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## Case Report

# Nested stromal epithelial tumor of the liver: A multidisciplinary approach to the treatment of an extremely rare malignancy ☆☆☆★

Ryan Mann, MD\*, Scott Kristenson, MD, Charles Kitley, MD, Kendra Thoren, DO

Madigan Army Medical Center, 9040A Jackson Avenue, Tacoma WA, 98431, USA

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## ABSTRACT

Nested stromal epithelial tumor (NSET) of the liver is an extremely rare and unusual liver neoplasm with limited evidence of best practice for management. We report a 28-year-old male with NSET managed with primary partial hepatectomy with subsequent disease recurrence with follow-up metastectomy and successful radiofrequency ablation (RFA). Management of NSET of the liver requires a multidisciplinary approach. RFA proves beneficial in a patient with disease recurrence following tumor resection. In order to validate the regular use of RFA, more long term studies would be required.

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## Introduction

Nested stromal-epithelial tumors of the liver are extremely rare and unusual primary hepatic neoplasms, with less than 20 cases reported in the literature [1–3]. Pathologic features include well demarcated nests of spindle and epithelial cells, myofibroblastic stroma, and intralesional calcification [3]. We report a case of a young male who underwent right partial hepatectomy of a primary tumor with subsequent episodes of disease recurrence necessitating additional hepatic wedge resection and finally radiofrequency ablation. Details about the

clinical, radiographic, and histopathologic features of the tumor are reported.

## Case report

A 28-year-old male was referred to general surgery following approximately 2-months of vague upper abdominal discomfort, constipation, and intermittent sharp right upper quadrant pain. On clinical exam the patient denied risk factors for hepatitis C, fevers, chills, malaise, or fatigue. Physical exam

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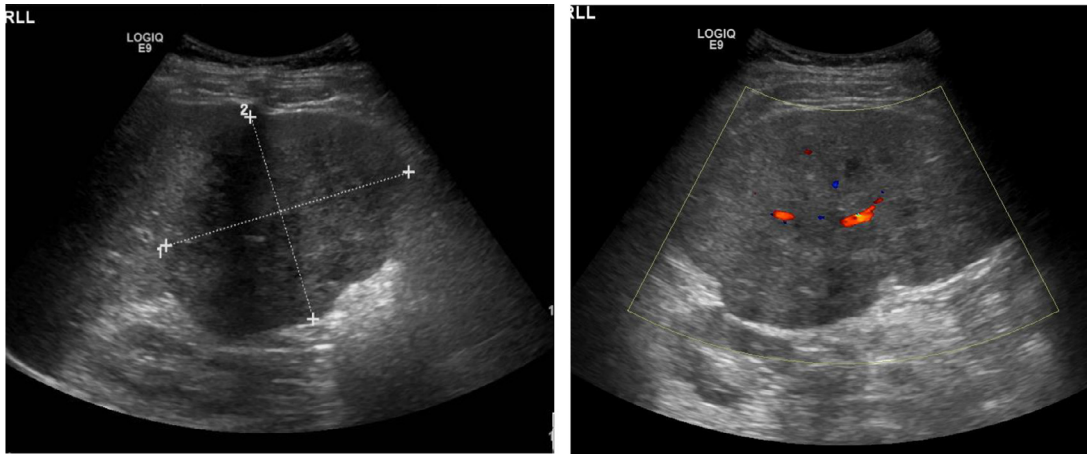
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\* Corresponding author.

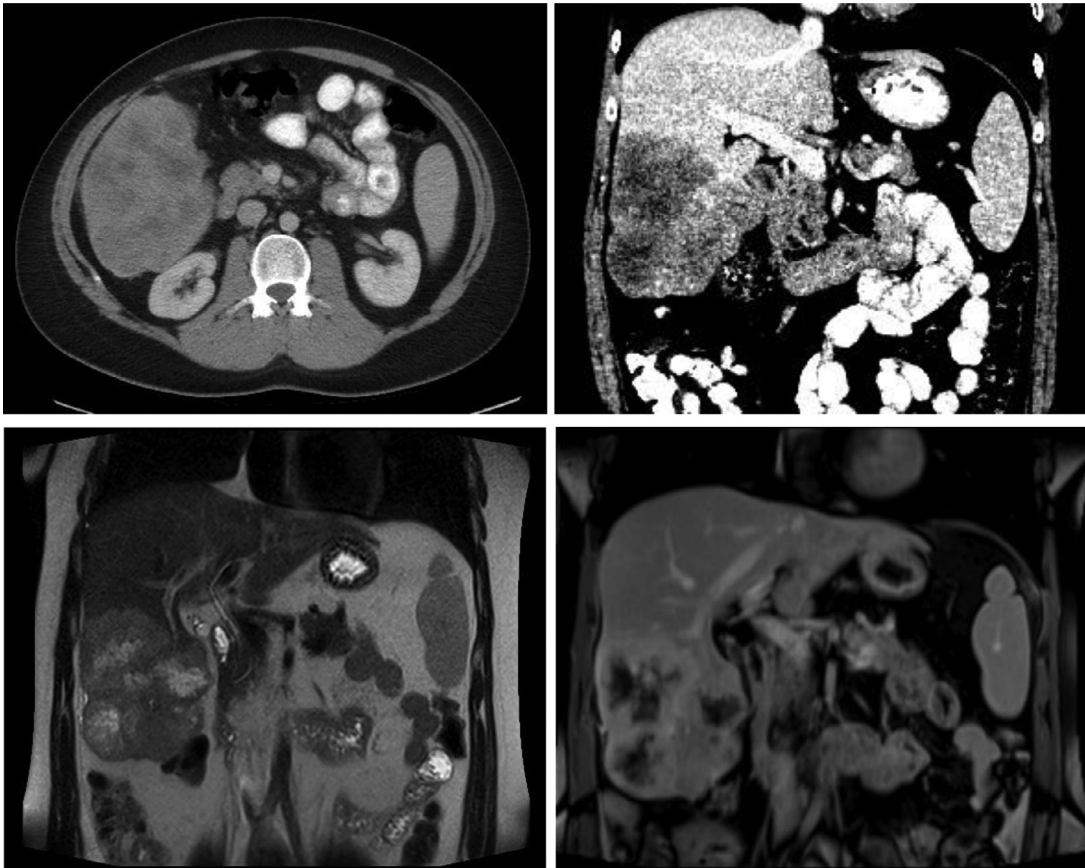
E-mail address: [ryan.d.mann6.mil@mail.mil](mailto:ryan.d.mann6.mil@mail.mil) (R. Mann).

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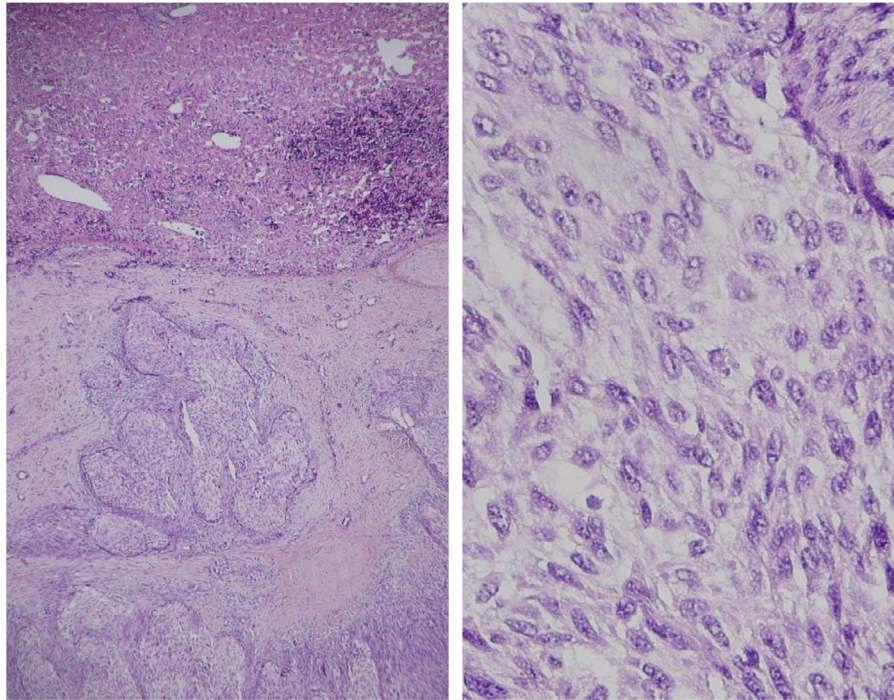
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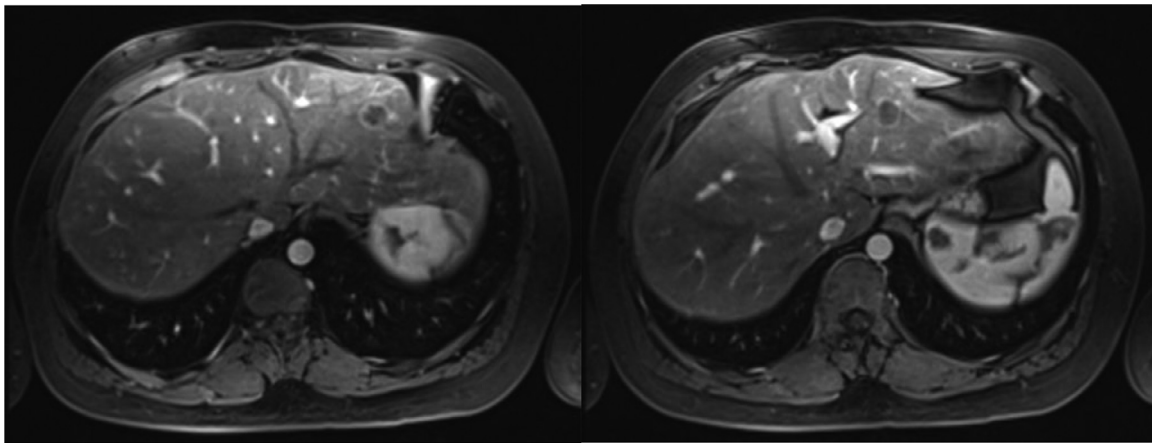
**Fig. 1 –** Grayscale and color Doppler images demonstrates a well circumscribed predominately hypoechoic mass with internal vascularity in the right lobe of the liver.



**Fig. 2 –** (A) Axial and (B) coronal contrast enhanced CT demonstrates a heterogeneously enhancing partially exophytic mass. (C) Coronal T2 weighted image demonstrates a heterogeneous T2 signal with more hyperintense areas centrally favored to represent cystic necrosis. (D) Contrast enhanced Coronal T1 weighted fat saturation MRI shows peripheral enhancement with central hypo-intensities.



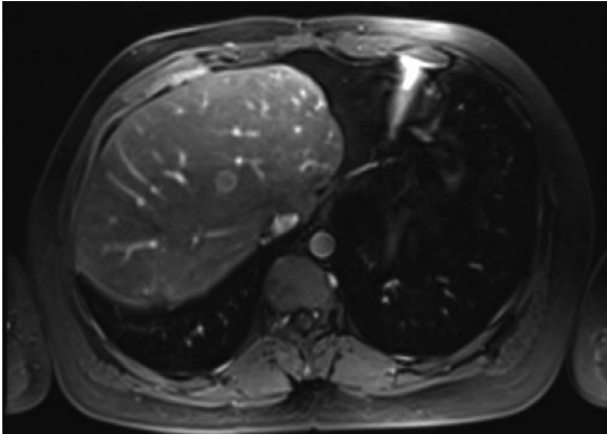
**Fig. 3 – (A) Tumor liver interface demonstrates well demarcated nests of epithelioid cells in a desmoplastic stroma. (B) Pale spindle cells demonstrating vesicular nuclei with mild atypia and some irregular mitosis.**



**Fig. 4 – Axial T1 FS postcontrast MRI demonstrated multiple peripherally enhancing metastatic lesions throughout the left and right liver.**

revealed hepatomegaly, with the liver edge measuring 2 cm below the costal margin. Initial imaging workup involved a right upper quadrant ultrasound (US) which revealed a 13 cm well circumscribed partially exophytic mass within the right lobe of the liver. Doppler US showed some internal vascularity. Subsequent MR and CT imaging confirmed a heterogeneous and lobulated soft tissue mass displaying peripheral rim like enhancement. Areas of internal hypo-enhancement were favored to represent tumor necrosis. US guided percutaneous needle core biopsy revealed a mitotically-active spindle cell neoplasm.

Given the lack of additional lesions on imaging and large exophytic size of the tumor, the patient underwent partial right hepatectomy. On laparotomy a well circumscribed firm tumor was discovered within the right lobe of the liver without evidence of extracapsular or intravascular extension. An uncomplicated partial right hepatectomy was successful and all surgical margins were tumor free. Specimens were sent out for expert consultation and returned with a diagnosis of nested stromal-epithelial tumor. Adjuvant chemotherapy was not warranted and the patient began regular surveillance to monitor for disease recurrence.



**Fig. 5 – Axial T1 FS post contrast MRI 6 months following metastectomy shows a round peripherally enhancing lesion within the right lobe of the liver consistent with a new metastatic focus.**

Approximately 8 months later, surveillance MRI demonstrated a new focus of enhancement along the peritoneum adjacent to the surgical margin. The case was presented at a liver multidisciplinary conference and the patient was referred for second opinion with the possibility of targeted genotyping and systemic therapy. This was unsuccessful and the patient underwent repeat hepatic metastectomy of up to 6 new lesions within the right and left hepatic lobes.

Surveillance MRI performed 6-months later demonstrated three new enhancing lesions which restricted diffusion suspected to represent disease recurrence. Given the location of the new lesions within the remaining liver and numerous prior surgical interventions leading to decreased liver volume and compromised function, further operative treatment was contra-indicated. Interventional radiology was then consulted for percutaneous ablation. The patient underwent successful

CT guided percutaneous radiofrequency ablation of all 3 lesions.

### Follow-up

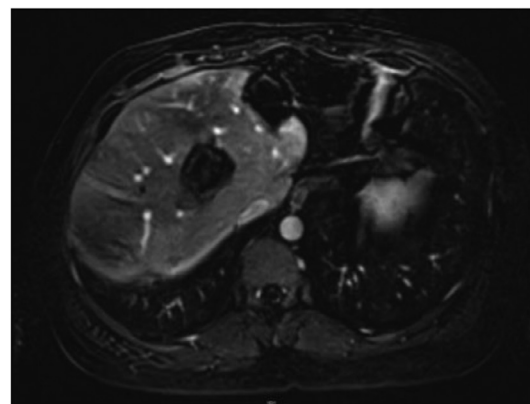
Although the initial postablation liver MRI was without evidence of disease recurrence, the immediate next MRI showed multiple enhancing foci near the prior ablation sites consistent with recurrence. The patient has since undergone 5 rounds of liver sparing CT guided ablation, using both cryo- and radiofrequency ablation.

### Discussion

Nested stromal-epithelial tumors of the liver are histologically nonhepatocytic, nonbiliary tumors demonstrating nests of epithelial and spindle cells with myofibroblastic stroma and calcification. Nested stromal epithelial tumors of the liver are extremely rare with limited information about clinical workup, prognosis, or outcomes.

Diagnosis based on imaging features alone is not possible at this time given limited number of cases and relatively non-specific findings. However, from an imaging standpoint our case is similar to others given findings as described within the figures to include hypoechoic echotexture, internal vascularity, and avid homogenous peripheral enhancement with central hypoenhancement on CT and MRI. Contrast enhanced US was not performed in the workup of our patient and there have been no reported cases that have used CE-US. This will, however, likely play an important role in future diagnoses of this tumor and its frequent recurrence. Multiple studies have demonstrated the usefulness of CE-US in characterizing focal liver lesions [7].

The differential diagnosis for the imaging features as described in our patient would include fibrolamellar hepatocel-



**Fig. 6 – (A) CT guided radiofrequency ablation using expandable electrodes with tines deployed. (B) Follow-up Contrast enhanced axial T1 weighted MR performed 6 weeks later shows a large field defect within the right lobe of the liver consistent with post-ablative changes.**

lular carcinoma, intrahepatic cholangiocarcinoma, or metastasis, all of which were considered at initial interpretation. Lymphoma could be considered, however, the patient's age would argue against this. One hallmark that our case did not demonstrate was punctate intralesional calcifications which has been described with multiple prior cases [1-6].

Current data suggests that these are low-grade tumors with an indolent course [3]. Our case is similar to prior cases in demonstrating the highly recurrent nature of the disease and need for long term follow-up [4]. Disease recurrence in our case was manifested by multiple enlarging peripherally enhancing lesions on surveillance MRI.

Wedge resection and partial hepatectomy are currently standard of care [5]. Some tumors have been treated successfully with chemotherapy; however, no preferred regimen has been adopted. Our case is the first to explore the use of percutaneous ablative techniques for this entity with relative short term success. Furthermore, it demonstrates the importance of a multidisciplinary approach for monitoring and managing this unusual and rare tumor (Figs. 1-6).

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