Differentiation of an Unclear Splenic Lesion in a Patient With Cholangiocarcinoma

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Abstract: Here we report on a 51-year-old man with the primary diagnosis of cholangiocarcinoma. Workup with CT and contrast-enhanced ultrasound revealed an additional lesion in the spleen, raising the concern for metastasis. Combined FDG PET/CT revealed a different metabolic pattern, making a metastasis unlikely. Histopathology of the splenic lesion confirmed sclerosing angiomatoid nodular transformation, a rare benign lesion of the spleen.

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REFERENCES

- Martel M, Cheuk W, Lombardi L, et al. Sclerosing angiomatoid nodular transformation (SANT): report of 25 cases of a distinctive benign splenic lesion. Am J Surg Pathol. 2004;28:1268–1279.
- Kuo TT, Chen TC, Lee LY. Sclerosing angiomatoid nodular transformation of the spleen (SANT): clinicopathological study of 10 cases with or with out abdominal disseminated calcifying fibrous tumors, and the presence of a significant number of IgG4+ plasma cells. *Pathol Int.* 2009;59:844–850.
- Kamisawa T, Funata N, Hayashi Y, et al. A new clinicopathological entity of IgG4-related autoimmune disease. J Gastroenterol. 2003;38:982–984.
- 4. Falk GA, Nooli NP, Morris-Stiff G, et al. Sclerosing angiomatoid nodular transformation (SANT) of the spleen: case report and review of the literature. *Int J Surg Case Rep.* 2012;3:492–500.
- Koreishi AF, Saenz AJ, Fleming SE, et al. Sclerosing angiomatoid nodular transformation (SANT) of the spleen: a report of 3 cases. *Int J Surg Pathol*. 2009;17:384–389.
- Lee HJ, Choi SY, Huang SM, et al. Sclerosing angiomatoid nodular transformation (SANT) in spleen: a case report. Korean J Pathol. 2011;45:111–114.
- 7. Feng YM, Huang YC, Tu CW, et al. Distinctive PET/CT features of splenic SANT. Clin Nucl Med. 2013;38:e465-e466.
- Subhawong TK, Subhawong AP, Kamel I. Sclerosing angiomatoid nodular transformation of the spleen: multimodality imaging findings and pathologic correlate. J Comput Assist Tomogr. 2010;34:206–209.

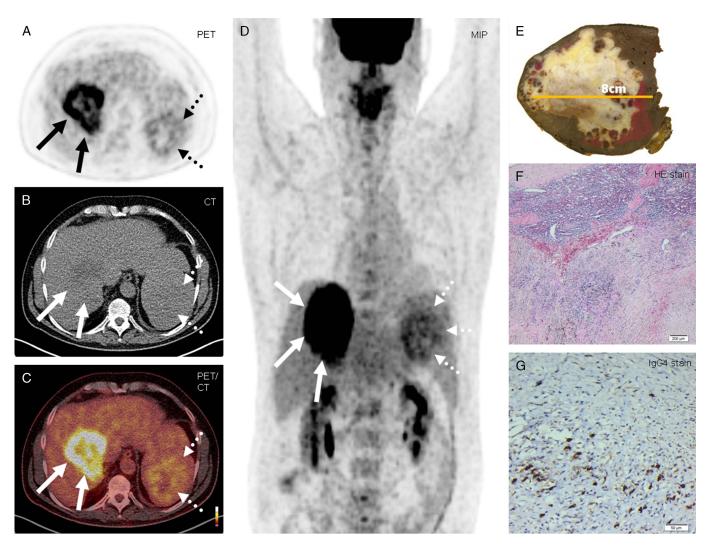


FIGURE 1. A 51-year-old man with the primary diagnosis of histologically proven cholangiocarcinoma (CCC) was referred for completion of staging and exclusion of distant metastasis. Contrast-enhanced CT revealed an additional, centrally hypodense splenic lesion measuring 8 cm in diameter, which could not be further specified. Contrast-enhanced ultrasound depicting inhomogeneous contrast media uptake of the splenic lesion and more rapid washout than of the surrounding tissue did not allow any further differentiation either. 18F-FDG PET/CT revealed a highly hypermetabolic (SUV_{max}, 19.4), centrally necrotic hepatic lesion, consistent with the previously confirmed CCC (A-D, arrows). However, the splenic lesion showed only a moderate peripheral FDG uptake (SUV_{max}, 5.4). With regards to the high FDG-avidity of the primary tumor, the uptake pattern of the splenic lesion made a metastasis unlikely (A–D, dotted arrows). Because potential differential diagnoses comprised hamartoma, hemangioma, inflammatory pseudotumor, and—though less likely—lymphoma, the patient underwent splenectomy. Histopathology confirmed a highly vascularized solid tumor with stromal sclerosis (E and F) and the presence of IgG4-positive plasma cells (G), consistent with sclerosing angiomatoid nodular transformation of the spleen. Sclerosing angiomatoid nodular transformation is a rare benign disorder of the spleen with no recurrence or malignant behavior. It usually affects middle-aged adults and is typically found incidentally. If symptomatic, most patients present with abdominal pain. Microscopically, tumors consist of isolated or confluent nodules of variable sizes separated by different degrees of internodular stromal sclerosis, which could be related to high numbers of infiltrating IgG4+ plasma cells. Because malignant diseases cannot be ruled out by imaging studies, the final diagnosis is based on surgical histopathology. 1-8 Consecutively, liver surgery and additional radiother apy for treatment of the CCC were performed.