

Clinical Quiz

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An unique and unusual lucent shadow seen on a plain x-ray abdomen anteroposterior view in erect position

Clinical Presentation

A plain x-ray anteroposterior erect film of a 28-year-old, well built and nourished, Bangladeshi expatriate man without any significant clinical complaint shows an irregular, large, lucent shadow in the right hypochondrium (Figure 1).

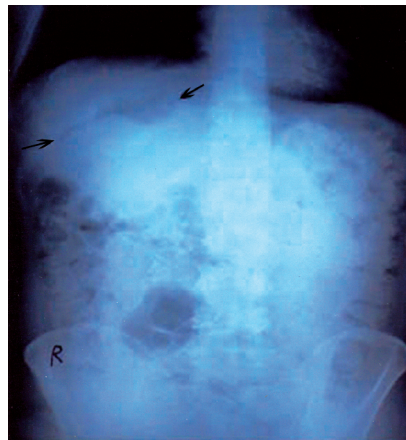


Figure 1 - X-ray abdomen anteroposterior erect. Note the irregular lucent shadow in right hypochondrium

Questions

1. What are the other findings? (Clue: Negative findings)
2. What is the likely Radiodiagnosis?

Clinical Quiz

Answers

1. Absence of stomach air bubble and absence of splenic shadow below the left diaphragm. The left subdiaphragmatic space is mostly occupied by transverse colon and splenic flexure.
2. **Figure 2** shows the answer very clearly. This is a case of “isolated stomach lying on the right side.” The irregular, lucent shadow is that of the air bubble in the so-lying stomach. This is a rare case of “heterotaxy involving the stomach” classified under the “right sided bilaterality sequence.” Hence, we coin a shorter term as “Dextrogastria” for this condition. **Figure 2** also shows the associated mirror images of abdominal esophagus and duodenum.



Figure 1 - X-ray anteroposterior abdomen after Barium ingestion. Note the right sided stomach.

Discussion

‘Situs’ means a state of affairs as regards to side and positioning of the viscera. ‘Situs solitus totalis’¹ implies normal asymmetry and positioning of all the viscera in the thorax and abdomen. ‘Situs inversus totalis’ indicates complete reversal of asymmetry in all organs of the thorax and abdomen. A complete mirror image, transpositioning, of all organs is seen. The incidence of situs inversus totalis is 1/7000, and is usually associated with normal physiology in the organs. In ‘isolated situs solitus,’ the thoracic viscera is in the solitus position and the abdominal viscera is reversed. In ‘isolated situs inversus,’ the thoracic viscera is reversed and the abdominal viscera is in the solitus position. ‘Heterotaxy’² refers to cases of random sidedness, such that some organs are reversed and others are not. These cases are classified as ‘laterality sequences’. Patients with these conditions appear to be either predominantly left sided or right-sided bilaterality. A left-sided bilaterality sequence is associated with polysplenia. A right-sided bilaterality sequence is associated with asplenia or hypoplastic spleen. The case of isolated stomach on the right-side presented here, therefore, is classified as “Heterotaxy involving stomach - right sided bilaterality sequence.” It is associated with asplenia and transpositioned mirror images of abdominal esophagus and duodenum.

During the gastrulation period (3rd and 4th week of gestation) of embryogenesis, the body axes, including left-right axis, are established. A secreted factor, sonic hedgehog (shh), first uniformly appears in the primitive node. Soon activin IIa receptors appear on the right side of the node, which when occupied by activin restricts ssh expression on the left side of the node where it activates transforming growth factor beta family genes namely nodal and lefty. Members of the snail family of zinc finger proteins are expressed and restricted to the right side

of the node. These growth factor genes, then, regulate the left-right asymmetry such as heart, spleen, stomach to lay on the left side and liver on right side. These genes further through a HOX code in splanchnic mesoderm and shh expression in the endoderm of primitive gut dictate differentiation, derivations, type of site specific organ formation by reciprocal interaction; for example lungs and stomach in foregut colon in hindgut and so forth. In heterotaxy, however, the above expressions are, somehow reversed at random to mirror image formation of a few organs either with left sided or right sided bilaterality sequence. In the present case of heterotaxy, the other organs and structures involved could be pancreas, gall bladder, common bile duct, omenta, neuro vascular supply including vagii, duodenal mesentery and so forth. However, this could not be confirmed radiologically.

References

1. Sutton D. A text book of radiology and Imaging. 4th ed. UK: Long man group UK Limited; 1987. p. 614- 616.
2. Sadler TW. Lang man's Medical Embryology. 8th ed. USA: Lippincott Williams and Wilkins; 2000.