Tumours of the spleen are infrequent entities with metastasis outnumbering the primary neoplasms. In the latter category, various benign and malignant vascular tumours constitute the majority, benign being the commoner. Hamartoma is a well-described tumour like benign lesion, which mostly remains clinically asymptomatic and is incidentally discovered during abdominal imaging. Infrequent presentation with haematological disturbance or splenic rupture have also been reported. Its morphological identification and differentiation from other splenic vascular neoplasms is greatly facilitated by immunocytochemistry.

Splenic Hamartoma is a rare lesion, which is usually an incidental autopsy/radioimaging finding. First described by Rokitanski in 1861, this entity remains the least important of splenic angiomatous lesions. Majority remain clinically asymptomatic. However, several cases presenting with hematological disturbances or even spontaneous rupture have been reported. Till to date nearly 150 cases have been described out of which only 20 were encountered in paediatric population. We report a case of splenic hamartoma in a 5 year-old child, where preoperative diagnosis was wrongly directed towards supra renal neoplasm resulting in laparotomy and splenectomy.

Case Report

A 5 year-old boy presented with a 2 months history of abdominal pain and 2 years history of vomiting. His CBC showed. WBC 9.2 x 10^9, Hemoglobin 11.6 gm/dl, platelets 327 x 10^9. PT and APTT were normal. Serum urea and creatinine values were within normal range. Abdominal ultrasound showed a well-defined hypoechoic area within spleen, 3.4-cm. sized. CT revealed 3.3 x 3.2-cm. soft tissue lesion occupying left upper abdomen medial to spleen and posterior to left kidney with a local mass effect on the spleen (Figure 1). Likely diagnosis was suprarenal involvement with neuroblastoma or pheochromocytoma. Further investigations revealed, Homovanillic acid, 9.33/umol/24 hrs (normal=4.4-l37/umol/24 hrs), vanillyl mandelic acid, 3/umol/24 hrs (Normal upto 35/umol/24 hrs), LDH, 510U/L. Patient was admitted for suprarenal exploration. On laparotomy a mass was detected in the spleen whereas suprarensals were unremarkable. Splenectomy was carried out for diagnostic purposes.

Pathological Findings

On gross examination spleen weighed 250 gms. with a normal capsule and hilum. On sectioning, a wellcircumscribed, tan colored, 3.5 cm. sized bulding mass was noted (Figure 2). A similar looking 1.2 cm. sized nodule was present 3.0 cm. away from the above-mentioned mass. On histological examination of both these nodules a non-encapsulated lesion of red pulp was defined surrounded by normal compressed splenic parenchyma (Figure 3). The red pulp cords were expanded & disorganized and splenic sinusoids were lined by flat to plump endothelial like cells (Figure 4). There was no representation of white pulp tissue within the lesion. However, the periphery of the lesion revealed encroachment at some foci within the normal splenic tissue. Reticulin stain outlined the sinusoids exaggerating the red pulp architecture. A panel of Immunocytochemical stains were employed. The sinusoidal lining cells stained with the vascular markers, CD34 and factor VIII Rag. In addition, CD8 was also positive in the lining cells but negative in the bulk of the lesion, which comprised of red pulp cords (Figure 5). Scattered lymphocytes in the main lesion were positive however with CD8. CD68 and CD21 were negative in the vascular lining cells (Figure 6).
Spleen is an uncommon site for primary tumours and in this context vascular tumors constitute the most common category. Both benign and malignant vascular tumors have been encountered, benign being the commoner of the two categories. Depending on the cell of origin, the benign vascular lesions are either hemangioma, littoral cell angioma or a red pulp cord lesion, the hamartoma. Arber et al conducted an elaborate analysis of splenic vascular lesions employing immunophenotypic and virological studies and reported 9.9% incidence of hamartoma in their series. Till today 150 cases have been reported in the literature. Its frequent asymptomatic presentation and lack of scrupulous examination of spleen at autopsy, probably accounts for its low quoted incidence. Majority of the cases are reported in adults whereas only 20 cases have been reported in paediatric population according to the English literature. One recent case report of spontaneous rupture of splenic hamartoma in a 5 month old child raises this count to 21. Advances in imaging techniques have resulted in increased frequency of its diagnosis even in asymptomatic subjects. On ultrasound examination our case presented as a well-defined hypoechoic area within the spleen contrary to the studies reporting the lesion as an echogenic well circumscribed mass. Varying results have been reported with CT scan and MRI. In homogeneous low density area on CT with I/V contrast with or without calcification have been reported. With MRI these lesions are visualized as isointense on T1-weighted and hyperintense on T2-weighted images. These studies also reported a delayed enhancement during arterial phase of dynamic MRI becoming isointense and prolonged on subsequent imaging. The authors regard prolonged enhancement as a characteristic feature for hamartomas and attribute the low signal intensity to lack of edema, necrosis and presence of fibrosis in the hamartoma. Difficulty of detecting these lesions due to Isodense nature compared to normal spleen has also been reported. CT and MRI findings in our case were misleading, however and were suggestive of a suprarenal pathology.

The histological diagnosis of our case was greatly facilitated by immunocytochemistry. Both benign and malignant neoplasms have to be considered in the differential diagnosis of splenic hamartoma. The benign categories of significance include haemangiomas, hamartoma and the recently described lesion of littoral cell angioma. The haemangiomas are distinct entities, which can occur in cavernous form with dilated endothelial lined spaces posing no diagnostic problem. Capillary type haemangiomas on the contrary may reveal overlapping features with hamartomas. Immunocytochemistry can play a major role here. The origin of haemangioma is believed to be ordinary vascular endothelium, factor VIII RAg positive. Whereas in hamartoma the lining characteristically shows CD8 positivity. These immunocytochemical differences suggest presence of different type of endothelium in these two tumours. Buckley et al. described for the first time monoclonal antibodies to T suppresser/cytotoxic lymphocytes (CD8) to react distinctly with the red pulp sinusoidal lining cells. The reaction was negative with pan T cell antigen or CD4 antigen. In our case CD8 positive T lymphocytes were scattered in large numbers in the splenic hamartoma and positive staining was also observed in the sinusoidal lining cells. A functional relationship between splenic sinusoidal lining cells and T lymphocytes have been postulated on the basis of sharing the common antigen. CD8 staining along with other immunocytochemical stains help to differentiate hamartoma from another well described benign vascular lesion, the Littoral cell angioma. Described for the first time in 1991 by Falk et al. as a novel type of vascular lesion of spleen, it has a characteristic and distinct histological appearance which most of the times helps in distinguishing it from hamartoma. It exhibits anastomosing vascular channels resembling sinuses with irregular lumina, often with papillary projections. The lining cells are characteristically tall endothelial cells, which slough into vascular lumina and exhibit haemophagocytosis. Immunocytochemistry reveals factor VIII RAg as well as CD68 positivity reflecting on dual differentiation potential of the cells lining the splenic sinuses i.e. endothelial / histiocytic differentiation. CD21 positivity is uniquely reported in the lining cells of LCA. In all the reported series Immunoreactivity of LCA is universally described as CD21+ve, CD 68+ve, CD8-ve. In some series the cases of diffuse haemangiomas and focal haemangiomas of spleen were also CD68+ve. However, this overlapping with LCA creates no diagnostic problem as CD 21 is consistently absent in the former lesions. Our case revealed CD21-ve, CD68-ve and CD8+ve pattern of immunocytochemical staining. Thus differentiation from Littoral cell angioma and haemangioma was straightforward.

The main clinically important differential of focal vascular lesions of spleen is from malignant tumours as haemangioendothelioma, angiosarcoma and even the rare entity as Littoral cell angiosarcoma. However these did not pose much diagnostic challenge in our case. Primary malignant vascular tumours are rare entities in spleen, angiosarcoma being the commonest with nearly 70 cases on record. Falk et al. in their series of 40 cases report a variable morphology; malignant vasoformative pattern along with solid sarcomatous pattern being present in varying proportions. The typical feature of capillary elongated slits or cavernous channels with atypical endothelial lining was not encountered in our case. Some studies
describe the presence of bland looking cavernous spaces or even pseudosinusoidal structures, which can simulate normal splenic sinuses. But despite these deceptive areas, cytological atypia definitely exists in all such cases.4,10

Symptomatic presentation of hamartoma is infrequent but has been well recorded. Till 1998 only 18 well-documented cases of symptomatic hamartomas were encountered and only 5 were reported in children.1 The symptomatic cases usually present with haematological problems as anaemia, thrombocytopenia, splenomegaly and pancytopenia.2 In these patients splenectomy alleviates the features pointing to a pathogenetic role. Few cases of spontaneous rupture are reported as the first manifestation, mostly in adults but occasionally in children as well.1,3 The presentation with vague abdominal symptoms in our case is unrelated to the primary splenic lesion and the case remains to be an incidental discovery in routine imaging abdominal evaluation. Since there is considerable overlap in the imaging pattern of various vascular lesions of spleen, the morphological evaluation remains the only definite mode of diagnosing these lesions. Thus in symptomatic cases splenectomy is the operation of choice which has been reported to alleviate the haematological symptoms as well as to serve the purpose of histological diagnosis.

The present report highlights the importance of including this rare benign lesion in the differential diagnosis of any radiologically detected focal splenic mass.

References


