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General Medicine

SANT(SCLEROSING ANGIOMATOID NODULAR TRANSFORMATION OF SPLEEN)-AN UNUSUAL SPLENIC TUMOR

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ABSTRACT Splenic tumors are uncommon entities. Lymphomas are the commonest splenic tumors. Vascular tumors are rarely seen in the spleen. We report a case of sclerosing angiomatoid nodular transformation spleen that occurred in a patient with acute severe viral hepatitis B infection probably due to reactive process and review the literature pertaining to this entity.

KEYWORDS : SANT, Hepatitis, Splenic tumors

INTRODUCTION

Sclerosing angiomatoid nodular transformation of spleen (SANT) is a rare benign tumor of the spleen. Less than 150 cases have been reported worldwide. It was first defined by Martel et al in 2004.[1] It is more predominant in females and presents around the third to fifth decade of life. It is usually found incidentally. Although it has specific radiological findings it is difficult to differentiate from other tumors of the spleen. Its etiology is unclear but many theories have been proposed. Three common theories are (A)abnormal inflammatory response [2,3] (B)autoimmune/dysimmune disorders (IgG4 related disease) (C)perturbations in red pulp blood flow. [1,4] It histologically comprises of angiomatoid nodules with interspersing fibrous stroma and three types of blood vessels capillaries, sinusoids, small vessels with specific immunophenotype.

CASE PRESENTATION

A 41-year-old male with no known co-morbidities or allergies presented to ER with generalized itching for 2 days. His clinical examination findings were unremarkable except for scratch marks. Basic investigation revealed elevated liver transaminases and normal bilirubin. Over the next week he developed worsening itching and yellowish discoloration of eyes. Evaluation revealed hepatitis B with high HBV DNA titre of 26638 IU/ml. Bilirubin increased from 0.8 mg/dl to 32 mg/dl over a period of one month. His first ultrasound abdomen was found to be normal. He developed progressively increasing jaundice, persisting pruritis, vomiting, and decreased appetite and he developed coagulopathy with INR of 1.8. Due to the possibility of fulminant hepatic failure, he was started on Tenofovir. His liver function was monitored and he was discharged after bilirubin levels reduced.

A week later he returned with high-grade fever with chills and rigors and with no localizing symptoms and signs. His complete blood counts were within normal limits. An ultrasound abdomen done showed a well-defined heterogenous lesion of 5×4.1 cm size in the upper pole of the spleen.

With splenic abscess as a provisional diagnosis, he was treated with antibiotics. Aspiration was deferred in view of coagulopathy. Serial imaging was done to monitor the reduction in size.

INVESTIGATION

As lesion remained persistent a CECT of the abdomen was done which showed hypodense lesion with peripheral arterial enhancement and centripetal filling in delayed images in the upper pole of spleen a differential of a primary vascular tumor was considered (Figure 1).

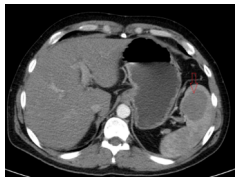

Figure 1: axial section of CT showing a well-defined hypodense lesion in the inferior pole of the spleen with peripheral arterial enhancement.

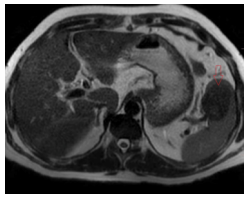

Figure 2: T2 sequence of magnetic resonance imaging showing hypointense lesion in lower pole of the spleen.

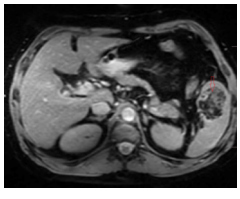

Figure 3: Gradient echo sequence of *magnetic resonance* sequence showing multiple areas of blooming within the lesion.

MRI was done and ruled out hemangioma and biopsy was done under frozen plasma cover (Figure 2, Figure 3).Biopsy showed a vascular lesion composed of disorganized blood vessels arranged as nodules which stained positive for CD 34 and Smooth muscle actin with Anaplastic lymphoma kinase being negative suggestive of Sclerosing angiomatoid nodular transformation of the spleen (SANT).

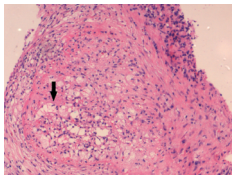

Figure 4: Histopathology showing disorganized blood vessels arranged as nodules

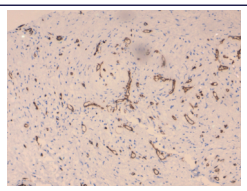

Figure 5: Histopathology showing immunohistochemistry positivity for CD 34

DIFFERENTIAL DIAGNOSIS

The common vascular tumors are littoral cell angioma [5], splenic hemangioma [6], lymphangioma, splenic hamartoma.[7]

Littoral cell angioma is a vascular lesion of spleen arising from the littoral cell, which lines splenic sinuses. They are usually multiple and positive for both endothelial and histiocytic markers. It has CD34-CD68+CD21+CD8+ immunophenotype. LCA is benign and is cured by splenectomy. There are reports of coexistence with other malignancies indicating long term follow up is strictly needed. [8,9]

Splenic hemangiomas are small <2cm benign tumors that arise from sinusoidal epithelial cells and show cavernous or capillary type. Positive for CD34,CD31 and negative for CD8,CD21,CD68.they lack the trivascular pattern of SANT.

Lymphangioma is a benign tumor of spleen involving lymphatic endothelium. It has cystic spaces filled with proteinaceous fluid. Lymphatic endothelium shows strong immunoreactivity to D2-40 and is negative for CD21 and CD8. Smaller lesions can be left alone while larger lesions require splenectomy.[10]

Splenic hamartoma is derived from cells lining the splenic sinuses and shows predominantly red pulp elements in a disorganized fashion. Positive for CD31, CD8, factor 8 and type 4 collagen.

Angiosarcoma has an irregular anastomosing vascular channel.Positivity for CD68,CD8.Primary modalities of treatment include surgery and radiotherapy, however, outcomes are very poor with an average survival of less than a year after diagnosis.[11]

All these lacs the nodular pattern of SANT and each stain for only a single type of vascular channel.

DISCUSSION

Vascular tumors are the second most common tumor of spleen after lymphoma, including hamartoma, hemangioma, littoral cell tumor, and hemangioendothelioma. SANT was first described by Krishnan et al in 1993.[14] It was initially named as cord capillary hemangioma and reinterpreted as a variant of hamartoma in 2003.SANT was defined by Martel et al according to histological and immunophenotype.

SANT has female preponderance and presents during the third to fifth decade. It is usually found incidentally. Among those presenting with symptoms, abdominal pain is the most common symptom.

The pathogenesis of this lesion remains unclear. It may be a Splenic hamartoma that has undergone an unusual form of sclerosis. SANT can resemble an inflammatory pseudotumor, as there have been reports of stromal cells being positive for Epstein-Barr virus as postulated by Weinreb et al.[13] Proliferation seen in SANT may be related to Ig G4 related sclerosing lesion due to plasma cells in the stroma. SANT could be a reactive lesion than a true neoplastic process due to several concomitant conditions. There has been one documented case of SANT, where the patient was also found to be positive for hepatitis-B.[15] Our patient had acute severe hepatitis –B infection which could have caused a reactionary transformation of the spleen.

Grossly it is a well-circumscribed non-encapsulated mass with multiple dark nodules interspersed with stroma. The distinctive feature is the presence of angiomatoid nodules surrounded by hyaline shell separated by fibrous stroma. The stroma is composed of myofibroblasts, plasma cells, lymphocytes, siderophages.

Three distinct type of blood vessels are seen:CD 34+/CD8-/CD31+ capillaries,CD 34-/CD 8+/CD 31+ sinusoids and CD 34-/CD 8-/CD 31+ small veins.CD 30,ALK-1 and EMP-LMP are negative.[13]

On CT, SANT lesions are well-circumscribed masses that are slightly hypodense. They demonstrate early peripheral enhancement which extends in a spoke wheel pattern toward the center with progressive central enhancement on delayed imaging.[16] Spoke wheel pattern is due to central stellate fibrous stroma with fibrous septa separating angiomatoid nodules. In an MRI lesion appears T2 hypointense

FOLLOW UP

The patient was put on careful observation with no definitive therapy. Over the last 1 year since diagnosis, he has had no symptoms and the lesion has not increased in size.

TREATMENT

Splenectomy has been considered the treatment of choice but bleeding has been a complication. Laparoscopic splenectomy has also been an option. In recent years conservative management in view of innocuous nature of the disease has been considered.[12][13]

LEARNING POINTS

1. SANT is a rare benign vascular tumor of the spleen.
2. It can be differentiated from other benign and malignant splenic tumors by biopsy showing typical trivascular pattern with distinct immunophenotype
3. While open and laparoscopic splenectomy is the usual modality of treatment, there is a role for conservative management.
4. It probably developed as an acute inflammatory response to severe acute hepatitis B in our patient.

REFERENCES

1. Martel M, Cheuk W, Lombardi L, Lifschitz Mercer B, Chan JK, and Rosai J: Sclerosing angiomatoid nodular transformation (SANT): Report of 25 cases of a distinctive benign splenic lesion. Am J Surg Pathol 28: 1268 1279, 2004.
2. Rosai J. Is sclerosing angiomatoid nodular transformation (SANT) of the splenic red pulp identical to inflammatory pseudotumor? Report of 16 cases. Histopathology.2009;54:494.23.
3. Diebold J, Le Tourneau A, Marmey B, et al. Is sclerosing angiomatoid nodular transformation (SANT) of the splenic red pulp identical to inflammatory pseudotumor? Report of 16 cases. Histopathology. 2008;53:299-310
4. Li L, Fisher DA, Stanek AE. Sclerosing angiomatoid nodular transformation (SANT) of the spleen: addition of a case with focal CD68 staining and distinctive CT features. Am J Surg Pathol. 2005;29:839-841.
5. Kutok JL, Fletcher CD. Splenic vascular tumors. SeminDiagn Pathol. 2003;20:128-139.
6. Hartmann M, Marx A, Geissinger E, Muller-HermelinkHK, Rudiger T. Vascular proliferations of the spleen. Pathologe. 2008;29:129-135.
7. Lee H, Maeda K. Hamartoma of the spleen. Arch PatholLab Med. 2009;133:147-151
8. Arber DA. Metastasizing splenic littoral cell hemangioendothelioma.Am J Surg Pathol 2006;30:1036–40.
9. Ben-Izhak O, Bejar J, Ben-Eliezer S, et al. Splenic littoral cell haemangioma endothelioma: a new low-grade variant of malignant littoral cell tumor. Histopathology 2001;39:469–75.
10. Morgenstem L, Bello JM, Fisher BL, Verham RP. The clinical spectrum of lymphangiomas and lymphangiomatosis of the spleen. Am Surg 1992;58:599-604
11. Montemayor P, Caggiano V: Primary hemangiosarcoma of the spleen associated with leukocytosis and abnormal spleen scan. Int Surg 1980;65:369-373.
12. Sclerosing angiomatoid nodular transformation of spleen masquerading as carcinoma breast metastasis: Importance of splenic biopsy in obviating splenectomy, Deep Dutta, Meha Sharma, Vrushali Maharajan, Prem Chopra.Indian J Pathol Microbiol 2016;59:223-226
13. Weinreb I, Bailey D, Battaglia D, Kennedy M, Perez-Ordo¯nez B. CD30, and Epstein–Barr virus RNA expression in sclerosing angiomatoid nodular transformation of spleen. Virchows Archiv: An International Journal of Pathology2007;451(July (1)):73–9.
14. Krishnan J, Frizzera G. Two splenic lesions in need of clarification: hamartoma and inflammatory pseudotumor. Semin Diagn Pathol 20: 94-104, 2003.
15. Sclerosing Angiomatoid Nodular Transformation (SANT) of the spleen: Case report and review of the literature. Gavin A. Falka, , Nishank P. Noolia, Gareth Morris-Stiff a, Thomas P. Plesecb, Steven Rosenblatta, International Journal of Surgery Case Reports • July 2012:492-500
16. Karaosmanoglu DA, Karcaaltincaba M, Akata D. CT and MRI findings of sclerosing angiomatoid nodular Sclerosing Angiomatoid Nodular Transformation of the Spleen / Koreishi et al389transformation of the spleen: spoke wheel pattern. Korean J Radiol. 2008;9(suppl):S52-S55.