Abstract: The most common primary tumours of the spleen are benign and originate from the vascular endothelium. Haemangioma is the most common primary benign neoplasm of spleen and usually represents as a small, localized tumour. Cavernous haemangioma is a rare disorder of the spleen with fewer than 100 cases reported in literature. This condition may occur together with haemangiomatosis of the liver or it may occur as a manifestation of systemic angiomatosis, or less commonly, is confined to the spleen. We present a patient with moderate splenomegaly caused by splenic haemangiomatosis. The patient had no clinical disorders resulting from the splenic vascular tumour.

Keyword: cavernous haemangioma of spleen, giant splenomegaly, haemangioma, partial splenectomy, splenic haemangioma

INTRODUCTION:
The most common primary tumours of the spleen are benign and originate from the vascular endothelium. Haemangioma is the most common primary benign neoplasm of spleen and usually represents as a small, localized tumour. Cavernous haemangioma is a rare disorder of the spleen with fewer than 100 cases reported in literature. This condition may occur together with haemangiomatosis of the liver or it may occur as a manifestation of systemic angiomatosis, or less commonly, is confined to the spleen. We present a patient with moderate splenomegaly caused by splenic haemangiomatosis. The patient had no clinical disorders resulting from the splenic vascular tumour.

CASE REPORT:
A 58 yr old male pt presented to the surgical opd with presenting complaints of occasional pain in the left upper quadrant of the abdomen for the past one year with progressively increasing pain which was dragging in nature. There was no h/o anaemia, weight loss, anorexia. The general examination was unremarkable. On p/a examination, the spleen was moderately enlarged, extending upto almost 5 cm from the costal margin which was firm in consistency. Patient was a diabetic. Otherwise the blood counts and serum chemistry was normal. Ultrasound examination of abdomen showed a significantly enlarged spleen including numerous, ill-defined, hyperechoic nodules and revealed a parenchymal heterogeneity and septation.

Abdominal CECT showed two hypodense lesion larger than 76*67 mm and the smaller one measuring 43*44mm which showed peripheral enhancement on contrast. A preoperative diagnosis of splenic haemangioma was made the patient was posted for elective spleenectomy for diagnostic and therapeutic purpose.

Patient was explored by a left subcostal incision under GA. Per operatively, there was a moderately enlarged spleen with two solid lesions of size 6 * 6 cms and 4 * 3 cms. Specimen was sent for histopathological examination. Post op period was uneventful.

HPE showed several thin walled blood vessels replacing most of the splenic tissue with adjacent splenic area showing congestion. Remnants of the red pulp were observed among the pathological vessels throughout the spleen.
Discussion:
Diffuse haemangiomatosis of the spleen is a rare benign vascular condition. Splenic haemangiomatosis may be asymptomatic or cause complications such as disturbances of blood coagulation, rupture of the spleen and portal hypertension. Small localized haemangiomas are common neoplasms of the spleen. Isolated diffuse splenic haemangiomatosis, however, is very rare. This lesion can be accompanied by severe hypersplenism and other complications. Clinically, the diagnosis is usually difficult to make, but computed tomography and ultrasound can be useful methods for evaluating this vascular disorder. In our case there was no radiologic or clinical evidence of other organ involvement and, therefore, the diagnosis of multiple splenic haemangiomatosis was made.

The differential diagnosis of diffuse splenic hemangiomatosis must take into account the other vascular tumors or tumor-like lesions of the spleen such as lymphangioma, littoral cell angiomia, hemangioendothelioma and primary angiosarcoma, peliosis of the spleen, and hamartoma. Lymphangioma and lymphangiomatosis are readily separated because they are often sub capsular and lymphatic channels contain eosinophilic, proteinaceous material without erythrocytes. Localized haemangiomas possessing a nodular pattern are easily eliminated.

US may show an inconsistent and nonspecific appearance of echogenicity and sharp margination sometimes with cystic regions. Unenhanced CT shows a low attenuation mass; but after injection of a contrast material, there is an increased attenuation of mass, which has been reported to be from periphery to the centre. MR imaging is more sensitive and specific than other imaging modalities in the diagnosis of splenic haemangioma. It can be used for imaging splenic lesions in which differential diagnosis is not reached by CT. They are typically hyperintense at T2-weighted MR imaging with a centripetal filling pattern after administration of gadopentetate dimeglumine. Hyperintense signal on T2-weighted images is due to long T2 relaxation times; this quality generally distinguishes it from solid neoplasms. However, regions of liquefied necrosis in solid tumours may also be hyperintense on T2-weighted images.

The usual reported treatment is splenectomy. Recently, there have been reports of benign lesions being treated with partial splenectomy after ligating the segmental supply of the involved half of spleen. The advantage of partial splenectomy is the preservation of its immunological function. Overwhelming postsplenectomy sepsis, although rare, can be a fatal untoward outcome of splenectomy. When the spleen is absent, its immunological functions are generally compensated for, but the phagocytic clearance of bacteria, especially the encapsulated pneumococci and Haemophilus influenzae, are reduced, particularly if the host has a deficient concentration of opsonising antibodies. Treatment options for large lesions usually consist of splenectomy, embolization, or both. Antiangiogenic treatment has not been reported previously as an effective alternative for this type of lesion. There is a case report of successfully using glucocorticoids in an infant with a large hemangioma of the spleen. Laparoscopic splenectomy appears to be a safe and effective procedure, in appropriately experienced hands, for patients with splenomegaly, given the spleen’s fragile anatomy and its relationship to other abdominal viscera.

CONCLUSION
Although unusual, haemangioma is nonetheless the most common primary splenic neoplasm. Haemangiomas are usually less than 2 cm in size. It is extremely rare to have such a massive solitary lesion. Most splenic haemangiomas are discovered incidentally, and their clinical importance lies in differentiating them from other space-occupying lesions of the spleen (both solid and cystic), particularly from metastases. Preoperative diagnostic investigations are often inconclusive and rarely distinguish between haemangiomas and metastasis. MR imaging is the most reliable imaging method. When possible, partial splenectomy is the treatment of choice for isolated splenic lesions.

References:
1) Maingot's Abdominal Operations. 11th ed. New York: McGraw Hill, 2007: 1075-96
b. Antiangiogenic therapy for a large splenic hemangioma. Islam S, Newman EA, Strouse PJ, Geiger JD. Department of Surgery, Division of Pediatric Surgery, University of Mississippi Medical Center.

c. Costello P, Kane RA, Oster J, Clouse ME. Focal splenic disease demonstrated by ultrasound and computed tomography. J Can Assoc Radiol 1985; 36:22-6.
d. Giant cavernous haemangioma of the spleen presenting as massive splenomegaly and treated by partial splenectomy. Ghuliani D, Agarwal S, Thomas S, Pathania Op.
e. Diffuse hemangiomatosis of the spleen – case report* Nuket Eliyatkin, Sibel Demir Kececi, Arsenal Sezgin, Hakan Postaci, Tahsin Tekeli , AliGalip
f. Burke JS. The Spleen. Sternbergs Diagnostic Surgical Pathology fourth edition. Philadelphia Lippincott Williams & Wilkins 2004: 849.

g. Elective laparoscopic splenectomy for giant hemangioma: a case report Kosmidis C, Efthimiadis C, Anthimidis G, Grigoriou M, Vasiliadou K, Sfikakis P, Tziris N, Fahantidis E

h. Isolated diffuse hemangiomatosis of the spleen: case report and review of literature. Steininger H, Pfote D, Marquardt L, Sauer H, Markwat R.