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Sclerosing Angiomatoid Nodular Transformation of the spleen, focal nodular hyperplasia and hemangioma of the liver: A tale of three lesions

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ABSTRACT

Sclerosing Angiomatoid Nodular Transformation (SANT) of the spleen is a benign vascular lesion with peculiar histological features. The pathogenesis of SANT is still largely unknown and variable etiologies have been proposed, including autoimmune, inflammatory, and/or vascular disorders. The present report describes a unique case of splenic SANT, associated with focal nodular hyperplasia and a sclerosing hemangioma of the liver. The clinic-pathological features of such an unusual case are thoroughly illustrated. Its possible pathogenic mechanisms are also briefly addressed.

1. Introduction

Sclerosing Angiomatoid Nodular Transformation (SANT) is a benign vascular lesion of the spleen, characterized by the proliferation of nodular vascular aggregates embedded in a dense fibro-connective stroma [1]. First described as "cord capillary hemangioma" in 1993, SANT has been thoroughly characterized by Martel and colleagues in a subsequent series of 25 cases [2]. Since then, approximately 130 cases of SANT have been reported in the literature.

The pathogenesis of SANT is still poorly understood. This lesion may indeed represent an abnormal vascular/stromal reaction to a broad spectrum of inflammatory or thrombotic/hemorrhagic accidents of the red pulp [2]. In recent years, a role for autoimmune/dysimmune disorders has also been highlighted [3].

The rarity of SANT and its nondescript radiological features represent a diagnostic challenge for the clinician [4]. On the other hand, its gross and microscopic features are relatively specific and allow the correct diagnosis in the majority of cases. The histologic appearance of SANT can however mimic other vascular and/or stromal lesions of the spleen [1]. In such cases, only a high degree of suspicion, together with adequate sampling and proper immunohistochemical analysis lead to the correct diagnosis.

In this report, we describe a case of splenic SANT associated with two primary lesions of the liver (focal nodular hyperplasia [FNH] and sclerosing hepatic hemangioma). The clinic-pathological features and differential diagnoses of SANT are thoroughly addressed. Its possible pathogenic mechanisms are also briefly outlined.

2. Materials and methods

2.1. Clinical and laboratory data

The patient's clinical and laboratory data were retrieved from the clinical charts of the Hepatobiliary Surgery and Liver Transplant Center (Padova University Hospital, Padova-Italy). The following clinical, radiological and laboratory parameters were considered: (i) symptoms and physical findings at presentation; (ii) laboratory tests at admission; (iii) abdominal ultra-sound (US), computerized axial tomography (CT) and magnetic nuclear resonance (MNR) findings before splenectomy. The patient's informed consent was obtained, consistent with the Declaration of Helsinki and with the Institutional rules on clinical research.

Abbreviations: FNH, focal nodular hyperplasia; SANT, Sclerosing Angiomatoid Nodular Transformation.

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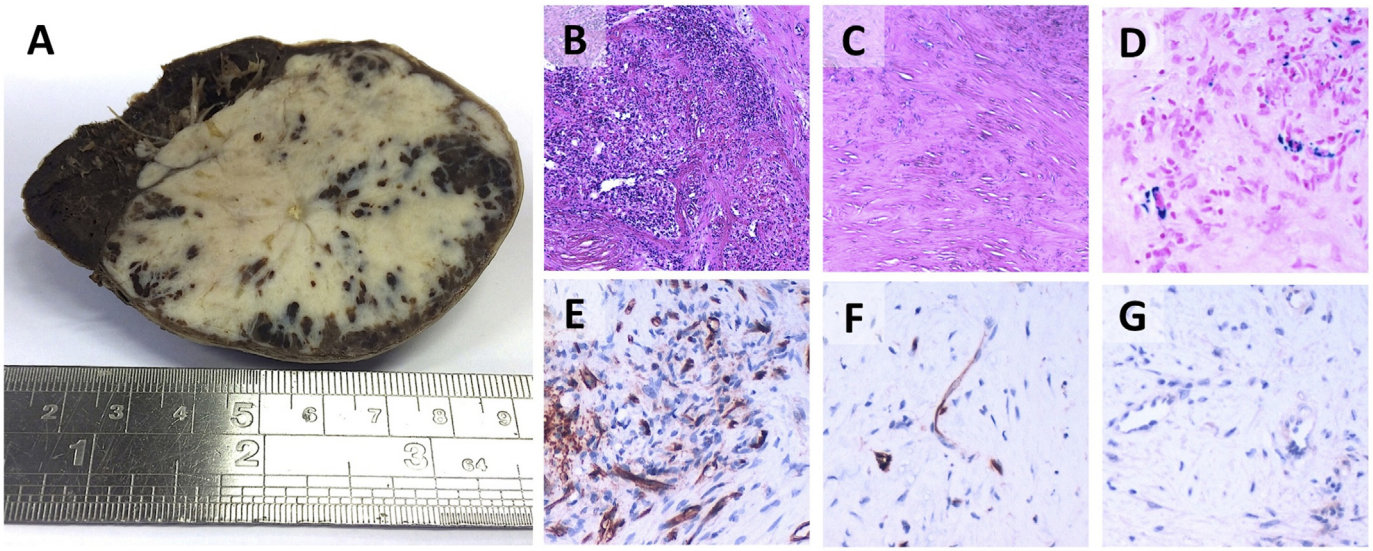

Fig. 1. Histological and immunohistochemical features of splenic SANT.
On gross examination, the lesion was composed of fibro-calcific tissue, with occasional angiomatoid nodules (A). Histologic evaluation disclosed the presence of nodular aggregates of micro-vessels, surrounded by scant inflammatory cells (B) and dense fibrous tissue (C); occasional hemosiderin-laden macrophages were also documented (D). Immunohistochemical analysis revealed the co-existence of CD34-positive capillaries (E) and CD8-postive red pulp sinuses (F). Endothelial cells were negative for CD30 (G) (H&E, Prussian blue and immunoperoxidase stains; original magnification 10× and 20×).

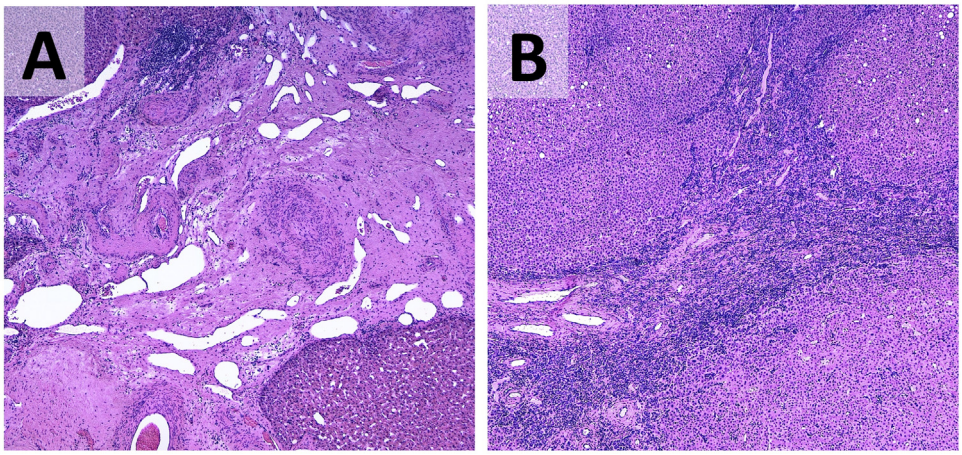

Fig. 2. Histological features of the hepatic lesions.
(A) The larger lesion was composed of irregular blood vessels, embedded in a dense fibrous stroma; the histological features were consistent with a sclerosing hemangioma of the liver. (B) Focal nodular hyperplasia consisted of a well-demarcated proliferation of hepatocytes, centered by a fibrous scar. Medium to large vascular structures with eccentric wall thickening were noted within the lesion. (H&E stain; original magnification 10×).

2.2. Histological evaluation

Formalin-fixed, paraffin-embedded tissue sections were stained with Hematoxylin and Eosin (H&E) for histological evaluation. Immunohistochemical analysis was performed on a fully automated immunostainer (Bond-maX; Leica, Newcastle Upon Tyne, UK) [5]. The following primary antibodies were used: CD34 (clone QBEnd/10, Leica; Newcastle, United Kingdom), CD31 (clone JC70A, Dako; Glostrup, Denmark), CD8 (clone C8/144B, Dako; Glostrup, Denmark), CD30 (1G12, Leica; Newcastle, United Kingdom), CD38 (SPC32, Leica; Newcastle, United Kingdom) CD68 (PGM-1, Dako; Glostrup, Denmark), IgG (polyclonal, Dako; Glostrup, Denmark) and IgG4 (clone HP6025, Thermo Scientific; Rockford, USA). Iron stain with Prussian blue was performed to confirm the presence of perivascular hemosiderin-laden macrophages.

3. Case report

A 57-year-old Caucasian man with unremarkable past medical history presented at Padova University Hospital (Padova, Italy) for mild left upper abdomen discomfort. Physical examination was negative, but for mild splenomegaly. Laboratory tests disclosed normal blood cell counts (red blood cells: 4.35×10^3/mL; hemoglobin: 12.9 g/dL; white blood cells: 6.15×10^3/mL; platelets: 376×10^3/mL) and normal liver and renal function tests. Reactive C protein, erythrocyte sedimentation rate and total serum immunoglobulins were within normal ranges.

An abdominal US disclosed a large, well demarcated, bosselated lesion of the spleen (main diameter 7 cm), with heterogeneous echogenicity and focal calcifications. CT and MNR scans confirmed the US findings and documented the presence of two focal lesions