An Incidental Finding of Polysplenia Syndrome in a Geriatric Patient: A Case Report

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ABSTRACT

Polysplenia syndrome is a rare congenital disease characterized by presence of two or more spleens and affection of other asymmetric organs including heart, lungs, liver and intestines. It is a rare congenital disease often recognized in childhood and rarely in adulthood. Here we report a case of polysplenia syndrome in a geriatric patient, as an incidental finding.

Keywords: Polysplenia, polysplenia syndrome

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CASE REPORT

A 75-year-old female patient was admitted in our hospital with a history of lower backache of 1 month duration. On enquiry, patient gave history of nonproductive cough, dyspnea, swelling of lower limbs and intermittent, colicky and vaguely located abdominal pain. On examination, she was having pulse rate of 80/min with irregularly irregular rhythm. Her blood pressure (BP) was 140/90 mmHg. Edema was present in lower limbs and neck veins were engorged with raised jugular venous pressure. Cardiovascular system showed systolic murmur in mitral and tricuspid area. Respiratory system showed scattered bilateral crepitations. On palpation, there was tenderness over lumbar vertebra. Per abdomen and central nervous system were within normal limit. Investigations revealed hemoglobin - 11.9 g/dL, total leukocyte count (TLC) - 5,200/mm³, differential leukocyte count (DLC) - N65L30E5, erythrocyte sedimentation rate (ESR) - 36 mm 1st/hour, platelet count - 1.96 lacs/mm³, blood sugar random (BS[R]) - 80 mg/dL, urea - 25 mg/dL, creatinine - 0.6 mg/dL, Na - 140 mmol/L, K - 3.5 mmol/L liver function tests (LFTs) - normal and renal function tests (RFTs) - normal. Electrocardiography (ECG) showed atrial fibrillation with left ventricular hypertrophy (LVH). Ultrasonography (USG) showed liver in midline and more towards left side, hilum leftward, but inferior vena cava (IVC) and hepatic veins on right side and were normal.

No spleen on left side was identified with an impression of situs ambiguous abdominis. Echo showed dilated right (R) atrium and R ventricle, LVH, mild mitral regurgitation, mild aortic regurgitation, mild tricuspid regurgitation with mild pulmonary arterial hypertension and normal left ventricular ejection fraction. High-resolution computed tomography (HRCT) thorax showed evidence of focal fibrosis in left lung, predominantly in peripheral aspect with suggestion of volume loss. Mild honeycombing with septal thickening in anterior segment of right lobe medially and a small emphysematous bulla in apical segment of left (L) upper lobe medially was seen.
Contrast-enhanced computed tomography (CECT) abdomen was performed on a 128 slice dual source dual energy scanner and it showed evidence of liver, gallbladder, IVC and common bile duct on left side of abdomen. Stomach, aorta and spleen with evidence of multiple accessory spleens (at least 3) were on right side of abdomen (Figs. 1-3). A final impression of situs inversus with polysplenia syndrome was made on CECT abdomen.

DISCUSSION

Polysplenia is a presence of two or more spleens in a patient and polysplenia syndrome refers to its association with multiple congenital abnormalities in abdomen and chest. It is a rare congenital disease with incidence of 1/2,50,000 live births and was initially described by Helwig in 1929.2,3

The exact cause of polysplenia has not been defined. The probable hypotheses are embryonic such as accelerated curvature of the embryonic body, genetic causes and teratogenic factors.2 There is slight female preponderance and rare familial association has also been found.2

The splenic mass is usually divided into fairly equal-sized masses, varying in number from 2 to 6 and ranging from 1 to 6 cm in diameter, which together equal the mass of a normal spleen. The location of the spleens is in either the left or right upper quadrant, along the greater curvature of the stomach.4

More than 40% cases of polysplenia syndrome have cardiac anomalies and majority of such children do not survive beyond 5th year of life.5 The various cardiovascular anomalies that may be encountered are atrial septal defect (ASD) (78%), ayzygous continuation of the IVC (65%), ventricular septal defect (VSD) (63%), bilateral superior vena cava (SVC) (47%), right-sided aortic arch (44%), partial anomalous pulmonary venous return (39%), transposition of the great arteries (31%), pulmonary valvular stenosis (23%) and subaortic stenosis (8%). The abdominal findings include midline liver (57%), situs inversus (21%), short pancreas, semiannular pancreas, preduodenal portal vein and malrotation. Renal agenesis or hypoplasia may also be seen. The pulmonary manifestations include bilateral bilobed lungs and hyparterial bronchi (58%).2,6,7

The presentation of polysplenia syndrome depends upon the presence and severity of the associated anomalies. About 10% of polysplenia cases reach adulthood and are commonly detected on diagnostic work-up of other associated disease.5

Plain chest X-ray and abdomen can give clue about abnormality in chest and abdomen. CT scan, magnetic resonance imaging (MRI), angiography and echocardiography are important tools for ascertaining the location and number of spleens, location of other...
organs in the chest and abdomen, and identification of other associated anomalies.

We report a case of elderly female of polysplenia syndrome with lumbar vertebra collapse, congestive cardiac failure and atrial fibrillation. On evaluation, she was found to have liver, gallbladder, IVC and common bile duct on left side of abdomen and stomach, aorta and spleen with evidence of multiple accessory spleen on right side of abdomen, as evidenced on CECT abdomen.

Peoples et al conducted a study on 146 cases of polysplenia, which showed both lungs with two lobes in 55% of patients, abdominal heterotaxy in 56% cases and cardiac anomalies occurring in at least half of the patients, which included bilateral SVC, interruption of the IVC with azygous continuation, VSD, ostium primum defect and morphologic left ventricular outflow tract obstruction. Fifty percent patient died by 4-month of the age and 75% before 5 years of age.7 Choh et al reported a case of polysplenia syndrome in a 40-year-old male patient. He was having multiple splenic masses, midline liver and stomach on right upper quadrant and also VSD with congenital lobar emphysema.6 Rasool and Mirza reported a case of polysplenia syndrome associated with situs inversus abdominis and jejunal atresia in a 2-day-old female baby.5

CONCLUSIONS

Polysplenia syndrome is a rare congenital disorder of childhood characterized by presence of two or more spleens with affection of other asymmetric organs and usually diagnosed incidental in adult.

Acknowledgment

I take this opportunity to extend my gratitude and sincere thanks to all those who helped me to complete this study. I am highly thankful to Dept. of Medicine, Pathology, Biochemistry and Radiodiagnosis for providing me adequate facility, which helped me to carry out this study. I owe great sense of indebtedness to Dean SRMS-IMS, Bhojipura, Bareilly, Uttar Pradesh for permitting me to carry out this study.

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