JD, Garrido JD. Imaging evaluation of the heterotaxy syndrome. Gastroenterol Hepatol Open Access 2019;10:75-80.
3. Ali M, Khan N, Shahid J, Shah SZ, Raees A. Heterotaxy polysplenia syndrome presenting with bleeding from the greater curvature of stomach: A case report. Egypt J Radiol Nucl Med 2022;53:135.
4. Ishibashi H, Oshio T, Sogami T, Nii A, Mori H, Shimada M.

Torsion of an accessory spleen with situs inversus in a child. J Med Invest 2012;59:220-3.
5. Alexander RC, Romanes A. Accessory spleen causing acute attacks of abdominal pain. Lancet 1914;184:1089-91.
6. Impellizzeri P, Montalto AS, Borruto FA, Antonuccio P, Scalfari G, Arena F, *et al*. Accessory spleen torsion: Rare cause of acute abdomen in children and review of literature. J Pediatr Surg 2009;44:e15-8.
7. Sadro CT, Lehnert BE. Torsion of an accessory spleen: Case report and review of the literature. Radiol Case Rep 2013;8:802.
8. Kamaya A, Federle MP, Desser TS. Imaging manifestations of abdominal fat necrosis and its mimics. Radiographics 2011;31:2021-34.
9. Corvino A, Campanino MR, De Rosa N. Corvino F, Gisonni P. Left-sided omental infarction without torsion: Report of a case with radiologic-pathologic correlation. Egypt J Radiol Nucl Med 2020;51:117.
10. Lam Y, Yuen KK, Chong LC. Acute torsion of a wandering spleen. Hong Kong Med J 2012;18:160-62.

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