CASE REPORT

CAVERNOUS HEMANGIOMA SPLEEN PRESENTING AS A CYSTIC MASS: A CASE REPORT

Muhammad Mumtaz Khan, Sadaf Alam, Fozia Rauf, Sajjad Ahmad

Department of Pathology, Peshawar Medical College, Peshawar, Pakistan, affiliated with Riphah International University, Islamabad, Pakistan.

ABSTRACT

Cavernous hemangioma is a rare disorder of spleen with fewer than 100 cases reported. We describe a case of cavernous hemangioma spleen in a 40 years old woman putting emphasis on its presentation. She presented with pain left upper quadrant and fever. Her spleen was palpable on abdominal examination. The lesion was picked on ultrasound abdomen which reported multiple cysts of variable sizes, more in the periphery, the largest measuring 2.4 x 1.4 cm. CT abdomen with contrast showed a normal sized spleen having multiple hypodense lesions more at the periphery. Splenectomy was carried out. Histopathological examination revealed diffuse involvement of the spleen by cystically dilated vascular channels most of which contained RBCs in their lumina. Therefore, a diagnosis of cavernous hemangioma spleen was made which was confirmed by a positive CD-34 immunohistochemical stain.

KEY WORDS: Cavernous hemangioma; Spleen; Cystic lesions.

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INTRODUCTION

Splenic hemangiomas (SH) range in size from capillary to cavernous and although rare, they are the most common benign primary tumor of the spleen. Fewer than 100 cases of cavernous hemangioma spleen have been reported in literature.[1] By definition cavernous hemangioma (CH) is an unencapsulated mass of dilated vascular channels lined by endothelial cells and filled with RBCs.[2]

Hemangiomas of the spleen are incidentally discovered because they are slow growing and are usually asymptomatic.[3] Patients with huge CHs may develop symptoms due to enlarged spleen. Large lesions with significant splenomegaly may cause fullness and left upper quadrant discomfort.[2]

Ultrasonographically capillary hemangioma is hyperechoic whereas CH is either a mixed echogenic or hypoechoic structure and may have focal calcifications or appear as a cystic mass.[4] On CT scan CHs show both cystic and solid components. The later appears as isoattenuating or hypoattenuating in relation to normal splenic parenchyma with solid tissue enhancement only.[5]

The differential diagnosis for a solid mass of the spleen include angiosarcoma, lymphoma, metastases and hamartoma. The differential diagnosis for a cystic lesion of the spleen includes lymphangioma, hematoma, abscess and parasitic cyst.[6]

Complications of CH include hypersplenism, malignancy and spontaneous rupture which is the most common complication and has been reported in 25% of cases. Patients with large hemangiomas may present with Kasabach-Merritt Syndrome which is a combination of thrombocytopenia, anemia and coagulopathy. Splenectomy is the choice of treatment for patients having symptoms.[5]

The aim of the article was to highlight the fact that pain and discomfort in the left hypochondrium may be due to splenic lesion most likely a hemangioma even though the spleen may not be enlarged.

CASE REPORT

A 40 years old female reported to surgical department with a history of pain left hypochondrium for the last four months. The pain was burning in type with gradual onset and occasional radiation to epigastric region. She also complained of abdominal fullness. This was accompanied by low grade fever and nausea. She reported having similar episodes

Corresponding Author:
Dr. Muhammad Mumtaz Khan
Professor
Department of Pathology, Peshawar Medical College
Riphah International University
Islamabad, Pakistan
E-mail: mmumtazkhan@gmail.com
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previously which resolved with self-medication (antacids). There was no history of abdominal trauma or infection.

All the vital signs were stable on general physical examination with no icterus or enlarged lymph nodes. Per abdomen there was tenderness in the left hypochondrium. The spleen was palpable. Rest of the systemic examination was unremarkable. The results of her lab investigations were as follows: Hemoglobin: 11.8 g/dL, WBC: 7.4x 10^9/uL, DLC: Neutrophils 60%, Lymphocytes 36%, Eosinophils 04%, Platelet count: 290,000/uL, Serum creatinine: 0.6 mg/dl, Blood urea: 23 mg/dL.

The patient was advised an abdomino-pelvic ultrasound to evaluate the mass. It reported a normal sized spleen having multiple variable sized cystic spaces more at the periphery but also involving the spleen diffusely, the largest cyst measuring 2.4 x 1.4 cm2. Rest of the viscera were unremarkable. This was followed by a CT scan abdomen with oral and I/V contrast. It revealed a normal sized spleen having multiple hypodense lesions with bulging contours (Fig. 1).

The diagnosis of a cystic mass of spleen was made with a differential of multiple angiomyolipoma, parasitic cysts, lymphangioma and hemangioma. Splenectomy was performed by conventional method by giving a left sub-costal incision along with resection of a large omental flap about 20 cm in size and was sent for histopathological examination.

Gross examination revealed a normal sized spleen measuring 14 x 9 x 4 cm3, weighing 230 g. Outer surface was grayish brown with protruding nodules measuring from 1 to 2.4 cm in diameter (Fig. 2).

The consistency was soft to firm and the cut surface showed numerous sub capsular, variable sized cysts as well as small cystic spaces scattered throughout and interspersed by firm grayish brown areas. The largest cyst measured 3 x 2.5 cm^2 filled with thick gelatinous material and surrounded by a thin rim of intervening normal looking splenic parenchyma (Fig. 3).

Microscopic examination revealed multiple dilated vascular channels of variable sizes, lined by a single layer of flattened endothelial cells most of which contained RBCs in their lumina (Fig. 4).

The intervening stroma showed sprinkling with lymphocytes and fibrosis. Blood vessels with partially organized clot and some with calcification were also present. The white pulp was normal in appearance and the red pulp showed congested sinuses. Immunohistochemical (IHC) stain CD-34 showed a strong positivity for the lining epithelium of the large dilated vascular channels as well as smaller blood vessels and capillaries scattered throughout the splenic parenchyma confirming the diagnosis of CH spleen (Fig. 5).

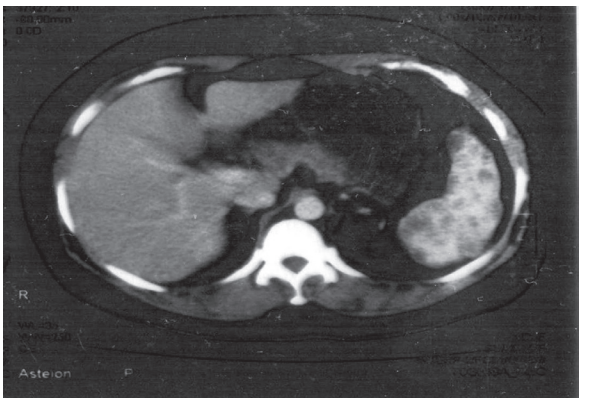

Figure 1: Spleen having multiple hypodense lesions

Figure 2: Spleen with bulging nodules

Figure 3: Spleen cut surface. Various size multiple cysts scattered throughout

Figure 4: Cystically dilated vascular channels with RBCs in their lumina. H&E x 20

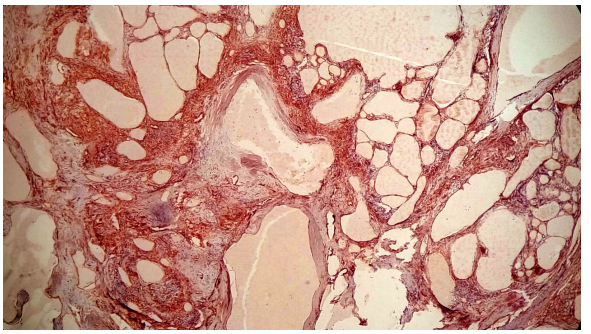

Figure 5: IHC stain CD-34 positivity of lining epithelium of cavernous vascular channels and small blood vessels in the splenic parenchyma

DISCUSSION

Regarding pathogenesis of hemangiomas two theories dominate. Firstly, it is stated that disrupted placenta embedded in fetal soft tissues during gestation or birth gives rise to hemangioma endothelial cells. The evidence being that immunohistochemical markers of hemangiomas coincide with those in placental tissue.[7] The second theory states that in the circulation of patients with hemangiomas endothelial progenitor stem cells have been found.[8]

Due to its slow growth, SHs are usually discovered in adult life and the spleen may or may not be enlarged. Willcox TM et al., reviewed an 8-year experience of SHs with 32 patients identified during this period. Their age ranged from 23 – 94 years with an average of 63 years, 17 being females and 15 males. Only six patients had symptoms related to SH. A large majority of patients (80%) were asymptomatic, and their SH was discovered during investigations for other disorders. Only four patients (12.5%) had a mass or palpable spleen. The size of SHs ranged from 0.3 to 7 cm in diameter. In 11 patients a diagnosis of SH was based on the computed tomography or ultrasound findings of a splenic mass with a size less than or equal to 4 cm.[9]

Our patient was a 40-year-old female who presented with gastric complaints and discomfort in the left hypochondrium. The cystic mass was easily picked on both ultrasound and CT scan, the largest cyst measuring 2.4 x 1.4 cm^2. Although her spleen was palpable but was found to be of normal size on imaging.[10]

In our case the spleen was misshapen grossly due to bosselated surface caused by bulging nodules and the cut surface was studded with numerous cysts of various sizes scattered throughout. Although usually the SHs are small measuring less than 4 cm diameter but literature has mentioned giant SHs of 12.2 x10.6 cm^2 and 11.8x9.7x7.8 cm^3 dimensions. In other instances multiple cystic spaces of various sizes diffusely involved the spleen[11] as in our case. There has also been a case report in which whole of the parenchyma of a grossly enlarged spleen was occupied by CH.[12]

Cystic hemangiomas often develop curvilinear or eggshell calcifications. Solid lesions have more commonly mottled central calcification, whereas long-standing thrombosis show coarse calcification in areas of necrosis.[5] In our case both curvilinear and coarse calcifications were seen in a thrombosed cystically dilated vascular channel suggesting long-standing duration of the lesion (Fig. 6).

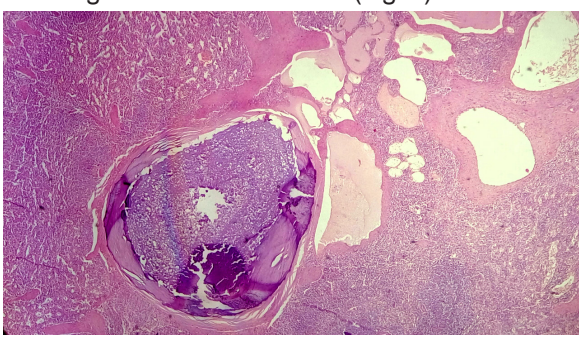

Figure 6: Curvilinear and coarse calcifications in a thrombosed vessel

Lymphangioma was the main differential considerations in our case which was ruled out by a positive CD-34 expression in the endothelial lining of vascular channels (Fig. 5).

It is essential that these patients are timely diagnosed and treated by splenectomy because 25% of hemangiomas more than 4 cm in diameter tend to rupture spontaneously.[13]

CONCLUSION

The possibility of SH must be kept in mind in patients coming with complaints of pain and discomfort in the left hypochondrium. Timely splenectomy in these patients may avert the danger of spontaneous rupture.

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CONFLICT OF INTEREST
Authors declare no conflict of interest.
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AUTHORS' CONTRIBUTION

Conception and Design:	MMK,
Data collection, analysis & interpretation:	SA, FR, SA
Manuscript writing:	MMK,