ISSN 2394-2118(Print)
e-ISSN 2394-2126(Online)

Research Communication

Congenital Anomalies of Spleen - cadaveric study with special reference to Multilobulated spleen

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Abstract: *Congenital anomalies of the spleen range from splenic lobulation, to accessory spleen to polysplenia, absence of the spleen, duplications, Splenoptosis or wandering spleen. Out of these the accessory spleen is the most common anomalies encountered. All these pathological conditions result from an altered embryogenesis of the spleen and are generally associated with other congenital abnormalities. A number of earlier studies reported on accessory spleens and splenunculi whereas, to our knowledge, there are very minimal studies in the literature concerning congenital anomalies of the spleen particularly multilobulated spleen. Hence the present study was taken up to study the frequency of congenital anomalies in cadavers with special reference to multilobulated spleen. Radiologists should be aware of this condition in order to avoid incorrectly diagnosing this ectopic tissue as metastasis, adenopathy or another solid tumor.*

Keywords: *Multilobulated spleen, Splenoptosis, Splenunculi, Embryogenesis.*

Introduction

Spleen is a largest lymphoid organ situated in left hypochondrium which is one of the major accumulations of lymphoid tissue in the human body. It is an organ which prenatally produces and in the postnatal period controls and removes worn out RBC's from circulation. It begins to develop during the 5th week of intrauterine life from a mass of mesenchymal cells originating in the dorsal mesogastrium as a localised thickening of coelomic epithelium.[1,2,3,4] It is nodular in development but as development proceed these nodules fuse with each other to form spleen. The notch at the superior border of spleen indicates its multinodular origin. Persistence of the nodules may lead to formation of lobulated spleen.

Some congenital anomalies of the spleen are common, such as splenunculi, ectopic spleen, asplenia and accessory spleen, while other conditions are rare, such as wandering spleen and polysplenia, splenogonadal fusion. Though most of these anatomical variants have no clinical significance, an accessory spleen may simulate a tumor in the adrenal gland, pancreas, stomach & intestine or may be the site of the relapse of a hematological disorder. Earlier studies have highlighted on the splenunculi, occurrence of many splenic flexures etc, however a very few cases reported pertaining to lobulated spleen hence an attempt to study the frequency of occurrence congenital anomalies in spleen is studied with special reference to multi lobulated spleen. Awareness of the various splenic congenital variants is important for the radiologist to interpret the imaging of the abdomen as it may be commonly misinterpreted as mass arising from kidney or pancreas.

Materials and Methods

During the routine dissection of abdomen for I MBBS students of J.J.M.Medical College, Davangere & ESIC Medical College & PGIMSR, Chennai, we observed spleen pertaining to its

location, blood supply and looked for any associated congenital variations. After opening the peritoneum, stomach was delineated and observed near the fundic portion of the stomach to locate for the position of the spleen. Further the branches of splenic artery were traced from celiac trunk to the spleen and looked out for any possible variations. The spleen was also observed for its abnormal shape and presence of lobulation, or notches over its superior, inferior and medial borders and presence of accessory splenic tissue. All the observations were recorded. Out of 50 spleens taken in the present study we observed one rare case of a multi lobulated spleen with three distinct lobules.

Observations and Results

In the present study of 50 spleens, we observed 45 spleens were normal in their location and had single branch of splenic artery supplying them and were drained by splenic vein, however 4 cases of splenunculi were also observed where accessory splenic tissue observed were single and situated particularly near the tail of the pancreas.

A case of multilobulated spleen in a male cadaver aged around 50 years was also seen which is been highlighted in the present study. Detailed study of the multilobulated spleen pertaining to its morphometric analysis was done and recorded. In this case after opening the abdominal cavity a single spleen was situated in the left hypochondriac region, near the fundus of stomach. Splenic tissue consisted of three lobules of varying size. Spleen was partially subdivided into lobules due to the presence of deep clefts transversing the medial part of the organ. The splenic notches or clefts traversed completely from superior to inferior border thus dividing it into three distinct lobules. All the three lobules were joined to each other & were covered by separate connective tissue capsule. Each lobule had a separate branch of splenic artery and splenic vein supplying them (Ref. Fig.no.1 & Fig no. 2). Histological features of the tissue were consistent with the spleen. The multilobulated spleen in present case can be explained on the basis of non-fusion of splenic lobules during development. The multilobulated spleen was photographed in situ and later removed from abdominal cavity along with the blood supply and the following parameters were recorded.

The weight of the spleen in the present case was 87 grams. Upper lobule measured about 4.5cm in length, 3.5cm in breadth and 2cm in thickness, Middle lobule measured about 2cm in length, 3.5cm in breadth and 2cm in thickness, with a partial cleft in the middle. Lower lobule measured about 3cm in length, 3.5cm in breadth and 2cm in thickness. Spleen was comparatively smaller in size with two distinct notches running from superior to inferior border. The splenic artery was seen dividing into branches outside the spleen to supply each lobule separately; it was also accompanied by tributaries of splenic vein as depicted in Fig. no 1 & 2. The branches of each lobule were traced to the splenic artery & vein respectively.

Table 1: Showing the incidence of congenital anomalies of Spleen in the present study

Type of spleen	No. of specimens	%
Normal spleen	49	98
Multilobulated spleen	01	2
Total no of spleens	50	100

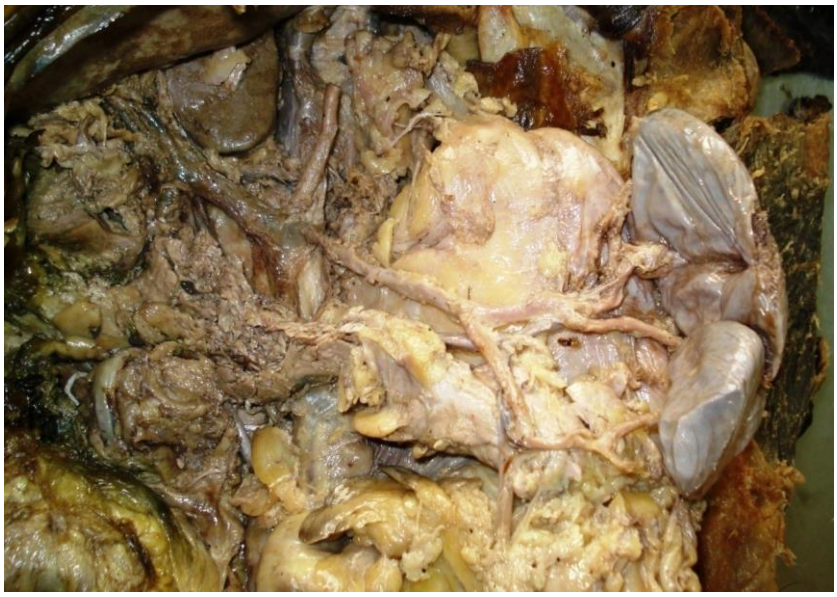

Fig 1: Showing the in-situ photograph of spleen with distinct branch of splenic artery and vein supplying each lobule in a case of multilobulated spleen.

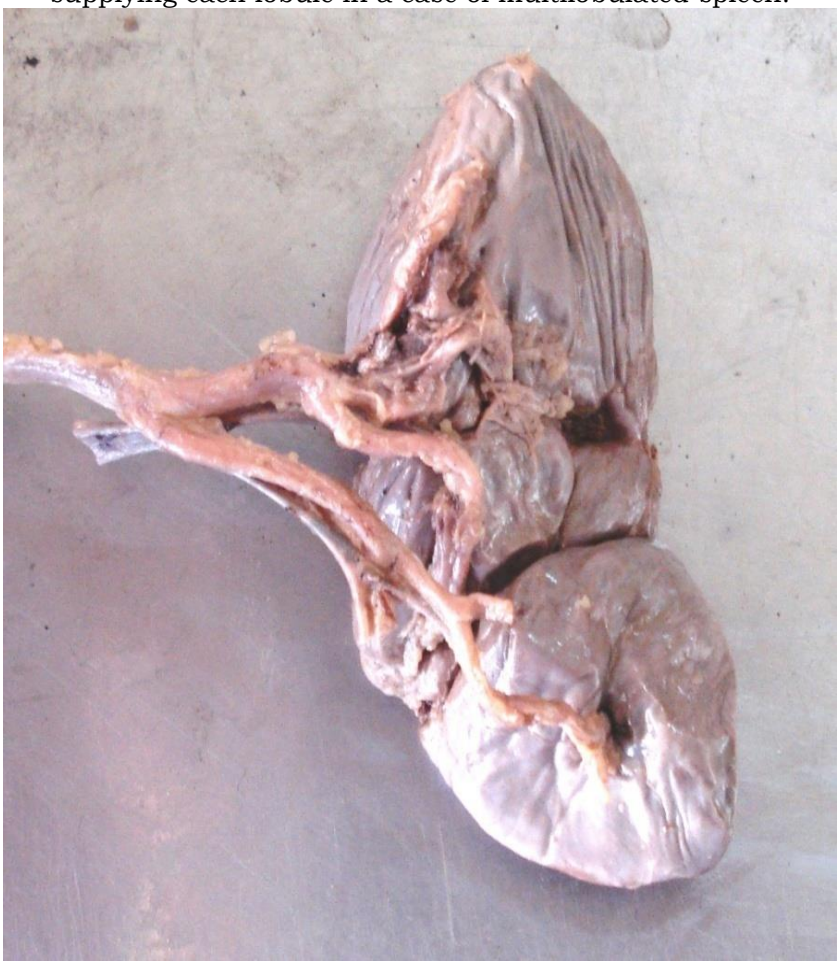

Fig 2: Showing the visceral surface of the Multilobulated Spleen with partial cleft in the middle lobe seen clearly.

Discussion

The congenital absences of spleen (splenic agenesis), congenital hypoplasia, lobulation, polysplenia, accessory spleen (spleniculi) are congenital malformation of the spleen. These malformations usually occur as a result of some changes which occur in the process of development of spleen. The spleen begins to develop during the sixth week of fetal life from a mass of mesenchymal cells originating in the dorsal mesogastrium. [1, 2, 3,4,5,6.] Rotation of the stomach and growth of the dorsal mesogastrium translocate the spleen from the midline to the left side of the abdominal cavity. Rotation of the dorsal mesogastrium establishes a mesenteric connection, the splenorenal ligament, between the spleen and the left kidney. Splenic lobulation is a rare anomaly, this low rate could be due to the fact that fetal splenic lobulation normally disappears before birth but sometimes may persist, causing a prominent lobule and may be erroneously interpreted for a mass in the abdomen. The gastrosplenic and splenorenal ligaments maintain the spleen in its anatomical position. [1, 2, 3,4,5,6.]

A persisting bulge or lobule of splenic tissue sometimes extends medially, anterior to the upper pole of the left kidney. Splenic lobulations may persist along the medial part of the spleen. Sometimes some of these lobules may develop independently leading to the formation of accessory spleens. The embryological reason for having notches on the superior margin is the improper fusion of the splenic nodules along this margin during development. [1,2, 3,4,5, 6,7].

According to Hollingshead, lobulated spleen is the presence of deep notches on superior border or both on superior and inferior borders.[8] Accessory spleen (supernumerary spleen, splenule or splenunculi) is usually a small nodule of splenic tissue found apart from the main body of the spleen.

The majority of accessory spleens, splenic lobulations are discovered by a relapse of chronic hematopathy, primarily treated by splenectomy. Accessory spleens are found in 10% to 30% of patients with hematological diseases. Thus, in this condition, it is important to detect the localization of this anomaly prior to initial splenectomy to reduce the risk of missing it and to allow complete removal of both the main and accessory spleen, in order to avoid recurrence of hematological disease after splenectomy[9].

In the earlier study by Shilpa N. Shewale, she has described a case report of multilobulated spleen consisting of 6 lobules of variable sizes. Each having their own separate artery coming from the main splenic trunk which correlates with the present study.[10] Multilobulated spleen were also reported in earlier studies of Dr. Muktyaz Hussein et.al, with an incidence of 9.3%[11] however the incidence in our study is 2% which is comparatively lower than the earlier study. The embryological basis for multilobulated spleen in the present study could be due to the failure of the fusion of fetal lobulations. However no other associated congenital malformations were noted with respect to other abdominal organs in the same cadaver.

The incidence of congenital anomalies in the present study was 10%. Splenic lobulation is usually a rare variation; this low rate could be due to the fact that fetal splenic lobulation normally disappears before birth. However in the present study the incidence of splenic lobulation is 2%. [1,2,3,4,8,9,10]

Splenic arterial divisions into superior, middle and inferior polar arteries have earlier been reported by Gupta[12] and Dixon[13]. In the present study splenic artery and vein divided into superior middle and inferior polar branches & tributaries respectively.

In our study all the spleens presented with anotched superior border. The presence of the notches on the superior margin is useful for the physician to palpate the spleen during enlargement of spleen. In the previous studies the splenic notches on the superior margin were

seen in 98% by Das et al., [14], 78.6 % by Skandalakis et al.,[15] and 50% by Satheesha Nayak B et al., [16]respectively.

Preoperative CT and preoperative selective spleen scintigraphy are able to identify only 25% of accessory spleen and lobulated spleen. However, an accessory spleen, enlarged spleen may interfere with diagnosis of a neoplasm of the left upper quadrant and, in particular, may be confused with solid tumors of the pancreas or the left adrenal gland.[11]

Conclusion

The notches or clefts on the superior border of the adult spleen are remnants of the grooves that originally separated the fetal lobules. These clefts can be sharp and are occasionally as deep as 2–3 cm. They may be erroneously interpreted as splenic laceration in patients with abdominal trauma. Multilobulated spleen should be kept in mind during clinical evaluations like splenomegaly, splenic traumas, lymphadenopathy and also in cases where splenectomy is required due care is needed to remove all the splenic lobules, otherwise the residual lobules even though quite smaller may take the function there by nullifying the effect of splenectomy. Congenital anomalies of the spleen though rare need to be taken into account in the differential diagnosis of pathological disorders, including neoplasm, when these are situated in the left upper quadrant of the abdomen. In the present era of laparoscopic splenectomy, more accurate knowledge of splenic variations is of fundamental importance to improve diagnostic and therapeutic approaches.

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