

## 99m-Techneium-Labeled Red Blood Cells' Single-Photon Emission Computed Tomography/Computed Tomography in Situs Ambiguous with Polysplenia

### Abstract

Situs ambiguous or heterotaxy syndrome is defined as the abnormal arrangement of organs and vessels within the body. Herein, we present an 8-year-old girl with growing right-sided suprarenal masses mimicking adrenal tumors. Technetium-99m-radiolabeled red blood cells' planar images were inconclusive for the diagnosis of splenic tissue in the right side. Single-photon emission computed tomography/computed tomography imaging clearly identified right-sided functioning spleens and confirmed the diagnosis of situs ambiguous with polysplenia.

**Keywords:** *Heterotaxy syndrome, polysplenia, selective spleen scintigraphy, single-photon emission computed tomography/computed tomography, situs ambiguous, technetium-99m-labeled red blood cells*

The term "situs" means "position, site, and location."<sup>[1]</sup> Situs solitus is the normal positioning of organs and vessels within the body. Situs inversus represents anatomic arrangement that is the mirror image of situs solitus. The condition which is called situs ambiguous or heterotaxy syndrome is less common, and it refers to visceral malposition and dysmorphism. The two main subgroups of situs ambiguous are situs ambiguous with polysplenia and situs ambiguous with asplenia.<sup>[1-3]</sup> Although the cases are classified into two main groups, it should be remembered that this anomaly has no pathognomonic feature and a single description is not possible.<sup>[4,5]</sup> Multiple spleens of variable size and number, bilateral bilobar lungs, centrally located liver, and dextrocardia are the most common anomalies; also, the stomach may be right sided in up to 75% of children with polysplenia that are typically in the same side with stomach along the greater curvature as in our case.<sup>[2,6]</sup> [Figure 1] A segmental or individualized approach has been recommended with the use of specific anatomic labels for describing patients with heterotaxy syndrome to achieve the best clinical management knowing the

clinical risks caused by malposition.<sup>[1]</sup> SSS is useful in characterizing an incidentally detected mass as functional splenic tissue.<sup>[7-9]</sup> It should be kept in mind that right-sided spleen tissue can mimic adrenal tumors and is more likely to be misdiagnosed as an adrenal tumor because of its growing in time, especially in children.<sup>[10]</sup> Although there are some studies in the literature using Tc-99m sulfur colloid in pediatric patients, damaged red blood cell (dRBC) scintigraphy is less commonly used in children because of its technical difficulties and special radiopharmaceutical preparation. However, dRBC has advantages over sulfur colloid in differential diagnosis due to its high specificity to splenic tissue.<sup>[9,11]</sup> Single-photon emission computed tomography/computed tomography gives valuable information if there are multiple or small lesions, where planar spleen scan images are inconclusive, especially in the presence of overlapping activity [Figure 2].

### Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images

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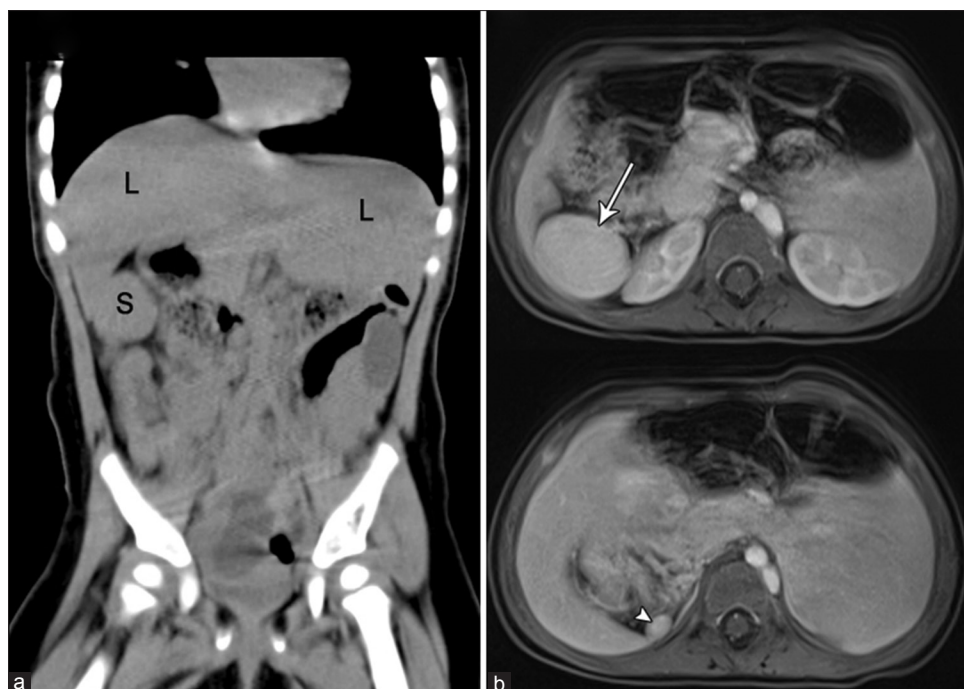
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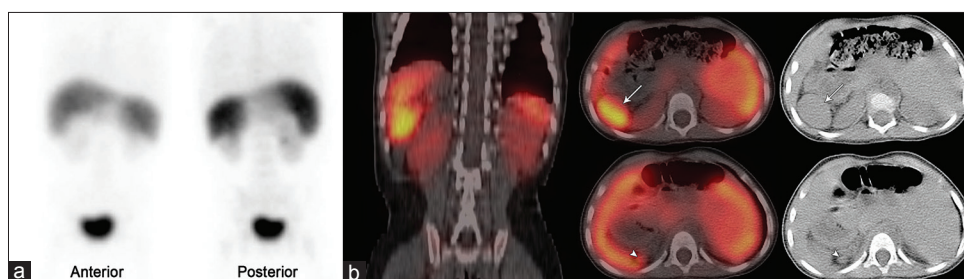


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**Figure 1:** An 8-year-old girl with left inguinal herniation was referred to abdominal ultrasonography. Ultrasonography revealed two homogeneous isoechoic solid masses in the right adrenal region. Further evaluation was needed to identify the adrenal masses due to the increase in size. Coronal magnetic resonance image showed a bridging liver (L) occupying both right and left upper quadrant of the abdomen (a). Axial magnetic resonance images showed 3 cm- (arrow) and 1 cm-sized (arrowhead) solid nodules adjacent to the liver that were suspected as the spleen (s) tissue (b)



**Figure 2:** The patient was referred to selective splenic scintigraphy to confirm the diagnosis. Splenic imaging was performed after the injection of Tc-99m-labeled heat-damaged red blood cells. The spleen was not observed in normal localization, and static planar images were inconclusive for the diagnosis of splenic tissue in the right side (a). Single-photon emission computed tomography/computed tomography clearly showed a prominent damaged red blood cell uptake in the suprarenal masses along the greater curvature of the right-sided stomach and was compatible with functioning splenic tissue (b). In addition, left-sided inferior vena cava was revealed, whereas the apex of the heart was left sided (levocardia)

and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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#### Conflicts of interest

There are no conflicts of interest.

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