SCLEROSING ANGIOMATOID NODULAR TRANSFORMATION (SANT) APPEARING AS METASTATIC DISEASE IN A PATIENT WITH MEN1

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BACKGROUND

- Sclerosing angiomatoid nodular transformation (SANT) of the spleen is a rare disease (incidence of 0.007% in all operating and autopsy specimens) with unknown origin.
- SANT affects adult women more than men (56% vs 44%) between 30 to 60 years of age.
- SANT usually is asymptomatic but sometimes is associated with vague, non-specific abdominal symptoms such as early satiety and pain. SANT can also cause anemia/pancytopenia and splenomegaly.
- SANT is likely benign as splenectomy has been curative in all cases. The natural history of these lesions, such as risk of rupture or malignant conversion, remains unknown as all patients had splenectomies to obtain their diagnosis.
- SANT has few defining radiological features and can mimic the appearance of malignant tumors on most imaging modalities. Imaging with PET is unhelpful due to background uptake of radiotracers in the spleen. Imaging with MRI and multiphasic CT scans also have not been shown to be able to rule out malignancy in these cases. Even ultrasound is inconclusive due to the variability of appearance of SANT.
- Percutaneous biopsy of the spleen is dangerous and generally avoided for splenic masses due to high risk of post-interventional bleeding and intraoperative seeding if the lesion was to be malignant.
- Due to the diagnostic dilemma of SANT, workup generally leads to splenectomy and subsequent diagnosis of SANT on final pathology.

CASE DESCRIPTION

We present the case of a 45-year-old female with a significant history of Multiple Endocrine Neoplasia type 1 syndrome confirmed by molecular testing at the age of 21. She had a spleen sparing pancreatectomy at the age of 21 due to multiple neuroendocrine lesions throughout her pancreas. Since her surgery she has had vague abdominal complaints including early satiety, bloating, intermittent pains, and eventually leading to slow weight loss. CT was obtained due to her symptoms and identified two indeterminate lesions of her spleen, but they were small (see “Initial Images”). The plan was to follow with serial imaging. A biopsy of a liver lesion at that time was benign. Eight months later, a CT scan was obtained on postoperative day 13 with resolution of her abdominal symptoms but with ongoing constipation. There was no evidence of postoperative complication.

INTERVENTIONS & TIMELINES

Due to the increase in number and size of her splenic lesions as well as her MEN-1 syndrome and history of NE tumors, there was concern for metastatic disease. A PET Dotatate CT scan was ordered which did not identify any neuroendocrine tumors in her abdomen, but evaluation of the spleen was limited due to background physiologic activity. An MRI of her abdomen was ordered which identified 4 distinct lesions previously seen on CT scan and were indeterminate but considered suspicious due to radiographic progression.

FINAL PATHOLOGY

- The peritoneal biopsies were consistent with dystrophic calcifications and scar tissue.
- The liver biopsy was consistent with scar and benign liver tissue.
- The spleen was consistent with sclerosing angiomatoid nodular transformation of the spleen (SANT) – Representative case slides below.

OUTCOME

Patient was discharged from the hospital on postoperative day 2 having recovered without complication. She presented to our office for routine follow-up on postoperative day 13 with resolution of her abdominal symptoms but with ongoing constipation. There was no evidence of postoperative complication.

REFERENCES