VOLUME - 11, ISSUE - 11, NOVEMBER - 2022 • PRINT ISSN No. 2277 - 8160 • DOI : 10.36106/gjra

Original Research Paper

General Surgery

MULTIPLE SPLENUNCULI IN HEMOLYTIC ANEMIA

Dr K Madhuri Reddy*	Post Graduate Resident in General Surgery, Prathima Institute Of Medical Sciences, Karimnagar, Telangana State *Corresponding Author
Dr Rajashekar Addagatla	Associate Professor in General Surgery, Prathima Institute Of Medical Sciences, Karimnagar, Telangana State
Dr E Soumya	Post Graduate Resident in General Surgery, Prathima Institute Of Medical Sciences, Karimnagar, Telangana State
Dr G Sai Pranathi Gaddipati	MBBS Graduate

ABSTRACT **Aim:** The aim of the present manuscript is to present the cases of multiple accessory spleens in hemolytic anemia patients and a review of the literature. **Methods:** We report two cases of accessory spleen in cases presenting with splenomegaly in hemolytic anemia operated for various reasons. On examination-both the cases had abdominal dissension and palpable spleen which was nontender, smooth in surface and firm in consistency, margins well defined. Contrast enhanced CT- showing mass in splenic hilum measuring 28.8mm X16.7mm, likely accessory spleen.(In case 2). On exploration-incidental finding(In case 1) **Results:** Histopathological examination revealed two or more nodules with encapsulated red pulp confirmative of Splenunculi. (In both cases). **Conclusion-** They are important in certain clinical scenarios like lymphadenopathy and tumors in other abdominal organs. They can cause life threatening complications like torsion, spontaneous rupture, hemorrhage. If left behind at the time of splenectomy it can lead to hypertrophy leading to recurrence of initial disease.

KEYWORDS : Splenunculi, accessory spleen, splenosis

INTRODUCTION:

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. They arise from the failure of fusion of the splenic anlage, located in the dorsal mesogastrium, during the fifth week of fetal life.Accessory spleens are relatively common and are seen in 10–30% of patients at autopsy. Accessory spleens are usually single (85%); in some patients there are two (14%) and only in particular cases are found three or more (1%). Although usually asymptomatic and incidentally discovered, they are clinically important in some patients.

Case Reports

Case 1-

A 24 year lady presented with complaints of abdominal mass since 11years and abdominal pain since 7 days. Received multiple transfusions. Gradually progressing mass, h/o of on and off fever. She was thin built, She presents with severe anemia, facial puffiness, thalassemia facies , pedal edema pallor. O/e- Abdomen was distended, umbilicus central in position, swelling extending from left coastalamrgin to 5 cm below umbilical towards right iliac fossa, non tender,smooth surface, margins well defined, firm in consistency.

Surgery-midline laparotomy incision given, massive spleen extending from right iliac fossa, umbilical and left hypochondrium seen, hilum identified, 2 Splenunculi close to each other identified in the hilum. Splenectomy was performed along with the excision of splenunculi

Macroscopic Examination:

On cut section hilum shows two nodules of size largest measuring 1.5x1cm, smallest measuring 1x1 cm(accessory splenic tissue). **(Figure 1)**

Microscopic Examination:-

Well encapsulated, shows red pulp with markedly decreased to absent white pulp areas. Shows red pulp congestion.

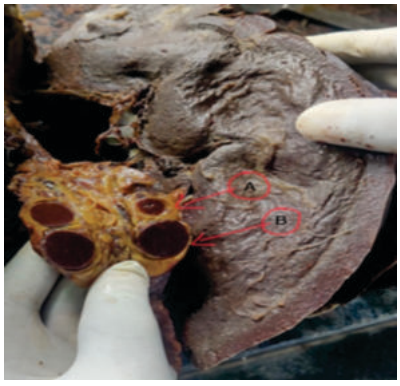

Figure 1- cut section of splenic hilum

Case 2-

A 9 years old girl(mother being the informant) child has complaints of abdominal mass and abdominal distension since 3 years. Known case of B thalassemia(have received 4-transfusions in the past).

On examination- abdomen was distended, spleen palpable upto umbilicus which was nontender, smooth in surface and firm in consistency, fingers could not be insinuated above swelling measuring 16x9 cms ,showing positive castell sign. CT abdomen showing mass in splenic hilum measuring 28.8mm X16.7mm, likely accessory spleen.(**Figure 2)**

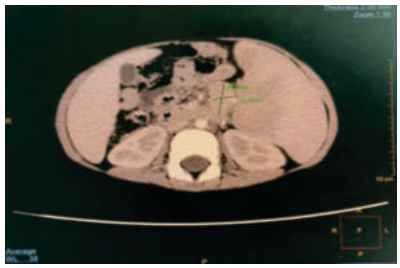

Figure 2- CT abdomen showing mass in splenic hilum.

Surgery -
transverse subcoastal incision of 6 cms given. 1cm accessory spleen found-at Hilum ,1cm accessory spleen found near gastrosplenic ligament of spleen, 3cm accessory spleen found embedded in tail of pancreas. Splenectomy was performed along with the excision of splenunculi**(figure 3)**

Macroscopic Examination:
On cut section Spleen measuring 16.5x9x7.5em. Externally grey black to slate color. Hilum shows two nodules of accessory spleen of size largest measuring 2x2x0.7cm, smallest measuring 0.8x0.5cm. Weight 456gm. **(Figure 3 and 4- showing Splenunculi in perisplenic area)**

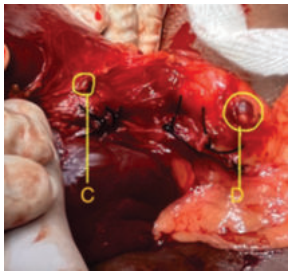

Figure 3-

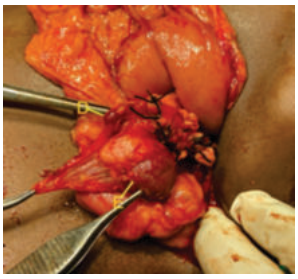

Figure 4-

Microscopic Examination:-
Sections examined show capsule composed of fibrous tissue. Subcapsularly splenic red and white pulp seen with expansion of red pulp.

Red pulp showing dilated, congested vessels along with dilated sinusoids and extramedullary hematopoiesis.

Section from accessory spleens show red pulp congestion with similar morphology. Splenunculi also seen adjacent to pancreatic tissue, and adipose tissue.

DISCUSSION:
The spleen develops as a collection of mesenchymal cells in the dorsal mesogastrium. The development occurs during sixth week of intrauterine life.Some of these cells are contributed by the coelomic epithelium lining the mesogastrium. The mesenchymal cells differentiate into lymphoblasts and other blood forming cells.As the mesenchymal cells proliferate, they Form a mass which projects to the left and is covered by peritoneum. Number of nodules develop which soon fuse to form a lobulated spleen.Notching of the superior border of the adult spleen is an evidence of its multiple origin. These nodules which fail to fuse, (while migrating from the midline to the left upper quadrant) form accessory spleens. **Figure 5** shows the usual sites of accessory spleens.

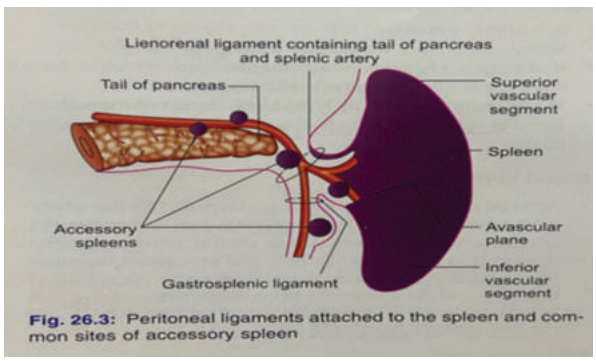

Figure 5- common sites of accessory spleen.

An accessory spleen can be found in In the derivatives of the dorsal mesogastrium, i.e. gastrosplenic ligament, lienorenal ligament, gastrophrenic ligament.(most common)

The common sites are:Table-1

a. Perisplenic Area	Hilum (50%),Splenic vascular pedicle, Tail of pancreas
b. Greater omentum	Along the greater curvature of stomac
C. Mesentery	Small and large bowel
d. Pelvis and Groin	-Left Broad ligament,Pouch of Douglas,Left testicles

Importance:
If left behind at the time of splenectomy, can hypertrophy leading to recurrence of initial disease.Accessory spleen can be present in 16% of the population , and they are found in 10–30% at autopsy. Accessory spleens are usually single (85%); in some patients there are two (14%) and only in particular cases are found three or more (1%) [2,17]. In this case, the total number and volume of all supernumerary spleens exceeded the normal volume of the only one natural spleen. Macroscopically , a typical accessory spleen usually appears as a solid mass, 1–2 cm in diameter And singular. The size bigger than 2cm and presence of two or more Splenunculi in a single patient is realtively rare. Microscopically, it reproduces a splenic pattern. An accessory spleen commonly has a well-defined fibrotic capsule that separates the surrounding normal tissue.

Other congenital and acquired splenic anomalies, including splenic clefts, lobulations, polysplenia, and splenosis, should be differentiated from accessory spleens. Splenic clefts are a result of incomplete fusion of the splenic anlage and usually are visible under the diaphragmatic surface. Splenic lobulations persist after fetal life. They usually are seen along the medial part of the spleen and sometimes are supplied by an early branch of the splenic artery. Polysplenia is a congenital syndrome seen in patients with bilateral left-sidedness, in which two to 16 splenic nodules of equal size can be found in the right or left upper quadrant (depending on the associated situs). Cardiovascular and pulmonary abnormalities are associated with polysplenia in most patients. Similar to accessory spleens, splenosis is a condition in which isolated foci of heterotopic splenic tissue are present .However, unlike accessory spleens, splenosis is an acquired condition and originates from seeding or implantation of splenic cells after splenectomy or splenic rupture (autotransplantation). Splenosis nodules usually are small as a result of limited blood supply. They show a sessile growth pattern and are found typically adjacent to small-bowel serosa, the greater omentum, the parietal peritoneum, and the diaphragm. Their blood supply derives from neovascularization and is not of embryologic origin, as is the case with accessory spleens.

Splenomegaly in hemolytic anemia-
Splenomegaly is defined as enlargement of the spleen measured by weight or size. The spleen plays a significant role in hematopoiesis and immunosurveillance. The major functions of the spleen include clearance of senescent and abnormal erythrocytes and their remnants, opsonized platelets and white blood cells and removal of microorganisms and antigens. The spleen also serves as a secondary lymphoid organ and is the site for maturation and storage of T and B lymphocytes, playing an important role in the synthesis of immunoglobulin G (IgG) by mature B-lymphocytes upon interaction with the T-lymphocytes. The spleen also synthesizes the immune system peptides properdin and tuftsin. Approximately one-third of circulating platelets are stored in the spleen.

The mechanism underlying splenic enlargement varies based on the etiology There are several potential causes of splenomegaly.

Liver disease (cirrhosis, hepatitis), Hematologic malignancies

(lymphomas, leukemias, myeloproliferative disorders), Venous thrombosis (portal or hepatic vein thrombosis), Splenic congestion (venous thrombosis, portal hypertension, congestive heart failure), Cytopenias (Immune thrombocytopenic purpura, autoimmune hemolytic anemia, immune-mediated neutropenia, Felty syndrome), Splenic sequestration (pediatric sickle cell disease, hemolytic anemias, thalassemias), Acute or chronic infection (bacterial endocarditis, infectious mononucleosis, HIV, malaria, tuberculosis, histiocytosis, abscess), Connective tissue diseases (systemic lupus erythematosus, rheumatoid arthritis, Adult-onset Still's disease, and some familial autoinflammatory syndromes), Infiltrative disorders (sarcoidosis, amyloidosis, glycogen storage diseases), Focal lesions (hemangiomas, abscess, cysts, metastasis).

Accessory spleen also enlarges in such conditions along with splenomegaly since it has the same function as spleen. Hence removal of

CONCLUSION-

The detection and characterization of accessory spleens are important in three clinical scenarios. First, an accessory spleen may mimic lymphadenopathy and tumors in other abdominal organs, such as the pancreas, the adrenal gland, and the kidney.Second, accessory spleens occasionally may become symptomatic because of torsion, spontaneous rupture, hemorrhage, and cyst formation. Third, a surgeon's awareness of their presence may be important when the intention is to remove all functional splenic tissue (e.g., hematologic disorders)

REFERENCES:

1) Splenunculi-Report of Three Cases, Harsh mohan https:// www.researchgate.net/profile/Harsh-Mohan-4 https:// www. researchgate.net/publication/265924003_Splenunculi-Report_of_Three_Cases
2) Palumbo, Valentina & Mannino, Maurizio & Teodoro, M. & Menconi, G. & Schembari, Elena & Corsale, G. & Carlo, I. & Toro, Adriana. (2019). An extremely rare case of an oversized accessory spleen: Case report and review of the literature. BMC Surgery. 19. 10.1186/s12893-019-0510-z.
3) Koenraad J. Mortelé1, Bart Mortelé2 and Stuart G. Silverman1 CT Features of the Accessory Spleen American Journal of Roentgenology. 2004;183: 1653-1657. 10.2214/ajr.183.6.01831653Read More: https://www.ajronline.org/doi/full/10.2214/ajr.183.6.01831653
4) Adalet Elcin Yildiz, Macit Orhan Ariyurek, Musturay Karcaaltincaba, "Splenic Anomalies of Shape, Size, and Location: Pictorial Essay", The Scientific World Journal, vol. 2013, Article ID 321810, 9 pages, 2013. https://doi.org/10.1155/2013/321810
5) Liu Y, Ji B, Wang G, Wang Y.Abdominal multiple splenosis mimicking liver and colon tumors: a case report and review of the literature. Int J Med Sci. 2012;9(2):174-7.
6) Chapman J, Goyal A, Azevedo AM. Splenomegaly. [Updated 2022 Aug 8]. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2022 Jan-.