DIAGNOSIS

In view of the histomorphologic and immunohistochemical evaluation, a diagnosis of Sclerosing Angiomatoid Nodular Transformation (SANT) of spleen was given.

DISCUSSION

The term SANT was introduced by Martel et al in 2004.[1] This entity was previously called splenic hamartoma, cord capillary hemangioma and multinodular hemangioma.[2]

SANT comes under the category of vascular neoplasm of spleen that include other benign and malignant vascular neoplasms namely, splenic hamartoma, lymphangiomas, hemangioendothelioma and angiosarcoma. SANT resembles a spleninc hamartoma in many aspects but in contrast to a splenic hamartoma that exhibits only sinusoids, SANT exhibits three types of blood vessels.[3]

This disease has a female preponderance and is found commonly in an age group of 30 to 60 years with abdominal pain being the most common symptom of this disease entity, as seen in the index case.

The pathogenesis of SANT is unclear, however it is hypothesized that it could be a splenic hamartoma that has undergone an unusual form of sclerosis with peculiar reactionary transformation of red pulp due to an exaggerated stromal response. In view of predominant plasma cells infiltrate in this lesion, it is also postulated that the sclerosis occurs due to IgG4 related pathogenetic process.[4, 5]

The differential diagnoses of SANT are Littoral cell angioma, hemangioma, lymphangioma, splenic hamartoma, and hemangioendothelioma. Littoral cell angioma comprises of splenic sinuses lined by tall endothelial cells with variable hemophagocytosis. The cells are positive for both endothelial and histiocytic markers with a CD34-/CD68+/CD21+/CD8- immunophenotype. Splenic hemangiomas show cavernous and capillary configuration of vascular channels that are lined by flattened endothelial cells. In lymphangiomas, the dilated vessels are filled with proteinaceous fluid. These vessels are highlighted by D240 immunostain in addition to vascular markers. Both the above entities however, lack the classical triphasic vascular proliferation seen in SANT.