The culprits behind a hyper-intense spleen

Sir,

A calcified and atrophic spleen is not a common finding and warrants a thorough work up to elucidate the etiology. Although there are relatively few differential diagnoses, the pattern and the setting in which such calcification presents, helps us pin-point the diagnosis. We would like to discuss one such case we came across. A 35-year-old male patient previously diagnosed as sickle cell disease, presented with fever, shortness of breath, multiple loose stools, occasional vomitings, abdominal pain and jaundice of one week duration. In the past, he received multiple blood transfusions, underwent cholecystectomy for gallstones and total hip replacement (THR) on the right for avascular necrosis. Examination revealed tachycardia, tachypnea, deep icterus, pallor, bilateral pitting pedal edema with raised jugular venous pressure and diffuse abdominal tenderness without rigidity or guarding. Peripheral smear showed tear drop cells, sickle cells, anisocytes, poikilocytes, and neutrophilia with toxic granulations. X-ray abdomen [Figure 1] showed air-filled bowel loops, calcified atrophic spleen (arrow) and Total Hip Replacement THR prosthesis on the right. Computed tomography (CT) scan of the abdomen [Figure 2] showed ascites and delineated the atrophic spleen (arrow).

Non homogenous or focal splenic calcifications encompass a wide variety of differential diagnoses like echinococcal cysts (visualizing daughter cysts helps differentiate), infection (pyogenic or tuberculous abscesses), infarction (presenting as triangular or wedge-shaped lesion with a broad capsular base), splenic artery aneurysm, dermoid, epidermoid, simple cyst, phleboliths, and old hematoma. The diffuse splenic calcifications can be either punctate (starry spleen) or uniform. Diffuse punctate calcifications of spleen are seen in brucellosis (associated with suppurating lesions), tuberculosis, pneumocystis jiroveci (commonly associated with kidney and lymph node calcifications), candidiasis, histoplasmosis (larger lesions and usually more than six in number), and amyloidosis (along with calcifications in liver). The homogenously calcified appearance is seen in sickle cell disease and occasionally in thorotrast administration. The higher density and the associated signal intensities of abdominal lymph nodes distinguishes thorotrast related diffuse splenic hyper intensity from that of sickle cell disease. Primary hemochromatosis is characterized by a unique peripheral egg-shell like splenic calcification and a rounded intra-splenic calcification. There are various mechanisms resulting in splenic calcification that depend mainly on the underlying disease process like calcification of thrombi in vasculitis and hematomas and deposition of calcium salts during the healing phase of focal, granulomatous inflammatory lesions in tuberculosis and
histoplasmosis, hyalinization, and calcification post inflammation in infections, etc.

Spleen in sickle cell patients may be atrophic and shrunken, a phenomenon termed as autosplenectomy. Radiological evidence of a diffuse uniform calcification (especially if associated with hemochromatosis from the repeated blood transfusions) strikingly evident in our case, is not very common. It points toward chronicity and functional asplenic state predisposing to fulminant sepsis (from overwhelming post-splenectomy infection) in which case the mortality may be more than 50%. The spectrum of causative organisms (particularly from polysaccharide-encapsulated bacterial infections[3]) is evolving and preventive strategies like education, prophylactic and standby antibiotics, preventive immunizations, optimal antimalarial advice when visiting endemic countries and early management of animal bites may help curb such devastating catastrophes.

Acknowledgement

We thank our colleagues and staff of internal medicine, nephrology, and critical care.

Dilip Gude, Dharam P. Bansal¹, Sashidhar Chennamsetty²,
Ratan Jha²

Departments of Internal Medicine, ¹Pulmonology and Critical Care, ²Nephrology, Medwin Hospital, Nampally, Hyderabad - 500 001, Andhra Pradesh, India

Correspondence to: Dr. Dilip Gude,
Department of Internal Medicine, AMC, 3rd Floor, Medwin Hospital, Chirag Ali lane, Nampally, Hyderabad - 500 001, Andhra Pradesh, India.
E-mail: letsgo.dilip@gmail.com

References

1. Singh S, Mukhopadhya A, Chandy GM, Korah IP. Splenic calcification in primary hemochromatosis mimicking hydatid cyst. Indian J Radiol Imaging 2000;10:268-89.
2. Topin J, Mutlu GM. Images in clinical medicine. Splenic and mediastinal calcifications in histoplasmosis. N Engl J Med 2006;354:179.
3. Pearson HA. Sickle cell anemia and severe infections due to encapsulated bacteria. J Infect Dis 1977;136 Suppl:S25-30.