

Hepatic Solitary Fibrous Tumors associated with recurrent hypoglycemia: A Case Report of Doege-Potter Syndrome

Simar Goyal, BA; Anna R Shajee, MD; Charles Neuwirth; Shankar Logarajah, MD; Anish Jomy, MD; Rohan D Jeyarajah, MD

Introduction

Solitary fibrous tumors (SFTs) are rare mesenchymal neoplasms with an incidence of approximately 1 case per million annually. They can arise in many anatomic sites, but hepatic SFTs are extremely uncommon, with only 117 reported cases in the literature.

Clinical and radiologic findings are often nonspecific, and diagnosis relies on histopathology and immunohistochemistry, particularly STAT6 nuclear positivity.

A small subset of SFTs are associated with Doege–Potter syndrome, a rare paraneoplastic condition characterized by non-islet cell tumor hypoglycemia due to tumor secretion of insulin-like growth factor II (IGF-II).

Case Presentation

- ❖ 64-year-old woman presented with 18 months of progressive abdominal distension, nausea, fatigue, and 40–50 lb weight loss. Examination revealed marked abdominal distension and tenderness.
- ❖ CT imaging demonstrated two large heterogeneous hepatic masses measuring $17.9 \times 20 \times 22$ cm and $21.7 \times 16.2 \times 21$ cm. The masses were causing **mass effect** on the stomach, pancreas, and inferior vena cava.
- ❖ Biopsy demonstrated a **spindle cell neoplasm** positive for **STAT6** and **vimentin** with **Ki-67 5–10%**, consistent with **solitary fibrous tumor**.
- ❖ The patient developed **severe hypoglycemia (40 mg/dL)** despite no insulin or oral hypoglycemic use.
- ❖ Laboratory evaluation showed:
 - ❖ Insulin <1.0 uIU/mL
 - ❖ C-peptide 0.1 ng/mL
 - ❖ **IGF-II/IGF-I ratio 8:1**

These findings supported non-islet cell tumor hypoglycemia consistent with Doege-Potter syndrome.

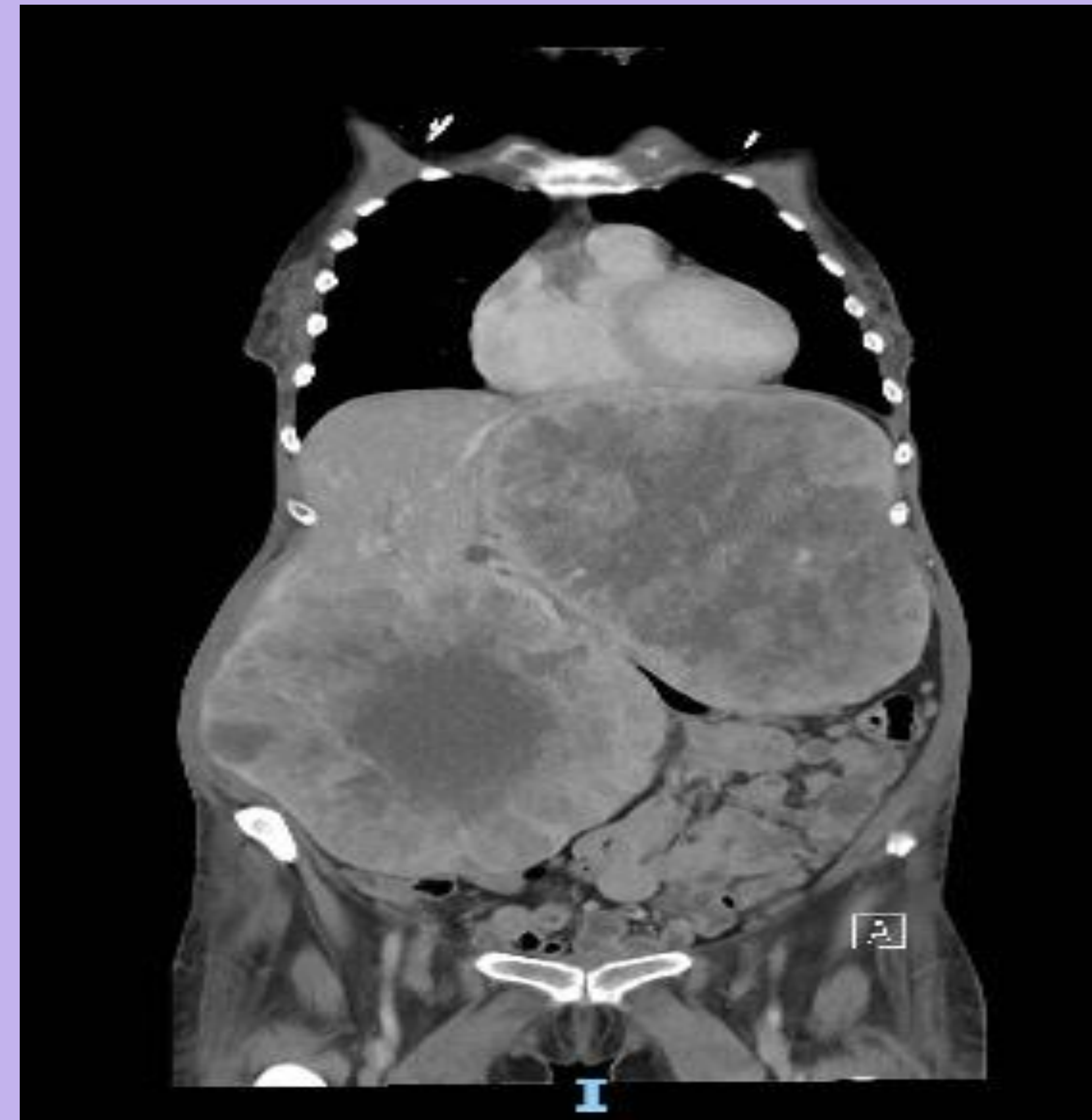


Figure 1: Coronal CT demonstrating two large hepatic masses with heterogenous enhancement

Operative Course

- ❖ Patient underwent **open resection of the hepatic tumors** through a bilateral subcostal incision
- ❖ Tumor removal required **left trisegmentectomy (segments II, III, IV, V, and VIII)**.
- ❖ The operation was complicated by massive hemorrhage, requiring:
 - ❖ 46 units PRBC
 - ❖ 41 units FFP
 - ❖ 3 units platelets
 - ❖ 3 units cryoprecipitate
 - ❖ 2 units whole blood
- ❖ Estimated blood loss was approximately **50 liters**.
- ❖ Temporary abdominal packing with vacuum assisted closure was performed prior to ICU transfer.

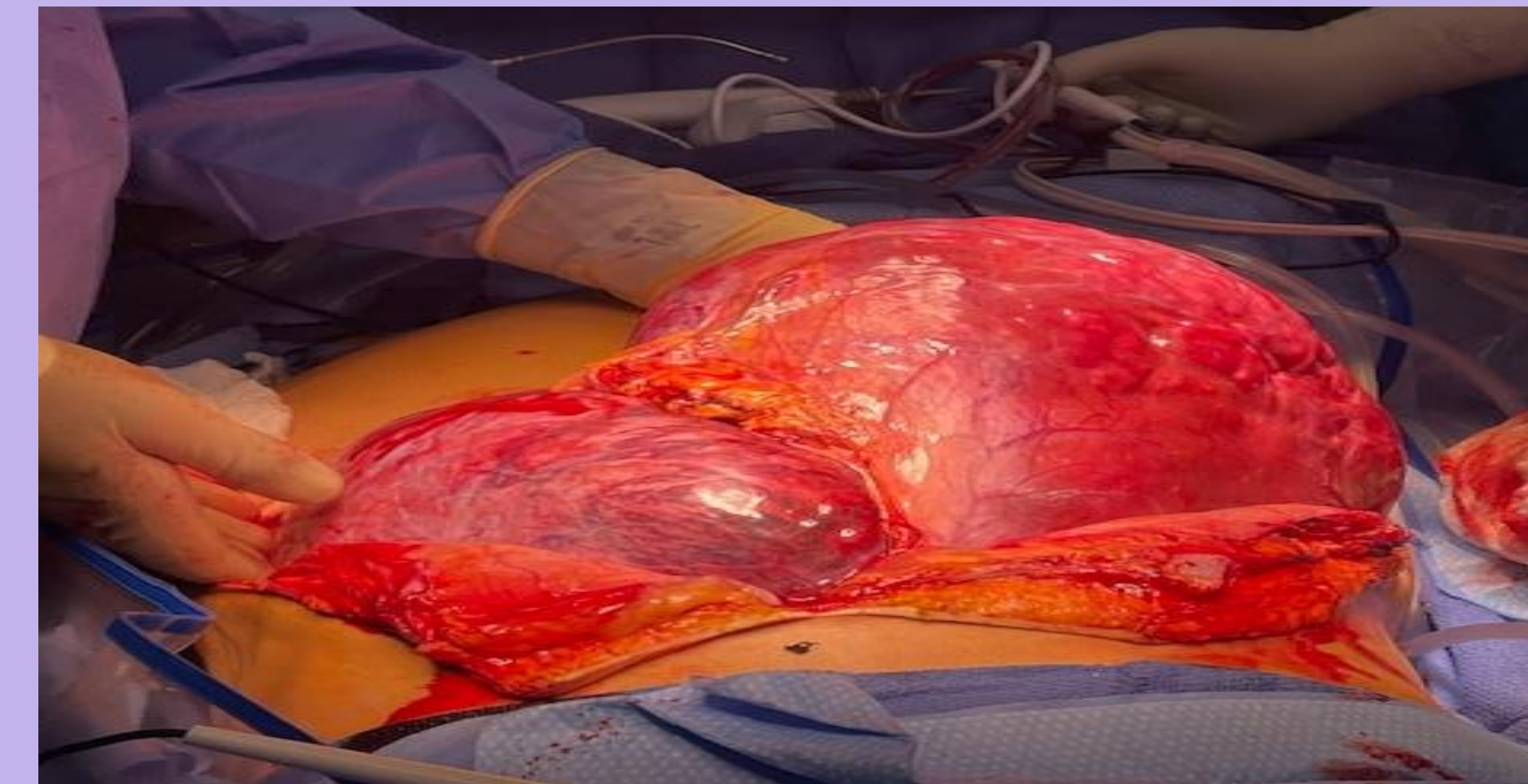


Figure 2a-b: Intraoperative view (top) and resected specimens (bottom) of hepatic solitary fibrous tumors

Post-Operative Course

- ❖ Returned to the operating room on **postoperative day 1** for washout and definitive abdominal closure
- ❖ **Extubated on postoperative day 2** with normalization of glucose and coagulation parameters
- ❖ Hospital course complicated by a **postoperative intra-abdominal abscess**, successfully treated with **IR-guided drainage and antibiotics**.
- ❖ Discharged **postoperative day 9** with resolution of hypoglycemia and improvement in presenting symptoms.

Discussion

- ❖ Hepatic SFTs are rare tumors that often present with nonspecific symptoms, making diagnosis challenging. Histopathologic confirmation with STAT6 positivity is essential
- ❖ Less than 5% of solitary fibrous tumors are associated with Doege-Potter syndrome, a paraneoplastic syndrome in which tumor secretion of IGF-II results in recurrent hypoglycemia
- ❖ An elevated **IGF-II/IGF-I ratio ($>3:1$)** is the most reliable biomarker supporting the diagnosis of Doege-Potter syndrome
- ❖ Complete surgical resection remains the cornerstone of treatment and is often curative, particularly for tumors associated with hypoglycemia.

References

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