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Exceptionally Enlarged Accessory Spleen Due to Portal Hypertension

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ABSTRACT

Accessory spleens, also known as supernumerary spleens, splenunculi, or splenules, are congenital foci of healthy splenic tissue that are separate from the main body of the spleen [1]. They are found incidentally in 10-30% of patient at autopsy, mostly at the hilum of the spleen and ad-jacent to the tail of pancreas in size of few millimeters to 2-3cm [1-3].
Reported 14 yr. old female child present with fever, pain abdomen, and generalized swelling. child have severe pallor and massive hepatosplenomegaly of 5cm and 7cm respectively with a lump of approximate size of 7cmx3cm involving umbilicus and left hypocondrium and confirmed as exceptionally enlarge accessory spleen in USG abdomen. CT scan confirmed presence of portal hypertension with collateral. Hematology revealed pancytopenia with normal hb elec-trophoresis.
It has been reported that accessory spleens undergo compensatory hypertrophy as a result of pre-vious splenectomy, which sometime reaches 3-5 cm in size [19] .The accessory spleen seen in the present case seems to be exceptionally large due to portal hypertension[20]. Unusual presentation of massive splenomegaly with significant pancytopenia and portal hypertension may lead to exceptionally enlarge accessory spleen.

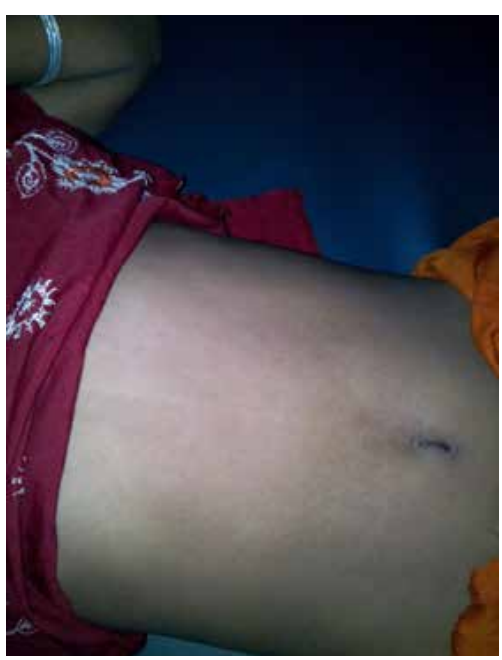
KEYWORDS

CASE REPORT

A 14 yr. old female child was admitted in department of pediatrics with a history of fever for 4 days, pain abdomen for 4 days and generalized body swelling. Fever was high grade, intermittent, associated with chills and rigor. Abdomen pain in left upper abdomen, dragging in nature and moderate in intensity and generalized body swelling started from feet, gradually increasing and progressing to face. In past, patient had similar complaints three times and for each episode patient was hospitalized for 3-4 days. First episode was at the age of 10 year and in each episode patient had multiple blood transfusions for severe anemia.

On general examination patient had severe pallor, no cyanosis, no clubbing, no lymphadenopathy on systemic examination on abdominal inspection bulging in left hypocondrium .On abdominal palpation patient had Liver 5cm below costal margin at mid clavicular line and spleen approximate 7cm below costal margin. One accessory lump was also palpable approximate size 7cm x 3cm involving umbilicus and left hypocondrium.no tenderness, no guarding, no rigidity. Other systems were normal on examination.

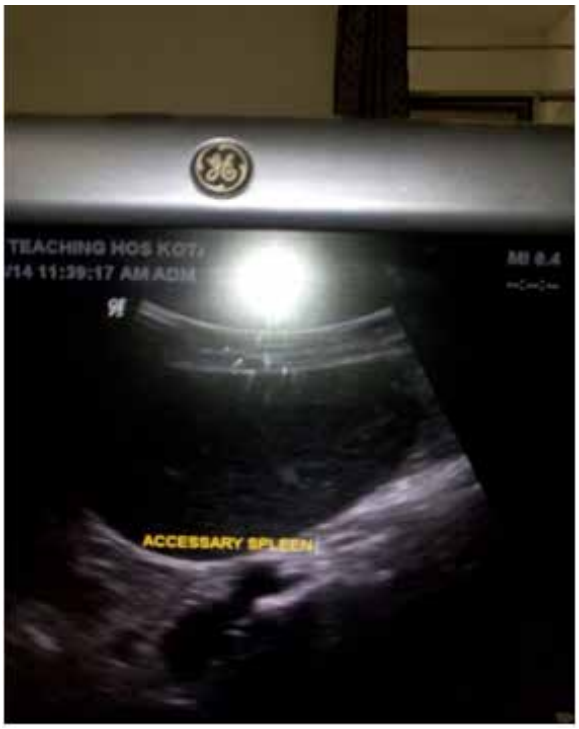
HEMATOLOGICAL INVESTIGATIONS revealed pancytopenia. Blood profile showing WBC .93X10‹3, RBC 1.44X10‹6 and platelet 42000, Peripheral smear showed: predominantly macrocytic normochromic anemia, hypersegmented neutrophil, few tear drop and target cell seen . Polychromatic RBC with few fragmented RBC present. Hb electrophoresis reveled HbA2 level 2.8% and Hb F level 1%.



NEUROIMAGING BY USG ABDOMEN

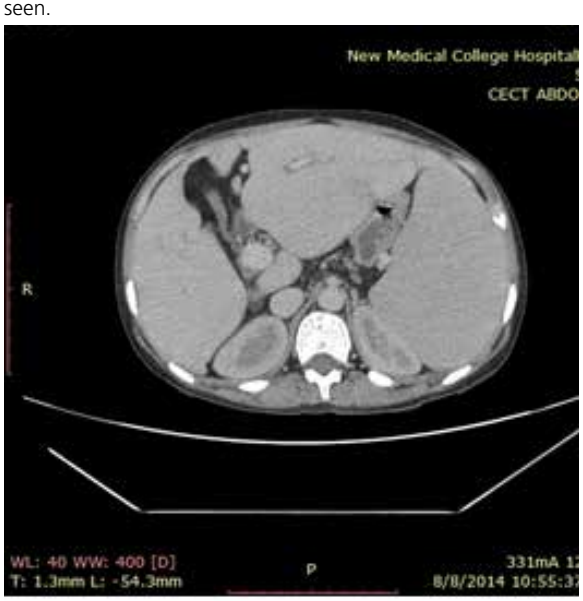
showed moderate hepatomegaly with massive splenomegaly with ACCESSORY SPLEEN total size of spleen 210mm with

accessory spleen 9x3 cm in size.



CT SCAN

splenomegaly with portal hypertension(dilated splenic 13mm and portal vein 16mm) and multiple umbilical, linorenal, and linrno portal collateral are seen. A 51x83x103 mm size accessory spleen at umbilical level is seen.



DISCUSSION

Ectopic splenic tissue arises from either of two discrete histological entities: splenosis or accessory spleen. Splenosis is due to heterotopic auto transplantation of splenic tissue following splenectomy or other splenic trauma. It occurs when disruption of the splenic capsule leads to spilling of viable splenic tissue implants into the abdomen and subsequent Seeding onto the peritoneum. Nodules can occur anywhere within the peritoneal cavity and even within the chest where intrathoracic splenosis has been reported following splenic injury accompanied by diaphragmatic tears[17].

Accessory spleens , are congenital foci of healthy splenic tissue, they arise from the failure of fusion of the splenic anlage, located in the dorsal mesogastrium, during the fifth week of fetal life [1-2]. Detection and characterization of accessory spleens are important in three clinical scenarios. First, an accessory spleen may mimic lymphadenopathy and tumors in other abdominal organ such as the pancreas, the adrenal gland, and the kidney [7-10]. Second, accessory spleens occasionally may become symptomatic because of torsion, spontaneous rupture, hemorrhage, and cyst formation [11–14]. Third, a surgeon's awareness of their presence may be important when the intention is to remove all functional spleen. Because remaining accessory splenic tissue may undergo compensatory hypertrophy after splenectomy, and may cause return of symptoms. [15-16] . The accessory spleen, which will inevitably enlarge in time, may grow significantly within a short period of time in the presence of portal hypertension and may thus be misdiagnosed as a tumoral mass[20].

CONCLUSION

An enlarged accessory spleen appearing as a mass on CT and ultrasonography is reported herein. Beahrs et al. observed no accessory spleen larger than 2.5 cm among more than 8000 abdominal CT examinations of patients with normal-sized intact spleen[18] .It has been reported that accessory spleens undergo compensatory hypertrophy as a result of previous splenectomy, which sometime reaches 3-5 cm in size[19] .The accessory spleen seen in the present case seems to be exceptionally large despite no history of splenectomy due to portal hypertension[20].

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