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Sclerosing Angiomatoid Nodular Transformation of the Spleen: A Case Report

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ABSTRACT

Sclerosing angiomatoid nodular transformation (SANT) is a relatively new entity in the spleen, which usually presents in the form of single nodule. The etiology of SANT is unknown. SANT of the spleen is a benign lesion that does not recur after splenectomy. Although histology can lead to the diagnosis of vascular tumor, immuno-histochemistry is the only way to confirm the diagnosis of SANT.

Keywords: Benign, Nodule, Sclerosing angiomatoid nodular transformation.

INTRODUCTION

Sclerosing angiomatoid nodular transformation (SANT) is a relatively rare, new and unique benign vascular lesion of the red pulp of the spleen with extensive sclerosis that was initially reported by Martel and colleagues in 2004.[1] In this paper, we shall report a case of SANT of the spleen. Its pathogenesis is not fully understood. Splenectomy is curative.

CASE REPORT

A 36-year old man presented with an accidentally discovered solitary splenic nodule during routine screening for abdominal pain. The nodule was located in the superomedial aspect of the spleen. It appeared as a well-circumscribed and hypodense vascular lesion in the CT scan. The patient's past history was non-significant. The patient was treated with splenectomy. The spleen weighed 100 g and showed a well-circumscribed but non-encapsulated nodule measuring 1.5 cm in maximum dimension, located at the superior pole. Cut surface showed focal calcification and a grey white stellate scar in the center. Microscopically, the lesion was sharply distinct from the adjacent splenic parenchyma (Fig. 1 and Fig. 2), and was formed of capillary sized vascular spaces surrounded by thick fibrosclerotic stroma (Fig. 2). These vascular spaces contained erythrocytes admixed with chronic inflammatory cells. Peripheral fibrin deposition (Fig. 4) and extravasated red blood cells were noted. Brisk mitoses & necrosis were absent. Reticulin and CD34 stains highlighted the vessels (Fig. 5). CD31 stain was negative. The adjacent uninvolved spleen showed the expected expression for the various vascular markers, with CD8 positivity and CD34 negativity in the normal splenic sinusoids. The case was diagnosed as sclerosing angiomatoid nodular transformation of the spleen (SANT). The patient did not received any further therapy, was alive, and well during the follow up.

DISCUSSION

Sclerosing angiomatoid nodular transformation (SANT) is a distinct pathological vascular lesion of the spleen newly described by Martel et al. in 2004.[1] In a series published by Diebold *et al.*, the majority of patients were asymptomatic, but had splenomegaly or abdominal pain.[2] In the present case, this lesion was incidental finding on imaging studies performed for other reasons. Vascular lesions in the spleen include hemangioma, littoral cell angioma, lymph - angioma, hemangioendothelioma and splenic hamartoma. SANT is best differentiated from these tumors by immunohistochemical staining, but it also has a different histological appearance. FNA and core biopsy carry risks of bleeding and needle tract seeding. Therefore, splenectomy may be the preferred modality to rule out malignancy or other pathological processes. These nodules had an angiomatoid appearance, in the sense that they were composed of slit-like, round or irregular shaped vascular spaces lined by plump endothelial cells and interspersed by a population of spindly or ovoid cells.[3] Nuclear atypia

was minimal, mitotic figures were extremely rare, and necrosis was consistently absent. The internodular stroma consisted of variably myxoid to dense fibrous tissue with scattered plump myofibroblasts, plasma cells, lymphocytes and siderophages. On Immunostaining these lesions show three distinct types of vessels: a cord capillary-like type that co-express CD34 and CD31 but not CD8, a sinusoid-like type that express CD8 and CD31 but not CD34, and small veins that express only CD31. These features are, therefore distinct from those of littoral cell angioma, conventional hemangioma and hemangioendothelioma of the spleen. The pathogenesis of SANT is unknown. Diebold *et al.* postulated that passive congestion of the red pulp may cause metabolic changes in those areas, damaging the sinus endothelial cells. This may cause fibrin deposition and inflammation, as seen in granulation tissue.[2] Martel *et al.* hypothesized that SANT was a response to stromal proliferation and that the internodular zones were very similar to inflammatory pseudotumor.[1]

On immunohistochemical staining, SANT appeared like a splenic hamartoma because of the red pulp tissue composition, as theorized by Awamleh and Perez-Ordonez.[4] Kuo *et al.* have connected the plasma cells and stromal sclerosis present in SANT to IgG4-related sclerosing disease.[5] This idea is supported further by a recent report of three cases by Koreishi *et al*., in which all three cases had positive IgG4 plasma cells.[6] According to Weinreb *et al.*, Epstein-Barr virus has some link with SANT, however, in their set of six cases, only one case was positive for the virus.[7] Koreishi *et al*. also tested for the Epstein-Barr virus, and in their three cases, all were negative.[6] SANT is a benign lesion, and splenectomy is curative. In the cases reported to date, recurrence of SANT does not occur after splenectomy.[1] More research about SANT is necessary, and as more cases are being described, an etiology will likely be discovered.

CONCLUSION

The diagnosis of SANT should be considered in any patient presenting with a splenic lesion that contains an angiomatoid or inflammatory component. There is a wide age distribution and the gender distribution appears to be equal. The majority of cases of SANT reported in the literature were incidental diagnoses, with the remainder presenting with non-specific symptoms. Differential diagnosis for SANT includes other vascular lesions including malignant pathologies. These entities cannot be differentiated on the basis of any radiological features, hence surgical histopathology remains the best option.

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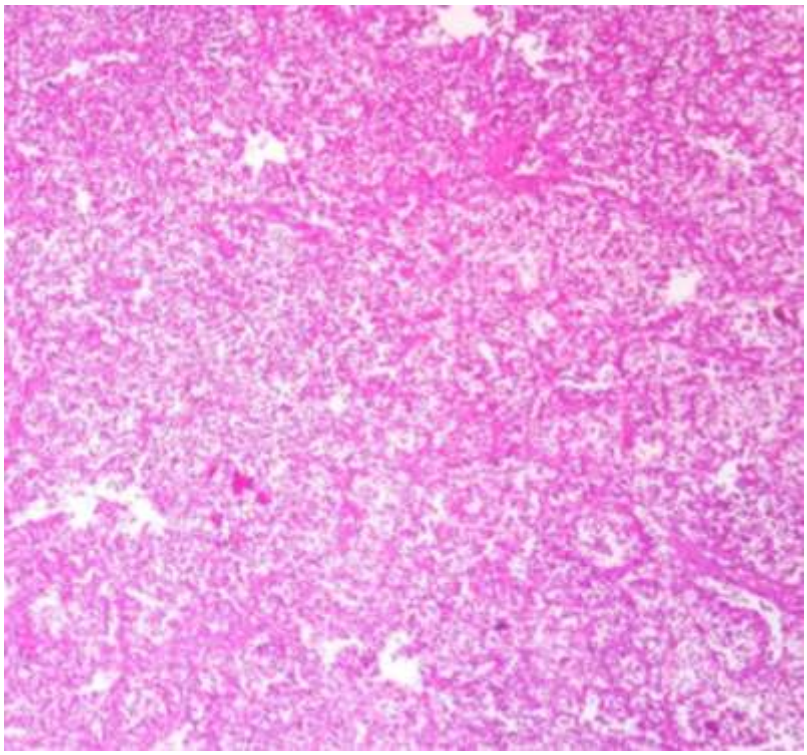

Figure 1: The lesion was composed of multiple, variably sized, circumscribed and confluent angiomatoid nodules and a fibrosclerotic stroma on periphery.(H&EX10)

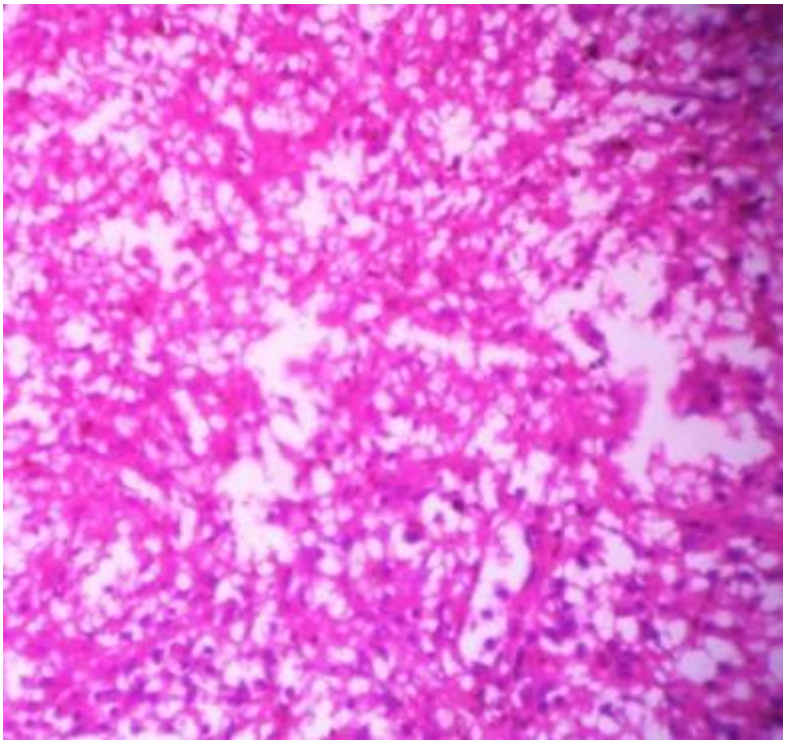

Figure 2: The lesion was composed of multiple, variably sized, circumscribed and confluent angiomatoid nodules and a fibrosclerotic stroma on periphery.(H&EX40)

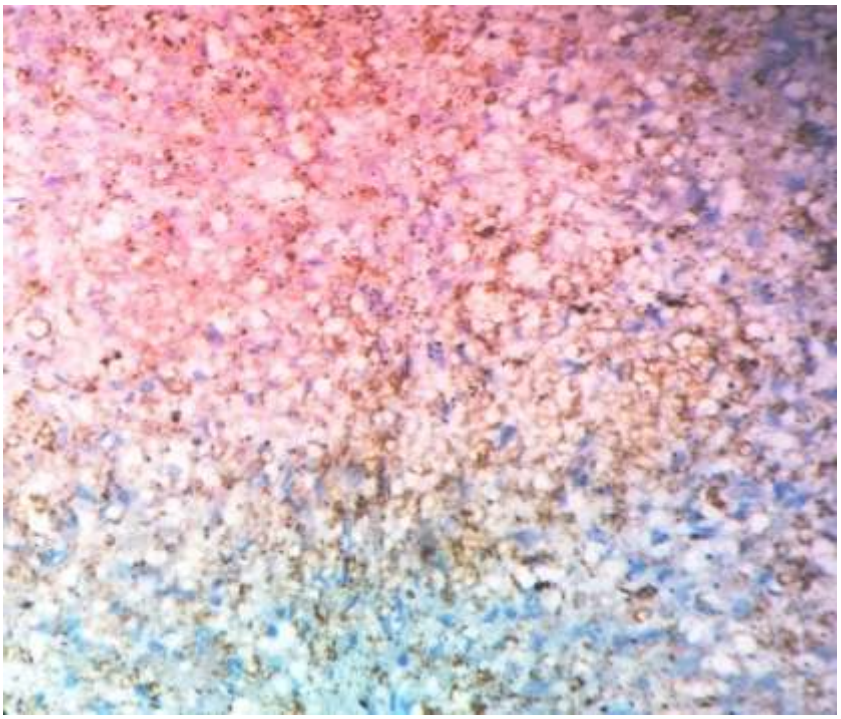

Figure 3: CD31 was positive in the lesion.(H&EX40)

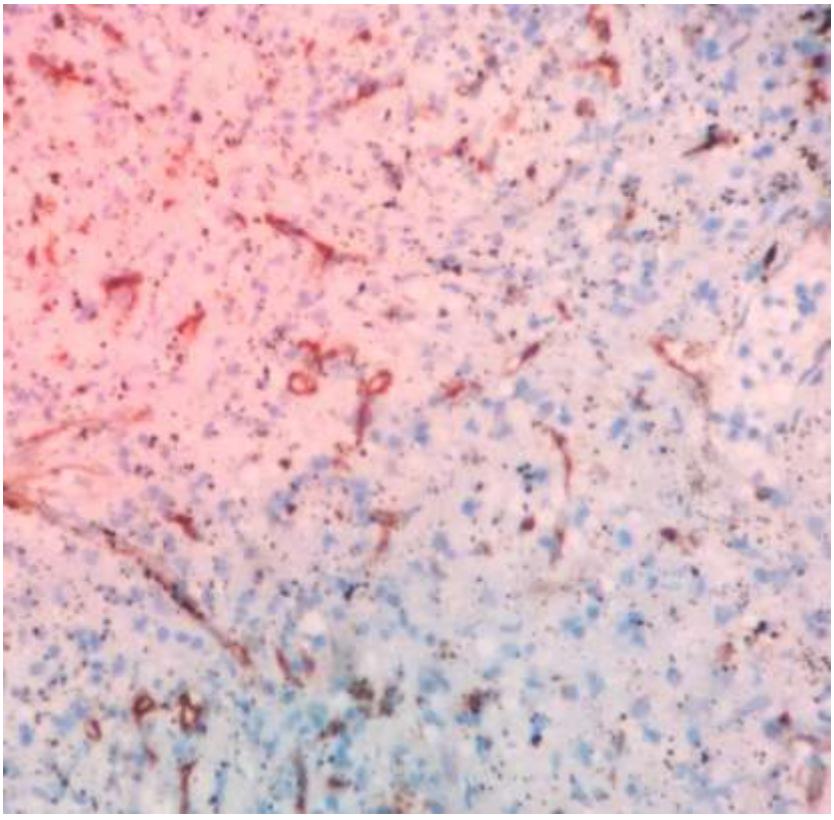

Figure 4: The capillaries were positive for CD34(H&EX40)