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Primary Spelnic Hemangioma with Spenunculi at Greater Sac: A Rare Case Report

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Abstract: Primary tumors of the spleen are rare and with the exception of malignant lymphoma are seldom considered in the differential diagnosis of splenomegaly. Such tumors may be either cystic or solid, and the cystic variety, composed mostly of epidermoid cysts, has occurred with slightly greater frequency. The most common primary solid tumor is the hemangioma (hamartoma) and the majority of these have been described as incidental autopsy findings. Most splenic hemangiomas are discovered incidentally, and their clinical importance generally lies in differentiating them from other conditions. Occasionally, they may be associated with splenomegaly, abdominal pain, dyspnea, diarrhea or constipation, anemia, thrombocytopenia and with larger lesions which spontaneously rupture. Although hemangioma is a very common benign lesion in the body, a huge and mixed splenic trype hemangioma is very rare.here we report a case of 50 yr female presented with lump in abdomen turned out to be splenic hemangioma. Splenunculi are single or multiple accessory spleens that are found in approximately 10–30% of the population.so primary splenic hemangioma with occurance of speninculi is rare finding.
Keywords: Spelnic Hemangioma, Spenunculi

INTRODUCTION

Splenic hemangioma is a vascular malformation that represents the most common benign primary neoplasm of the spleen. Its prevalence at autopsy ranges from 0.03% to 14% and it is the hemangioma most commonly seen in adults in the mid-30to mid-50year old range [1].This lesion is even less common in infants and young children [2]. Most hemangiomas are small asymptomatic lesions that are found incidentally.The indications for surgical intervention and other therapy remain unclear. There is an increased risk of splenic injury and rupture, as well as the possibility of developing hypersplenism or Kasabach– Merritt syndrome, especially in larger lesions [3]. In addition there is the concern for malignancy, which prompts splenectomy in most cases [4].

CASE REPORT

A 51-year-old lady presented with a 1 month history of vague abdominal pain, with a one week history of increasing fatigue and dyspnea on exertion.He had 1 episode of sudden fainting 2 days back .She denied any history of vomiting,constipation,diarrhea,haematemesis,bleeding PR or use of non-steroidal anti-inflammatory agents (NSAIDs). Her past medical history was insignificant. Social history was negative for alcohol, tobacco, or illicit drug use. Her family history was unremarkable. On physical exam, the patient was severely anaemic. Her heart rate was 80/min and blood pressure 118/78.She never noticed any swelling or lump in the abdomen. Her abdomen was soft and non tender, and auscultation revealed normal bowel sounds. There was E/o intra abdominal firm palpable lump in left hypochondrium extending up to left iliac region (size - 15cm x 10cm x 3 cm), margins well defined ,smooth surface with upper border not palpable, mobile with respiration.Clinically diagnosis of splenomegaly was done. The rest of her exam was unremarkable. Laboratory evaluation revealed pancytopenia -HB being 4.8 with hematocrit of 21.7% (38.4-51.7 gm/dL), mean

corpuscular volume (MCV) of 86 (80-97 fl) with WBC count 3400/cu mm and platelets being 96000/cu mm. Other lab investigations including the remainder of liver enzymes, basic chemistries, and a thyroid panel were all within normal limits. Iron studies and a plasma iron 21 lg/dL (34–175 lg/dL), total iron binding capacity 207 lg/dL (250–400 lg/dL), transferrin saturation of 10% (15–50%). Indirect Coomb's test was negative.Her urine analysis and stool guiaic were negative for blood.Patient was given 4 units of PCV to built up his HB upto10 for surgery.

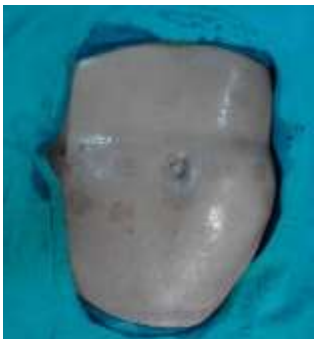
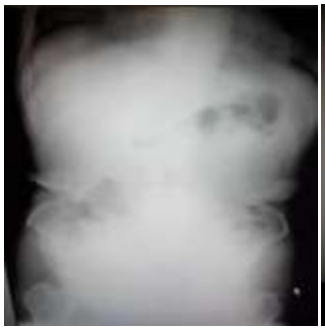


Fig. 1 : **Fig. 2**



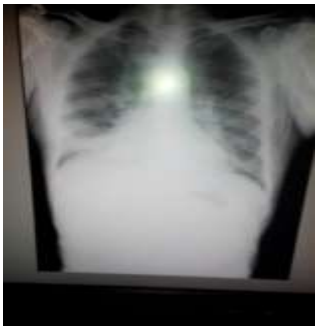
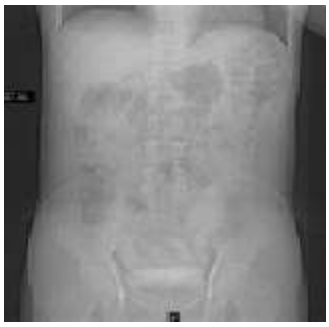


Fig. 3 **Fig. 4**

Her X ray abdomen erect and chest was normal (Fig. 3 & 4).USG abdomen and pelvis suggested E/o massive splenomegaly. No evidence of free fluid or obvious lymphadenopathy. CECT abdomen and pelvis reported demonstrated a splenic lesion measuring 20cm*15 cm (Fig. 6 & 7). This exerted a mass effect on the tail of the pancreas and the splenic hilar vessels. The splenic lesion was not amenable to percutaneous biopsy,making it difficult to differentiate between a benign and malignant process.



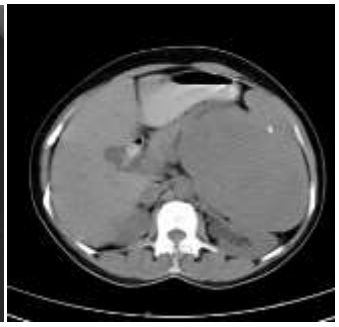


Fig. 5 **Fig. 6**

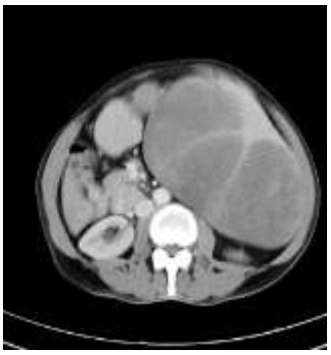


Fig. 7

Fig. 8

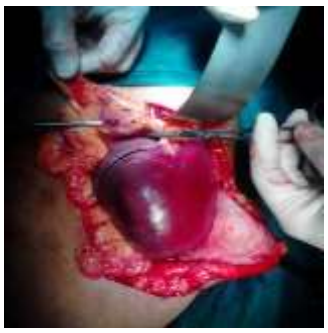


Fig. 9

Fig. 10

Fig. 11

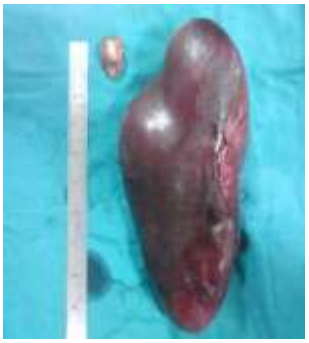


Fig. 12

Given the risk of lymphoma, which often is treated non surgically, a bone marrow biopsy was performed. Staining of her bone marrow aspirate revealed iron present in the normo cellular sample,also present was a para trabecular lymphoid aggregate suggestive, but not diagnostic, of splenic lymphoma. Therefore, it was decided to perform a splenectomy for diagnostic and possibly therapeutic purpose. Exploratory laparotomy was done with cheveron incision extending left subcoastal margins. Intraoperatively, the spleen was found to be markedly enlarged with the lower pole reaching below the umbilicus. There were thickened lateral attachments with dense adhesions to the splenic flexure of the colon and the tail of the pancreas. Mobilization of the spleen required extensive lysis of all attachments with care being taken to avoid damage to the tail of the pancreas. There was evidence 1 splenincul of size about 3cm *2cm*1cm at greater sac which was excised .There was minimal blood loss during the procedure.The specimen weighed 1700 gms. The spleen measured 25cm * 15 cm*10 cm.Post operative course was uneventful. Liquids and Oral feedings started on 2rd day. Sutures were removed on 10^{th} day and patient discharged. Histopathology of the resected specimen showed splenic parenchyma, which was significantly replaced by a lobulated proliferation of vascular structures. These structures were separated from the parenchyma by dense fibrosis. The proliferation was composed of ill defined vascular channels admixed with stromal cells,extravasated red cells, fibrin, and significant hemosiderin deposition. Finally diagnosis of splenic hemangioma was made. Histopathology of mass at greater sac was consistent with Splenunculi. Follow-up 5 weeks later revealed normalization of her labs with a hemoglobin of 12.4 g/dL (11.7–15.5 g/dL), hematocrit 43.3% (35–45%),

and an MCV of 88fL (80–100 fL). Post operative iron studies also normalized with a plasma iron of 90 lg/dL (35–175 lg/dL), total iron binding capacity 344 lg/dL(250–400 lg/dL), and transferrin saturation of 26%(15–50%).She remains symptom free with normal haemoglobin after 6 months of follow up.

DISCUSSION

Splenic hemangioma was first described by Virchow in 1863 [1]. Etiology of splenic hemangiomas is unknown. It has been suggested that they represent a congenital nevus that may or may not enlarge to become symptomatic [3, 5]. Most are found in patients in their third to sixth decades of life, although some pediatric literatures have been reported[6, 7].

Bostick surveyed 16 patients with splenic hemangiomas and a palpable abdominal mass and found that 62% had pain, 12% had anemia, 12% had ascites, and 18% had weight loss [5]. Thrombocytopenia has also been described [8]. Generalized splenomegaly may be present,hypersplenism is most common association. Splenic hemangioma may occur as part of generalized angiomatosis, as seen in Klippel-Trenaunay syndrome [9, 10]. There was no clinical or laboratory evidence associating either syndrome to our case.

There are some reports in the literature that describe the synchronous presence of liver and spleen hemangioma [11]. The presence of central hematomas in the giant hemangioma are prone for possible rupture. Serious complications of hemangiomas include rupture and malignant transformation. Spontaneous rupture of a giant hepatic hemangioma (diameter >4 cm) with hemoperitoneum occurs very rarely. Willcox et al. report a spontaneous rupture incidence of 25% [1]. It is reported that malignant transformation occur more frequently with large or multiple hemangiomas and most surgeons to favor splenectomy [1,12].

Sonography is known as a useful examination tool of soft tissue masses, including thosethat are suspected to be hemangiomas or vascularmalformations [13]. TheDoppler examination is crucial to distinguishlow- and high-flow vascular malformations [14]. As reported by Chatzoulis *et al*. [15] CT and MRI failed to achieve appreciable sensitivity and specificity to define the origin of the giant splenic hemangioma. Velkova and Neveda [16] report a combined sensitivity of 61.3% for ultrasound and CT scan in the recognition of liver and spleen hemangiomas.

The treatment of splenic hemangiomas usually involves splenectomy [17].Folkman demonstrated that hemangiomas secrete angiogenic agents (such as B-FGF)that can be detected in the urine, and are reduced with anti angiogenic therapy [18]. Other drugs successfully used have been the COX-2 inhibitors and Thalidomide [18, 19].

CONCLUSION

Primary tumors of the spleen are rare and with the exception of malignant lymphoma are seldom considered in the differential diagnosis of splenomegaly. Primary splenic hemangioma attending a large size requires definitive treatment. Other medical modalities like steroids, thalidomide or arterial embolisation is not that effective in case of massive splenomegaly due to large splenic hemangioma. Splenectomy is treatment of choice. Splenunculi can be an incidental finding and complete excision is of utmost importance.

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