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Robotic Splenectomy for Sclerosing Angiomatoid Nodular Transformation

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Abstract Splenic masses represent a clinical challenge when encountered by health care providers. These lesions must be excised completely and assessed histologically to differentiate benign and malignant diseases. Biopsy mass bleeding and false negative results are common. We report a 59 year old woman with no previous cancer history, with an incidental splenic mass found by abdominal ultrasound for left abdominal pain. Computer tomography scan and magnetic resonance imaging identified a solid nodular splenic lesion lacking definitive criteria for benign or malignant disease. Furthermore, liver/spleen scan ruled out hemangioma. After discussing the patient at tumor board, a robotic splenectomy was performed. The patient was discharged home 20 hours postoperatively. Sclerosing Angiomatoid Nodular Transformation (SANT) was confirmed by histology and immunohistochemistry. Imaging modalities are not helpful to clinch the diagnosis, leaving minimally invasive splenectomy and pathology to treat and assess this very rare splenic disease.

Keywords: *Splenic mass, Robotic splenectomy, Immunohistochemistry, Triple Vessels Disease, Sclerosing Angiomatoid Nodular Transformation*

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1. Background

Sclerosing Angiomatoid Nodular Transformation (SANT), first described by Martel, represents a rare benign vascular disease of the splenic pulp, involving the formation of angiomatoid nodule surrounded by fibrosclerotic stroma [1]. The diagnosis is confirmed by histological evaluation and supported by immunohistochemistry. Those lesions are incidentally discovered by imaging modalities due to abdominal pain, splenomegaly, weight loss or previous history of malignancy [2,3]. Originally recognized more commonly in women, the incidence is equal among genders and the mean age upon presentation is 46 year old [4]. Since the final diagnosis of vascular splenic lesions can be only confirmed by immunohistochemical evaluation, hamartomas, inflammatory pseudotumor, lymphomas, angiomatoid endothelioma, littoral angioma or metastases are still considered in the differential diagnosis [5,6,7].

Open or minimally invasive splenectomy is recommended in treating this solid angiomatoid transformation. Robotic splenectomy for SANT has not been previously described. As aim of this case presentation, we emphasize the paramount role of immunohistochemical technique to confirm the diagnosis of this rare splenic entity and we describe the robotic surgical steps in treating splenic solid masses.

2. Case Presentation

A 59 year-old woman presented to our multidisciplinary clinic with intermittent left abdominal pain and mild weight loss for few months. She had no prior malignancy and had normal laboratory values. Upon presentation, her abdominal US showed a 5,2 cm hypoechoic splenic mass extending into the splenic hilum (Figure 1). Coccidiodal serology was negative, as well as the carcinoembryonic antigen (CEA).

3. Investigation

Triple phase computer tomography scan (CT scan) of the abdomen showed a complex splenic lesion of 5 cm in diameter with central irregular calcifications. On unenhanced CT the mass was hypoattenuating with similar spleen density, while on arterial phase the lesion was heterogenous with rim enhancement and radial lines (Figure 2). On venous and delayed phases, the lesion appeared with progressive filling and spoke wheel feature

was noticed within the mass (Figure 3). Magnetic resonance imaging confirmed a 4.6 x 5.2 cm enhancing splenic lesion suspicious for atypical hemangioma on T1 weighted and T2 weighted images (Figure 4, Figure 5). She underwent liver / spleen nuclear medicine with single photon emission tomography scan. The test showed photopenia in the region of the mass of the spleen ruling out the presence of atypical hemangioma (Figure 6). The case was discussed at the multidisciplinary tumor board and, considering the risks for a splenic biopsy and the inconclusive imaging findings, splenectomy was recommended.

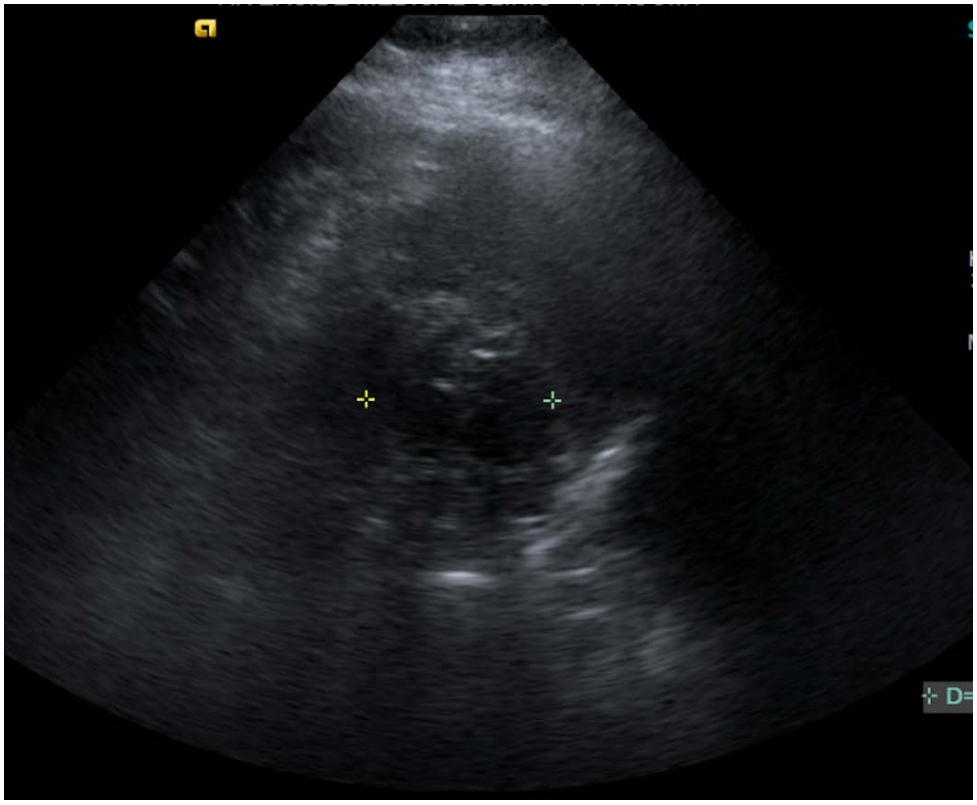

Figure 1. Ultrasound of splenic mass

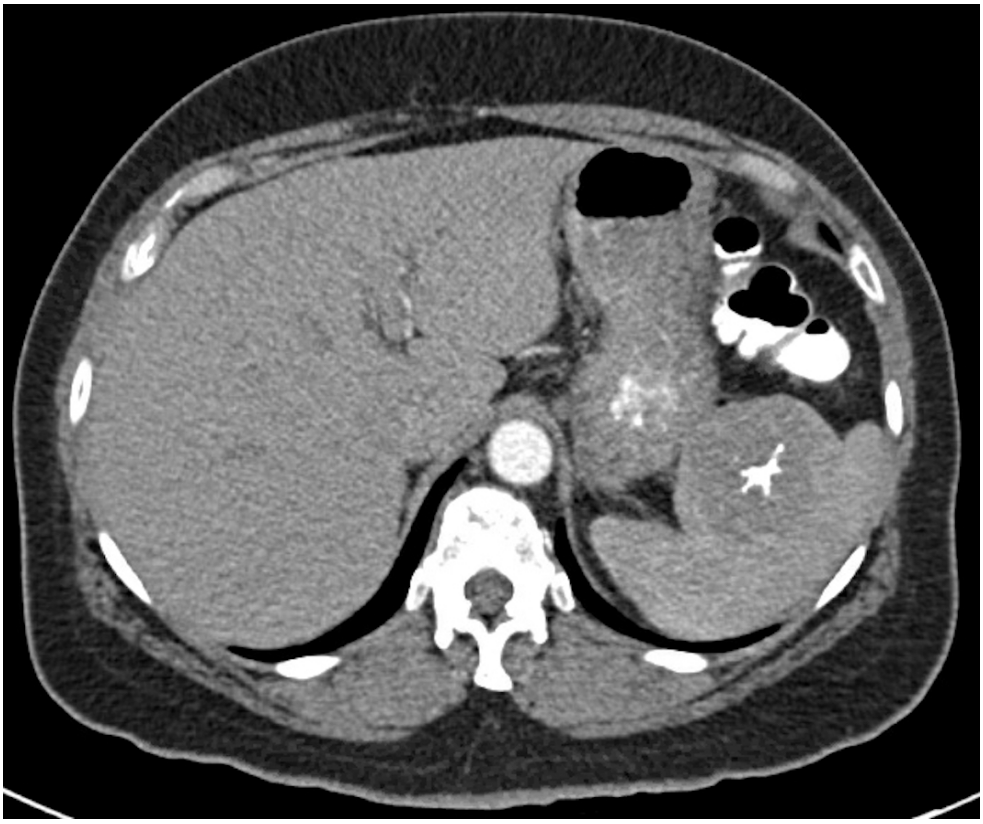

Figure 2. CT scan of splenic mass - arterial phase

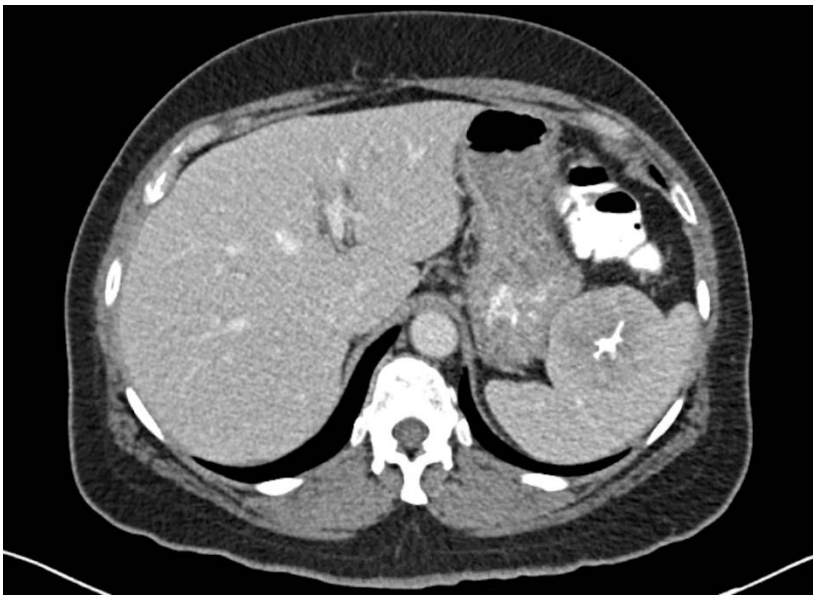

Figure 3. CT scan of splenic mass - Portal 4 minutes phase

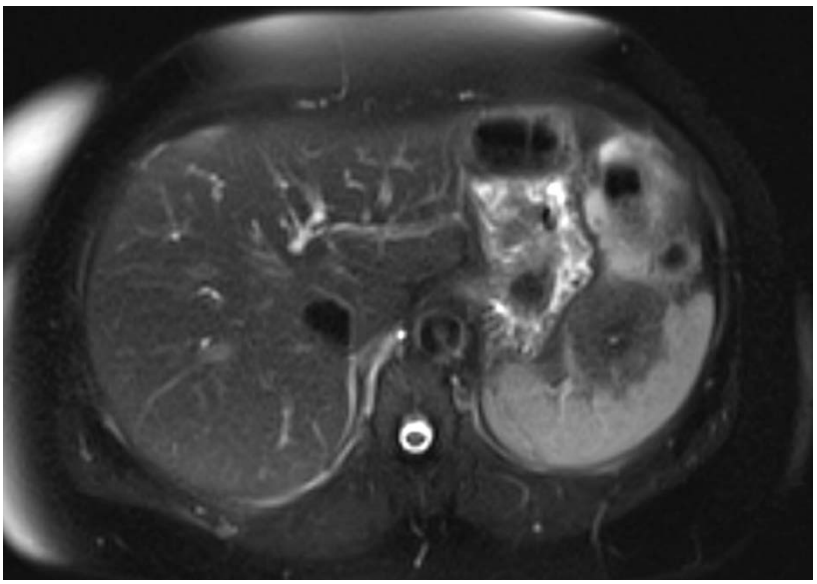

Figure 4. MRI of splenic mass - T1 – weighted

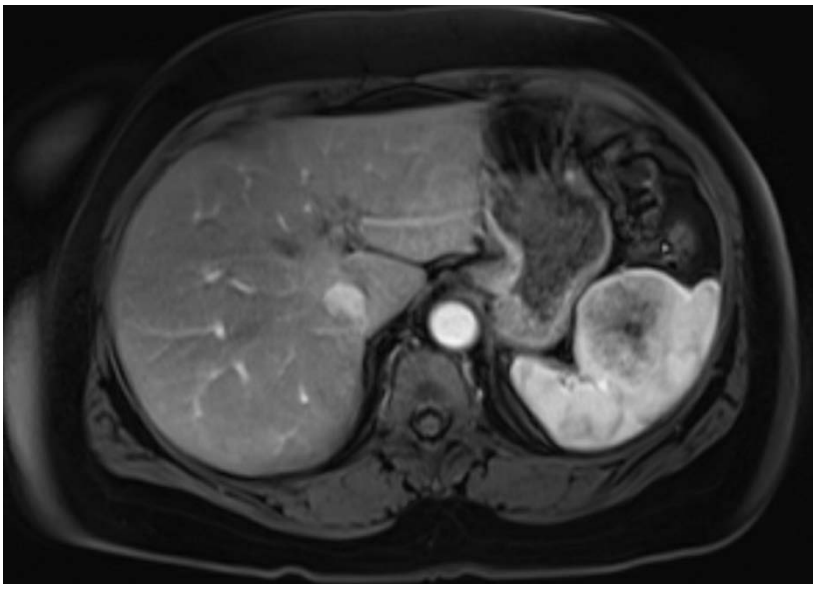

Figure 5. MRI of splenic mass - T2 – weighted

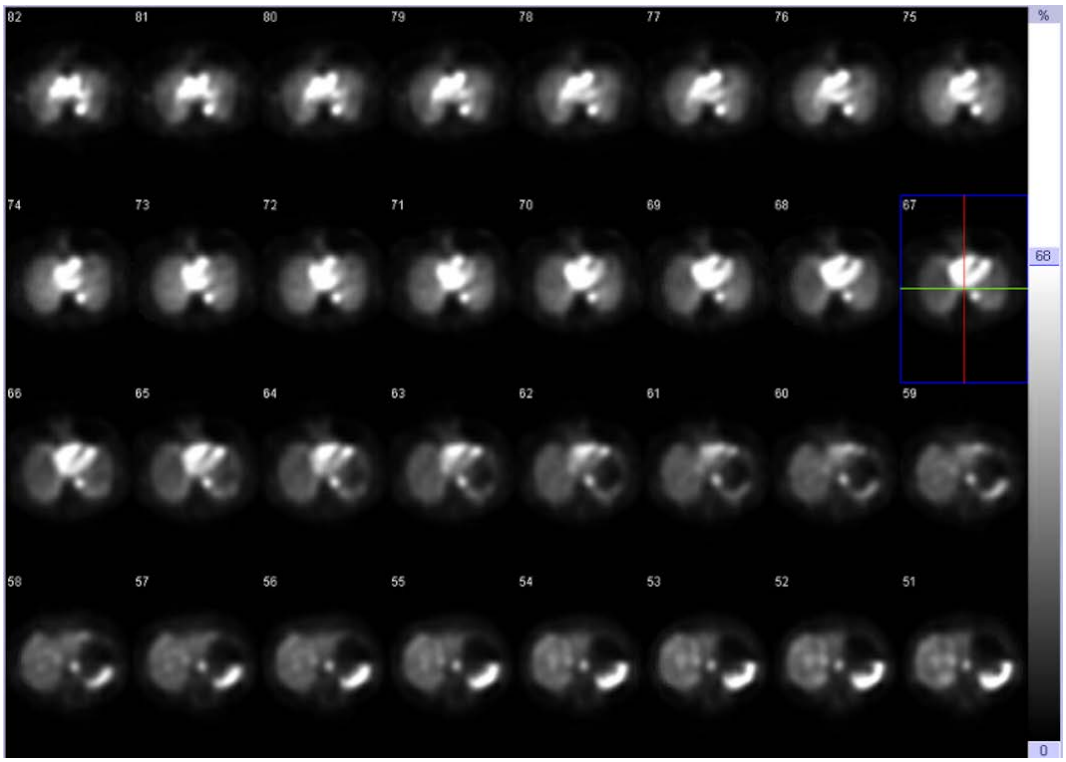

Figure 6. Liver / spleen nuclear medicine with single photon emission tomography scan showing photopenia of the splenic lesion

4. Treatment

The patient was given post-splenectomy vaccination including pneumonia, hemophylous influenza and meningococcus vaccines according to the Centers for Disease Control and Prevention guidelines (CDC). A robotic splenectomy was performed. A Xi robotic model (Intuitive Surgical Inc., Sunnyvale, CA) was utilized to perform the operation. Figure 7 shows the incisions for robotic splenectomy and extraction of the spleen in its entirety. Operative time was 157 minutes from docking to undocking the robot. The main steps of the operations included opening the gastro-colic ligament, taking down the short gastric vessels to mobilize the stomach from the gastro-splenic ligament (Figure 8A), mobilizing the splenic flexure to visualize the lower pole of the spleen and the pancreatic body. The splenic artery was then identified, dissected and stapled with a vascular cartridge controlled robotically (Figure 8B) and all branches of the splenic vein were dissected and divided individually using intra-corporeal clips or ligatures (Figure 8C). After controlling the splenic hilum, the spleno-renal ligament was taken down preserving the integrity of the spleen to avoid potential seeding in case of malignancy (Figure 8D). The robotic platform is ideal to avoid this potential risk. The spleen was then placed in an endo catch bag (Medtronic, Minneapolis, MN) and extracted using a mini Pfannenstiel supra-pubic incision. A closed drain was left in the splenic fossa at the end of the procedure. Figure 9 shows the extracted spleen with the solid mass.

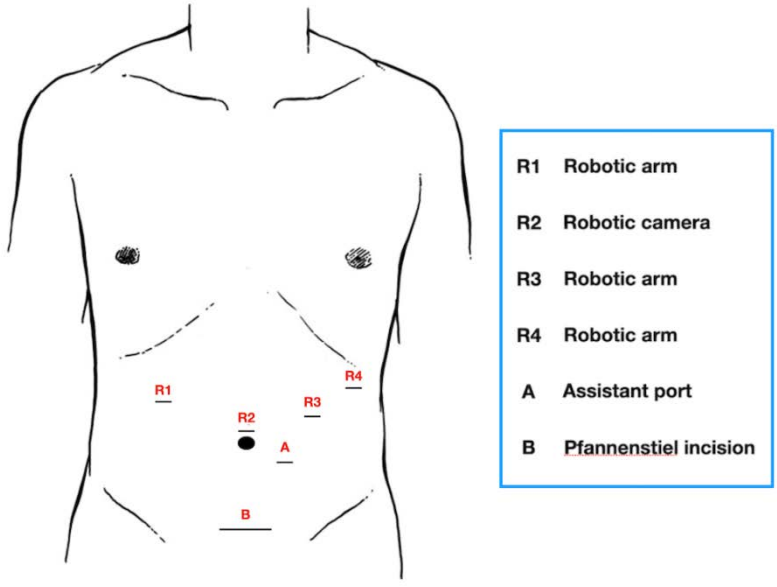

Figure 7. Position of trocars and extraction incision for robotic splenectomy

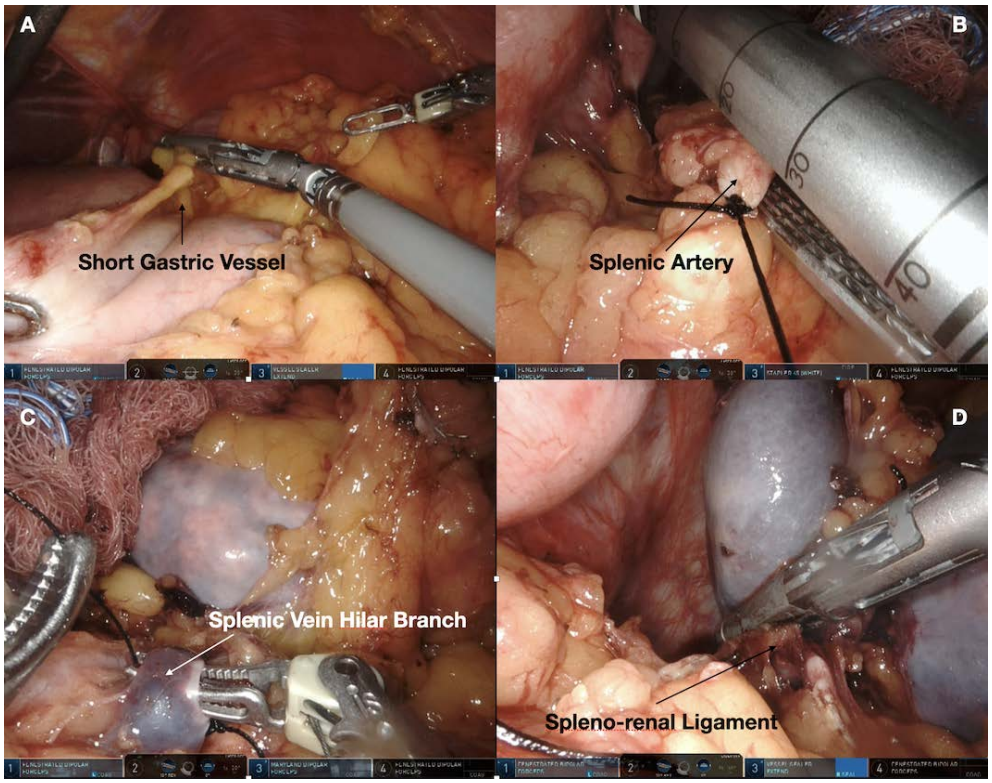

Figure 8. A: Division of short gastric vessels; B: Encircling splenic artery with its division; C: Ligation and division of hilar splenic vein branches; D: Division of the spleno-renal ligament

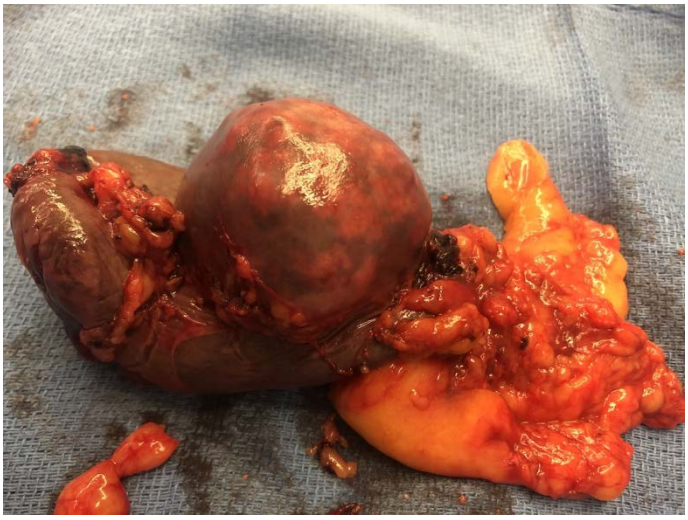

Figure 9. Specimen of the spleen with the solid mass

5. Outcome and Follow-up

The patient was discharged after 20 hours of hospital stay tolerating regular diet. No pancreatic leak was detected based on the normal value of lipase from the drain. The drain was removed before her discharge. In follow up, she is doing remarkable well and she received all the required vaccinations based on CDC recommendations.

6. Pathology

The spleen revealed a well-circumscribed sub-capsular lesion with a tan-grey cut surface measuring 5.3 x 5.0 x 4.0cm in overall dimension without necrotic changes (Figure 10).

Microscopically, the lesion revealed micronodular structures with slit-like and irregular shaped vascular spaces, lined by endothelial cell, intermixed with epithelioid and spindle cells. These structures can be described as angiomatoid nodules. The stoma contained myxoid and dense fibrous tissue with scattered myofibroblasts and lymphoplasmacytic infiltrate. Immunohistochemical staining of the small vessels within the nodules was positive for CD34, CD31 and predominantly negative for CD8 as well as AE1/AE3, Desmin and S100.

In addition, EBER ISH study was performed and was negative. Specific morphology and immunohistochemical findings distinguish sclerosis angiomatoid nodular transformation from other splenic vascular tumors.

Figure 10. Gross appearance of splenic SANT with well-circumscribed sub-capsular lesion with a tan-grey cut surface

7. Discussion

First described by Martel [1], SANT represents a very rare benign entity of the pulp of the spleen which can be confused with splenic hemangiomas. There is still lack of agreement how this benign disease undergoes an angiomatoid transformation with associated sclerosis. In the original paper, it was speculated that a hamartoma undergoes nodular transformation with associated sclerosis, or the SANT could be secondary to a transformation of an organized hematoma. More recently, Chang et al hypothesized that the etiology of SANT is secondary to vascular insufficiency of the spleen which is subsequently repaired by vascular proliferation [9]. In the past, the prevalence of this rare entity favored the female gender like in our described patient, but with recently published case reports, the gender distribution is equal today [4]. This entity of the spleen has similar features of previously described splenic lesions. About 10 years ago, a similar splenic mass, characterized by angiomatoid transformation associated with peripheral nodularity, was described in an abstract as "Cord Capillary Hemangioma" [8]. More recently, Diebold et al. accredited in their review the nomenclature utilized by Martel, and retrospectively named more appropriately their inflammatory pseudotumor series as SANT, based on the presence of angiomatoid nodules and not follicular dendritic cells [7]. Contrary to our patient presenting with abdominal pain and mild weight loss, majority of patients, in whom SANT is incidentally found, are asymptomatic [10]. Currently there are no definitive radiographic criteria that can help the clinician narrow the differential diagnosis when facing angiomatoid solid lesion of the spleen. When detected incidentally, the ultrasound findings are characterized by hyperechoic or sometimes hypoechoic features based on the moment in time the lesion is analyzed, and this variability is more confusing when internal septa are encountered [11]. The utilization of innovative CT and MRI methodologies help narrow the possible diagnosis of this very rare splenic disease. Like our patient, Vigorito et al. described in a non-enhancement CT scan a hypoattenuating splenic lesion, which enhanced peripherally in the arterial phase with centripetal filling and hypodense center (the spoke wheel) in the delayed phase [12]. A very elegant paper by Liao et al, was recently published and confirmed Vigorito's findings. The Authors performed an image analysis with CT scan and MRI in 18 patients with pathologic features suggesting SANT, primarily characterized by nodular transformation, fibrous scar and occasional calcifications. They confirmed that enhancement characteristics like centripetal enhancement, nodular enhancement and delayed enhancement of central fibrous scar were all typical characteristic findings of SANT. Particularly, the presence of hemosiderin deposition and fibrous scar tissue were correlated with the hypoattenuation on T2-and diffusion-weighted images on MRI [13].

Red cell scan was utilized in our patient and the findings were not diagnostic for splenic hemangioma. Our study showed a photopenia with only a thin rim of patchy tracer activity at the periphery of the lesion which ruled out splenic hemangioma, but raised the possibility of a splenic hamartoma. This nuclear medicine technique was described only one time in all the literature related to SANT [14]. We believe this methodology should be considered as part of our armamentarium, narrowing our differential diagnosis in preparation for surgical treatment.

To the best of our knowledge, our patient represents the first robotic splenectomy for SANT. All previous case series or case reports were compiled with open or laparoscopic splenectomy or partial splenectomy [4,15]. We described in details our surgical steps using robotic surgical platform. Whereas there has been significant improvements in laparoscopic instruments over the years, they are still limited in their ability to perform surgery in tight spaces or challenging angles. Those limitations make traditional minimally invasive splenic surgery challenging, particularly in the treatment of splenomegaly. The recent introduction of robotic surgery reduces those limitations by granting the user increased dexterity of movements with the advent of wristed instruments. Being diagnostic and therapeutic, we believe robotic splenectomy should be considered an upgraded technique to maintain the integrity of the organ in possible malignant conditions, and to use the ergonomic properties of the instruments in encircling and controlling the splenic vessels achieving minimal blood loss.

Recently, Jin et al, performed a retrospective analysis on the largest patient population with diagnosis of SANT and compared open splenectomy in 12 patients to laparoscopic splenectomy in 12 patients and in laparoscopic partial splenectomy in 13 patients. The outcome measures included operating time, blood loss, hospital course and postoperative complications and all parameters were comparable among all three groups, proving the laparoscopic partial splenectomy might be an alternative approach in treating this rare disease [15].

The morphology and immunohystochemical findings represent typical angiomatoid nodularity that is the

landmark feature for the SANT [1]. Some studies described a correlation between Epstein-Barr virus and SANT, suggesting that the pathophysiology of this splenic lesion could be similar to an inflammatory tumor [16]. We did not find this evidence in our patient. The only modality to confirm the diagnosis of SANT is histological evaluation supported by immunohistochemistry [1]. Three different types of narrow capillary vessels were noticed within the angiomatoid nodules, similarly seen in the normal red pulp. All three vessels are positive for CD31. However, they can be distinguish from other CD31 expressions, because the presence of other receptors (CD34 and CD8) varies based on their morphologic features and distribution [17]. Our patient presented positive immunohistochemistry staining of the small vessels within the angiomatoid nodules for CD34 and CD31, but predominantly negative for CD8.

8. Learning Points

- Sclerosing Angiomatoid Nodular Transformation (SANT) is a rare splenic disease characterized by the presence of angiomatoid nodules.
- Imaging modalities lack of criteria to identify any splenic solid mass as benign or malignant disease.
- Minimally invasive splenectomy has diagnostic and therapeutic roles in SANT.
- Robotic splenectomy has a paramount role in treating solid lesions of the spleen, treasuring the dexterity of movements, granting the integrity of the spleen and minimizing risks of bleeding.
- Immunohystochemistry is the landmark to identify SANT, assessing the presence of CD31, CD 34 with rarely CD8 expression in this three narrowed vessels capillary disease.

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