SYNDROME! POLYSPLENIA IN A 10 YEAR OLD GIRL

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ABSTRACT:

We present a case of 10 year old patient with polysplenia, situs inversus abdominus, malrotation of bowel and renal anomalies.

Polyspenia Syndrome refers to presence of two or more spleens associated with various abnormalities in chest and abdomen. Situs inversus is present in about 20% of cases Polysplenia Syndrome in association with situs inversus abdominus, incomplete malrotation of bowel and renal anomalies is rarely reported.

Keyword: Polysplenia, situs inversus abdominus, incomplete malrotation and ectopic kidney.

INTRODUCTION

Polysplenia syndrome was described by Helwig in 1929. It is a rare hereditary syndrome that presents in childhood and only 10% of the patients without cardiac anomalies reach adulthood.

The syndrome is associated with heterotaxy of various organs, cardiac malformations, bilary atresia, abnormality of vena-caval bowel malrotation, pulmonary and genitourinary malformations, less frequent anomalies include intestinal atresia, short and annular pancreas and pre duodenal portal veins.

Case Presentation:

A 10 year old girl presented to the emergency room with 3 months history of recurrent abdominal pain. No associated vomiting and nausea. Physical examination was normal except mild tenderness in the suprapubic region. Laboratory results were also normal. Abdominal ultrasound revealed left sided liver and multiple spleens in the right upper quadrant. The left kidney was ectopic in the pelvis and there was moderate hydronephrosis of right kidney. C. T. Scan of abdomen and Pelvis was done to corroborate the ultrasound findings.

C. T. Scan Results:

1. The Liver was on the left side.
2. Three spleens of various sizes were located in the left upper quadrant.
3. The stomach was located in the right upper quadrant.
4. The small bowel was mostly on the right side of upper with abnormality high position of ileocecal junction.
5. The left kidney was on the left side of the pelvis and there was pelvi-ureteric junction obstruction of right kidney.

The chest radiograph revealed right sided aortic arch.

Echocardiogram was normal.

DISCUSSION:

Polysplenia is the presence of two of more spleen and polysplenia syndrome refers to its association with various organ abnormalitites in abdomen and chest.

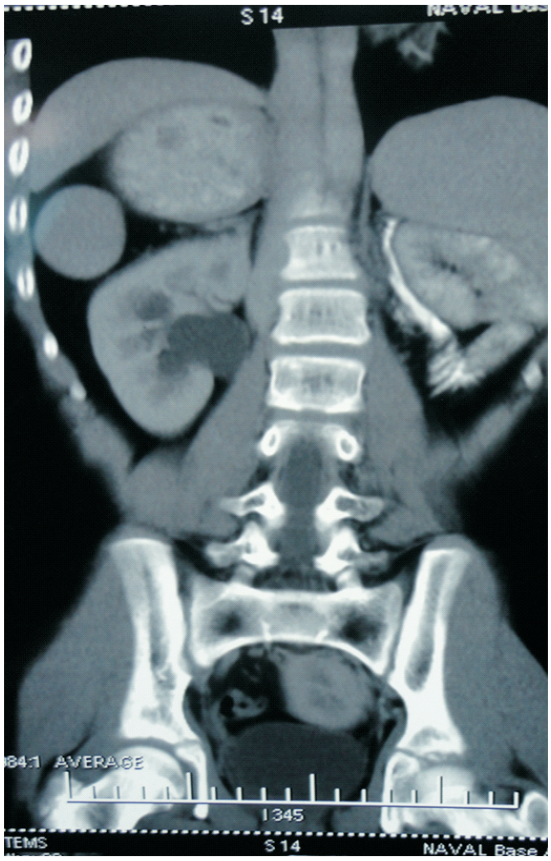
Polysplenia syndrome is common in females and usually presents in infancy and early

childhood. Situs inversus is present in 20% of cases. More than 40% of patients have cardiac anomalies and majority do not survive to adulthood. Only 23 cases have been reported in adults.

Most cases are diagnosed incidentally during abdominal surgery for some other reasons. The presentation depends upon the presence and severity of associated anomalies. The common symptoms are usually vague abdominal pain, nausea and vomiting.

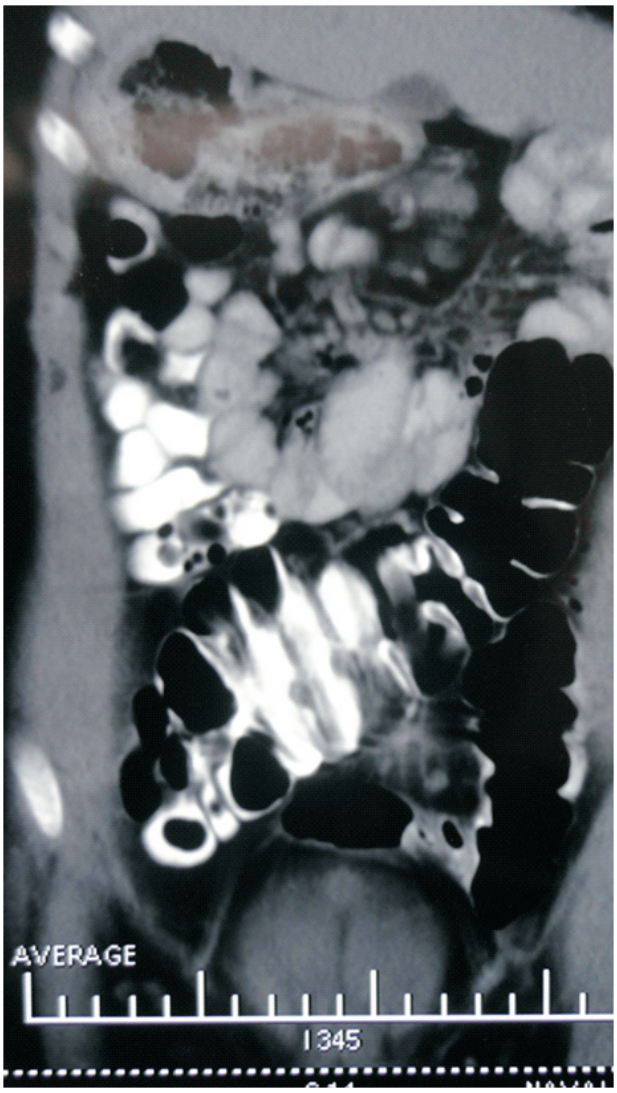
The case we present unites most of the characteristics of polysplenia syndromes. Very few reports have described the presence of polysplenia with situ-inversus abdominus, incomplete malrotation of bowel and anomalies of kidneys –left ectopic kidney and pelvi-ureteric junction obstruction of right kidney.

Fig. 1 Coronal Sections of CT scan of Abdomen.



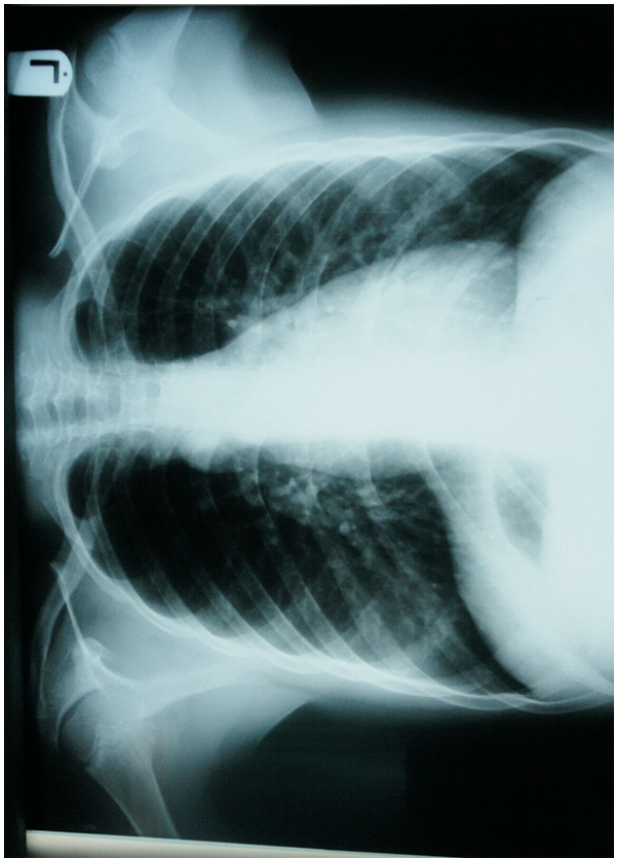
1. Left sided liver.
2. Right sided spleen and Stomach
3. Pelvic-ureteric obstruction of right kidney.
4. Left ectopic kidney.

Fig.2 Coronal Sections of CT scan of Abdomen.



1. Abnormal positions of small bowel and transverse colon.

Fig. 3 PA Radiograph of the Chest.



Right sided aortic arch.

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