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Detection of an asymptomatic non-palpable non-lymphomatous CD8$^+$ splenic hamartoma

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Abstract
Present case report relates to the incidental finding of a splenic hamartoma during a medical check-up. Later, splenectomy was done. Tumor measured 3.5×3×3 cms. Microscopic examination of tumor showed unencapsulated disorganized tortuous vascular spaces of variable width, lined by plump endothelial cells. Immunohistochemical (IHC) examination of lining endothelial cells with anti-CD8$^+$ antibody revealed a positive reaction. Anti-CD8$^+$ positivity suggested the tumor to be a splenic hamartoma. It is a rare benign tumor. Present tumor was arising from red pulp (type II splenic hamartoma).
Keywords: *Benign, embryonic, immunohistochemistry, splenectomy, tumor malformation, vascular.*

Case Report

A 20 year old man was found to have a round hypoechoic lesion near the splenic hilum on ultrasonographic (USG) examination. Later, CT examination with contrast revealed a well circumscribed round tumor. The lesion was detected incidentally during a medical check-up. Preoperatively, CBC, bleeding time and coagulation time were done. Results were within normal range. The lesion was provisionally diagnosed as benign neoplasm of spleen. Subsequently, splenectomy was done. The resected spleen weighed 225 gm which appeared mildly enlarged (normal weight of spleen in Indian adult male is 90 $\pm$ 60 gm). Outer surface of spleen was normal. Serial cut sections showed a bulging homogenous grey tan nodule. Nodule measured 3.5×3×3 cm. Microscopically, nodule showed unencapsulated disorganized tortuous slit-like vascular channels of variable caliber, lined by plump endothelial cells (figure 1a). At few places, focal sclerosis was seen. It suggested its origin from red pulp (type II). Lymphoid follicles were not seen in tumor tissue. Compressed vascular channels were surrounded by loose aggregates of lymphoid cells. The adjoining splenic parenchyma showed normal red and white pulps traversed by fibrous trabeculae. Immunohistochemical (IHC)

examination of tumor tissue was done. Lining endothelial cells of tumor showed positive reactions with anti-$CD8^+$ (Fig. 1b), anti-$CD31^+$ (Fig. 1c), anti-$CD34^+$ (Fig. 1d) and anti-factor VIII antibodies (Fig.1e). Both endothelial cells and suppressor/cytotoxic T-cells (CTL) are $CD8^+$; $CD8^+$ antigen positivity of present tumor ruled out the possibility of most of other vascular neoplasms, e.g. hemangioma, littoral cell angioma, hemangioendothelioma, angiosarcoma, lymphangioma, angiomatoid transformation and metastatic tumors. Triple positivity ($CD8^+$,$CD31^+$ and$CD34^+$) of tumor cells was seen in present case as well as in 2 earlier reports[1,2]. Anti-$CD68^+$ antibody gave a diffuse positive reaction with scattered stromal macrophages and focal endothelial lining cells (figure 1f). Anti-$CD21^+$ antibody gave a negative result (figure 1g), Anti-Vimentin antibody gave a positive reaction with endothelial cells (figure 1h). Similar vimentin positivity has been reported earlier in this tumor. Present patient was finally diagnosed as a case of type II splenic hamartoma on the basis of histopathological and IHC findings. The patient could not be followed further.

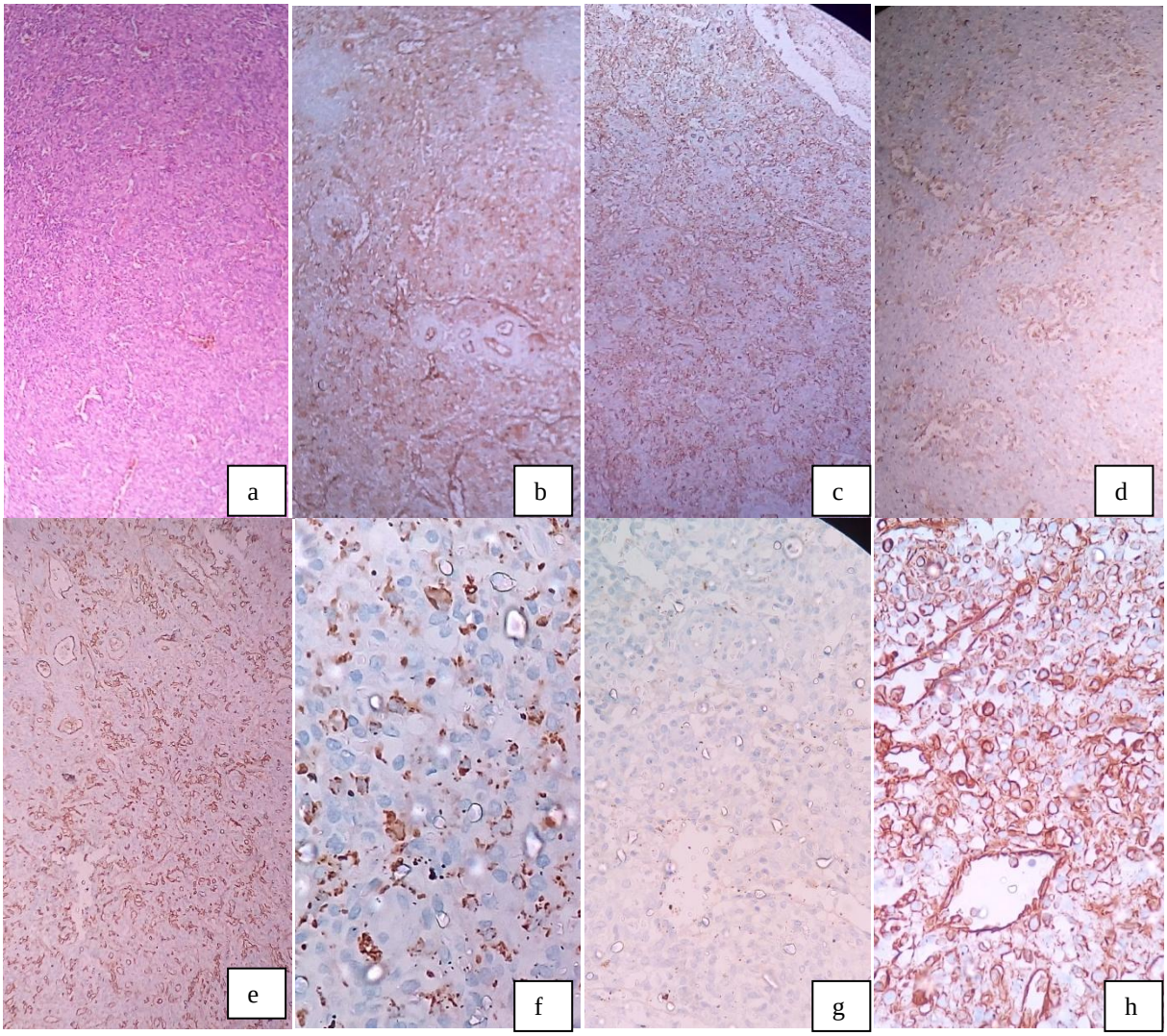

Figure 1 [a] shows tortuous disorganized vascular channels of variable width (HE × 100). [b]shows $CD8^+$ antigen positivity of endothelium (× 100). [c]shows $CD31^+$antigen positivity of endothelial cells (× 100). [d]shows positivity of tumor cells with anti-$CD34^+$antibody (× 1000). [e]shows reaction with factor VIII antibody (× 100). [f] shows staining of macrophage cells and focal endothelial cells with anti-68^+antibody (× 450). [g] shows negative result with anti-$CD21^+$antibody (× 100). [h] shows positivity of tumor cells for anti-vimentin antibody (× 450).Arrowheads indicate endothelium on macrophage cells.

Discussion

Current case was not associated with hematological dysfunctions while several of earlier cases developed anemia and/or thrombocytopenia or pancytopenia. Most of authors consider it to be a malformation. However, a few authors consider it to be a neoplasm of red pulp or a post-traumatic reactive lesion[3,4].

Most important feature of current case was the detection of a tumor consisting of disorganized proliferation of mature vascular channels of spleen. In addition, IHC examination of tumor revealed $CD8^+$antigen positivity of lining cells of vascular spaces. Similar $CD8^+$positivity has been described in several earlier studies[1,2,5-7]. Dual positivity of lining cells for $CD8^+$(a cytotoxic T-cell marker) as well as for VIII-related endothelial cell marker suggested a diagnosis of splenic hamartoma[8]. $CD8^+$ antigen positivity of lining endothelial cells ruled out the possibility of other vascular neoplasms. Another interesting feature of current case was the detection of $CD31^+$ and $CD34^+$ antigens in lining endothelial cells of tumor tissue. Similar finding has been reported earlier[2]. CD31 is a platelet endothelial cell adhesion molecule (PECAM-2); it plays a key role in removal of aged neutrophils. CD34 is a lineage marker of hematopoietic stem cells and may be expressed on endothelium. $CD68^+$ antigen positivity suggested staining for scattered macrophages and focal endothelial lining cells. Moreover, Vimentin antigen positivity indicated mesenchymal character of current tumor tissue.

Present case was asymptomatic. Similar asymptomatic presentation has been reported earlier[1,2,5-7]. Rarely, larger splenic hamartomas may be associated with various features, e.g. fever, thrombocytopenia and abdominal pain[7-9]. Thrombocytopenia may be due to anti-platelet antibody formation[8] and may not be related with the lesion. In a previous study[1], both anti-nuclear and anti-EBV antibodies were also detected. Later observation suggested possible role of EBV infection in pathogenesis of symptomatic splenic hamartoma. The previous patient responded to intravenous gamma-globulin therapy with a rise in platelet count[8].

Splenic hamartoma was first described by Rokitanski in 1861. Since then >150 cases have been reported. Its frequency in autopsy series is reported to vary from 0.024% to 0.13%. Clinically, it may be associated with hamartomas in other organs. In addition, its association with other neoplasms has been reported[10,11]. Present case was different from previously reported cases because it occurred in a young man, aged 20 years while most of earlier cases occurred in females in the age group of 21 to 60 years[1,2,9]. It may be incidently diagnosed due to compression of surrounding structures[12]. Rarely, asymptomatic multinodular splenic hamartoma has been described in a child with sickle cell disease[13].

Conclusion

Splenic hamartoma is a benign vascular $CD8^+$ endothelial proliferative embryonic tumor. Generally, it is asymptomatic. However, larger tumors may be associated with cytopenias and other systemic clinico-hematological features. Total or partial splenectomy appeared to reverse these features.

Financial and other Competing Interests: None

References

1. Lee H, Maeda K. Hamartoma of the spleen. Arch Pathol Lab Med 2009; 133:147-151.
2. Sim J, Ahn HI, Han H, Jun YJ, Rehman A, Jang SM, Jang K. Splenic hamartoma : A case report and review of the literature. World J Clinical cases 2013, 1(7):217-219.
3. Silverman ML, Livolsi VA. Splenic hamartoma. Amer J Clin Pathol 1978; 70 : 224-229.
4. Sankar S, Thanka J, Jagdishchandrabose S, Rajendran S. Splenic hamartoma : a rare vascular space occupying lesion of the spleen. Indian J PatholMicrobiol 2011; 54: 223-225.

5. Zhang L, Tou J, Wang X, GuW, Ma X, Qin Q. Splenic hamartoma in two children. World J Surgical Oncology 2014; 12:180-184.
6. Vlachou P, Fagkrezos D, Tzivelopoulou, Kyriakopoulou G, Maniatis P, Triantopoulou C, Papailiou J. A rare case of a splenic hamartoma in a patient with a huge palpable abdominal mass : a case report. J of medical case reports 2015; 9:4.
7. Basso SMM, Sulfaro S, Marzano B, Fanti G, Chiara GB, Lumachi F, Incidentally discovered asymptomatic splenic hamartoma with rapidly expansilegrowth : A case report. In Vivo 2012; 26 : 1049-1052.
8. Hayes TC, Britton HA, Mewborne EB, Troyer DA, Saldivar VA, Ratner IA. Symptomatic splenic hamartoma : Case report and literature review. Pediatrics 1998; 101 (5) : 1-6.
9. Eker T, Kocaay AF, Sevim Y, Cakmak A. Splenic hamartoma is a rare cause of abdominal pain : case report and literature review. Ulus Cerrahi Derg 2015, DOI : 10.5152/UCD. 2015.3048.
10. Darden JW, Teeslink R, Parish A. Hamartoma of the spleen : a manifestation of tuberous sclerosis. Ann Surg 1975; 41 : 564-566.
11. Huff DS, Lischner HW, Go HC, DeLeon GA. Unusual tumors in two boys with Wiskott-Aldrich-like syndrome. Lab Invest. 1979; 40 : 305-306.
12. Tavangar SM, Abdollahi A. Splenic hamartoma: Immunohistochemical profile. Acta Medica Iranica 2017; 55(1) : 77-78.
13. Elenga N, Labbe S, Leduc N, Sika A, Cuadro E, Long L, Njuieyon F, Kom-Tehameni R, Basset T. Asymptomatic multinodular splenoma (splenic hamart-oma) in a child with sickle cell anemia. Int Med Case Rep 2017; 10:233-236.