ACG CASE REPORTS JOURNAL

CASE REPORT | PANCREAS

Unusual Case of a Pancreatic Neuroendocrine Tumor Containing a Central Scar

Binit Sureka, MD, DNB, MBA-HCA, MNAMS, PGDHA[1], Vaibhav Varshney, MS, MCh[2], Poonam Elhence, MD[3], Jyotsna Bharti, MD[3], Taruna Yadav, MD, PDCC[4], Pawan Kumar Garg, MD[4], and Pushpinder Singh Khera, MD, FRCR[4]

[1]*Department of Radiology, All India Institute of Medical Sciences (AIIMS), Jodhpur, Rajasthan*
[2]*Department of Surgical Gastroenterology, All India Institute of Medical Sciences (AIIMS), Jodhpur, Rajasthan*
[3]*Department of Pathology, All India Institute of Medical Sciences (AIIMS), Jodhpur, Rajasthan, India*
[4]*Department of Radiology, All India Institute of Medical Sciences (AIIMS), Jodhpur, Rajasthan*

ABSTRACT

We report a previously unreported case of a pancreatic neuroendocrine tumor with a central scar mimicking a serous neoplasm. To our knowledge, this atypical imaging morphology of pancreatic neuroendocrine tumor has not been described before. Our report adds to the body literature that describes atypical imaging variants of neuroendocrine tumors and highlights that clinicians should be aware of the broad imaging characteristics of neuroendocrine tumors.

INTRODUCTION

Pancreatic neuroendocrine tumors (PanNETs) represent a considerable diagnostic challenge because of varied clinical presentation and imaging features and comprises about 2%–10% of all pancreatic tumors. According to recent studies, up to 40% of PanNETs may not show typical arterial hyperenhancment. Atypical imaging features of PanNETs are purely cystic, solid-cystic, calcified variety, and diffuse forms. We highlight a very rare radiological presentation of a PanNET with a central scar mimicking a serous neoplasm. This radiological pattern has not been described previously in the literature.

CASE REPORT

A 23-year-old man presented with complains of pain in the central abdomen for the past 10 days. The pain was mild to moderate in intensity, dull aching in nature, and nonradiating. No history of jaundice, fever, anorexia, weight loss, diabetes, or alcohol abuse was present. The patient also did not undergo any kind of major surgery in the past. Physical examination revealed mild abdominal tenderness at the epigastrium. His hemoglobin was 16 g/dL. Anti-hemoglobin core was nonreactive, and hepatitis C virus RNA was not detected in the plasma. Liver and kidney function tests were normal. Carbohydrate antigen 19–9 was 23.13 U/mL and was carcinoembryonic antigen was 1.02 ng/mL.

Contrast-enhanced computed tomography showed a low-attenuation minimally enhancing lesion in the body-neck junction of the pancreas. Upper abdominal ultrasound revealed a hypoechoic lesion within the pancreas without evidence of internal vascularity. Contrast-enhanced magnetic resonance imaging (MRI) showed a mass lesion of size 4.2 × 3.5 × 3.5 cm, which was hypointense on T1WI and mildly hyperintense on T2W magnetic resonance images. The mass lesion had a central T2 hyperintense scar. On postcontrast magnetic resonance images, the periphery of the lesion showed minimal enhancement. The central portion of the lesion showed a spoke-wheel type of delayed enhancing scar (Figure 1). The upstream main pancreatic duct and common bile duct were not dilated. Based on the MRI, a provisional diagnosis of solid serous adenoma with a central scar was made. Endoscopic ultrasound or biopsy was not performed because it would not change the management.

ACG Case Rep J 2019;6:1–5. doi:10.14309/crj.0000000000000047. Published online: March 18, 2019

Correspondence: Binit Sureka, MD, DNB, MBA-HCA, MNAMS, PGDHA, Department of Diagnostic & Interventional Radiology, All India Institute of Medical Sciences (AIIMS), Jodhpur, Rajasthan 342005, India (surekab@aiimsjodhpur.edu.in).

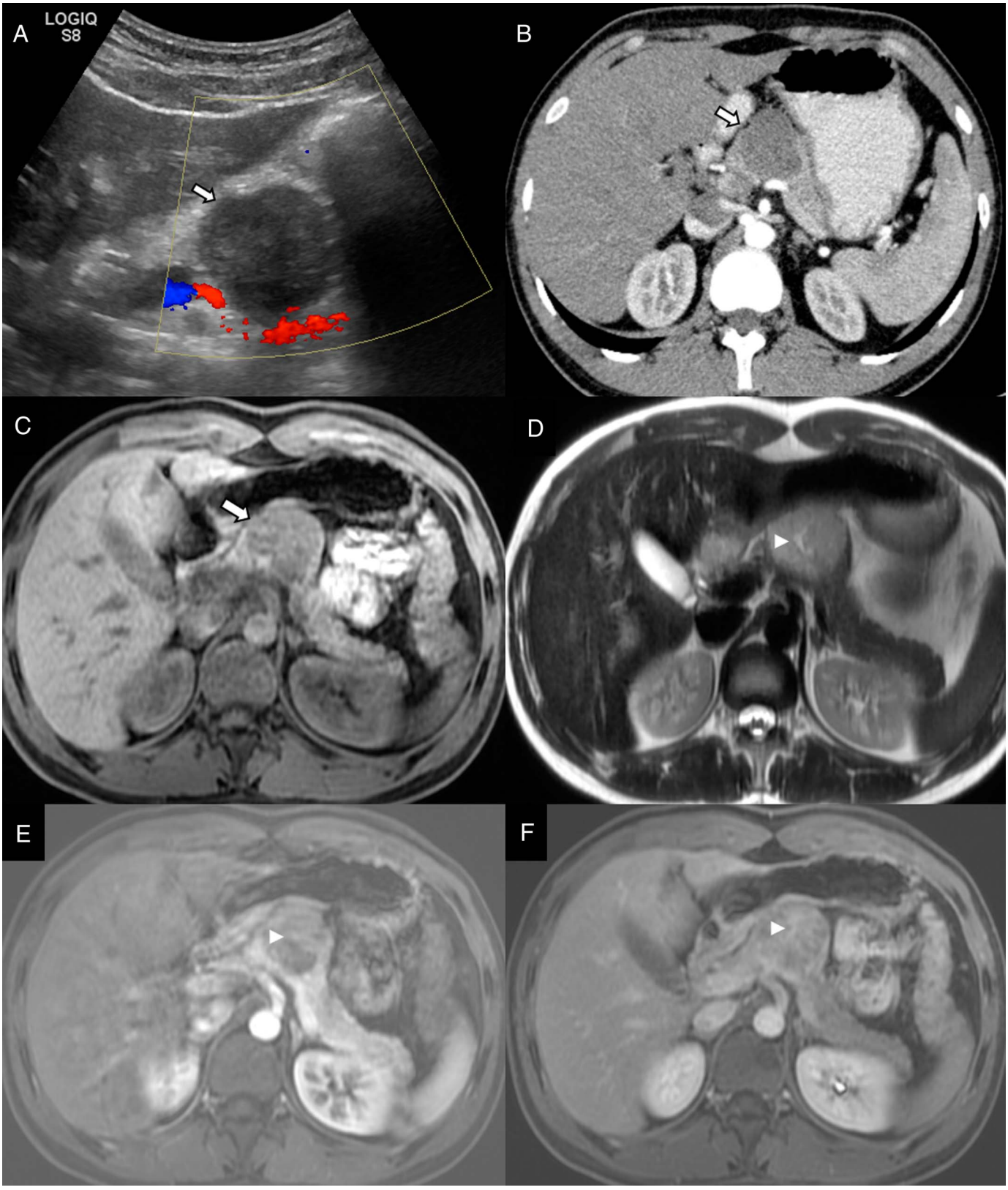

Figure 1. (A) Transverse ultrasound image showing predominantly hypoechoic solid mass lesion (arrow) in the neck of the pancreas without internal vascularity. (B) Axial contrast-enhanced computed tomography arterial phase showing hypoenhancing hypoattenuating lesion (arrow) in the neck of pancreas. (C) Axial T1-weighted magnetic resonance image depicting hypointense lesion (arrow) in the neck of the pancreas. (D) T2-weighted magnetic resonance image showing mildly hyperintense lesion with a central hyperintense scar (arrowhead), (E) minimal enhancement in arterial phase, and (F) gradual delayed enhancement of the central scar (arrowhead) with radiating septae.

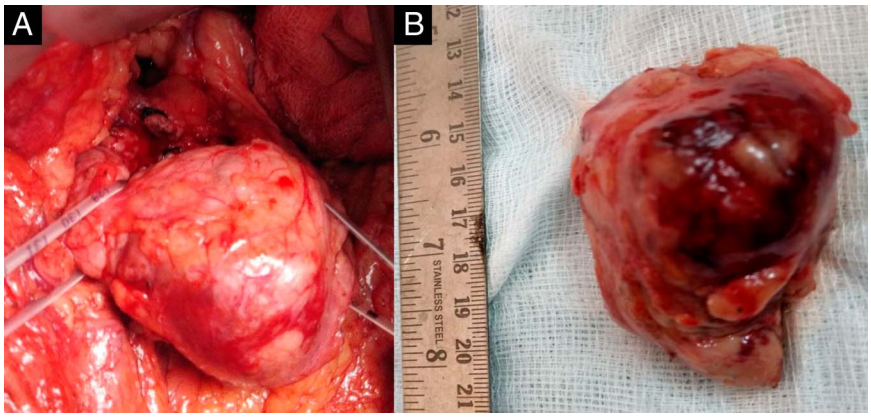

Figure 2. (A) Intraoperative and (B) postoperative surgical specimen.

The patient was advised surgery and underwent median pancreatectomy with pancreatojejunostomy. On gross examination, the cut section of the tumor was homogeneous, gray-white, and firm (Figure 2). Microscopic examination showed tumor arranged in solid nests with trabeculas of small to medium cells separated by the eosinophilic hyaline material. The tumor cells showed monomorphic round nuclei, salt-and-pepper chromatin, and scanty to moderate amounts of cytoplasm.

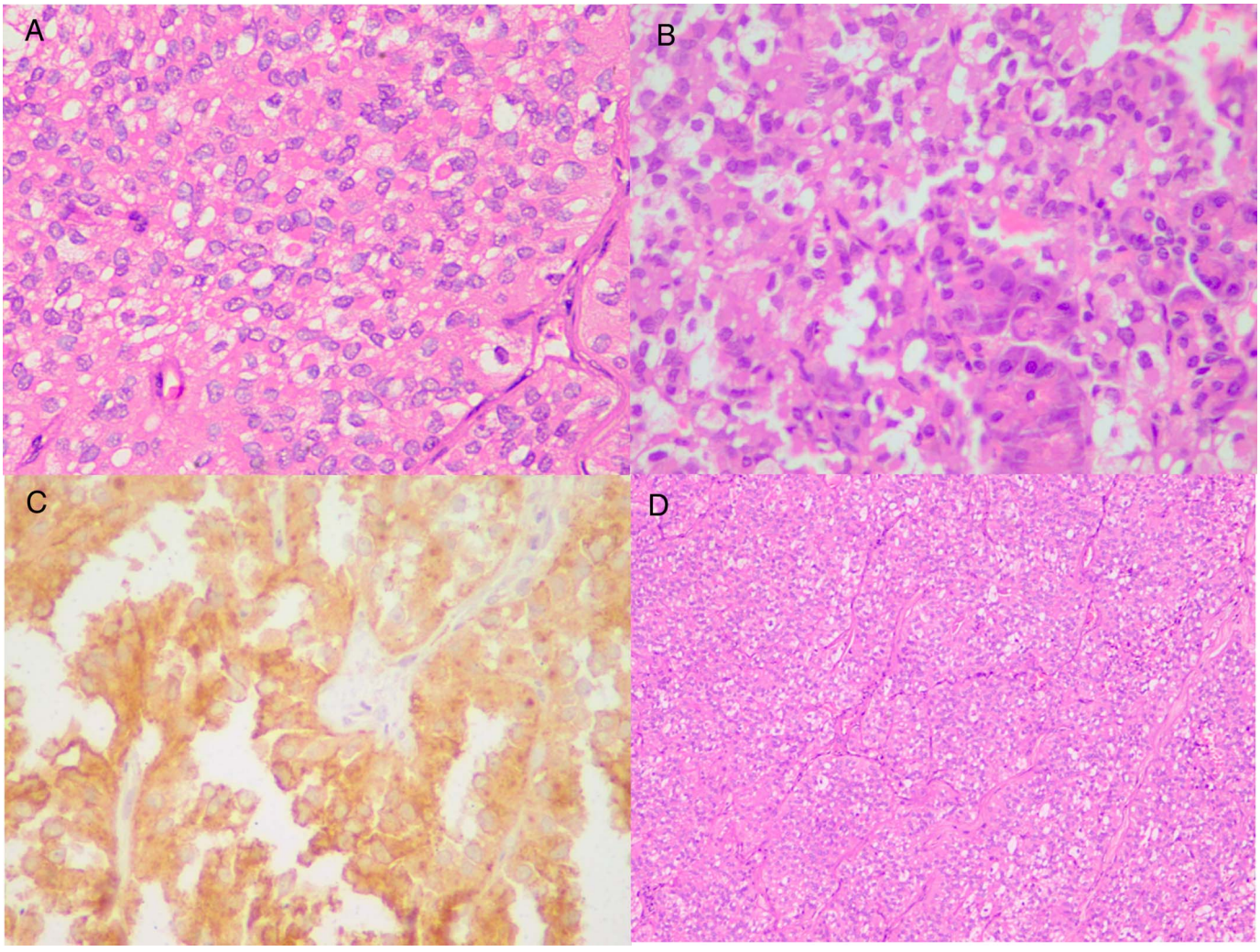

Figure 3. (A) Well-differentiated neuroendocrine tumor, nests, and few cytoplasmic hyaline globules seen, 40×, (B) tumor nests infiltrating the pancreatic acini, 40×, (C) immunohistochemistry positive for synaptophysin, 40×, and (D) tumor nests divided by hyalinized fibrous septae, 10×.

Mitosis was frequent (<2/10 hpf). No areas of necrosis/vascular and perineural invasion were seen. On immunohistochemistry, tumor cells are reactive for synaptophysin, chromogranin, and cytokeratin. The Ki67 was <2%. The final diagnosis was well-differentiated neuroendocrine tumor (grade I) (Figure 3). At one-year follow up, the patient is asymptomatic and doing well.

DISCUSSION

PanNETs account for less than 3% of all pancreatic neoplasms.[1] They can be broadly divided into the following 2 types: nonfunctioning and functioning neoplasms. Nonfunctioning neoplasms are more common than functioning neoplasms.[2] Currently, available tools for the detection of PanNETs are divided into three categories—anatomical (CT, MRI, and ultrasound (US)), functional (scintigraphy and positron emission tomography (PET)—^{68}Ga-DOTATOC, ^{68}Ga-DOTANOC, and ^{68}Ga-DOTATATE), and hybrid imaging (PET/CT, single-photon emission computed tomography/CT, and PET/MRI). Anatomical imaging tools such as CT and MRI are the initial investigations of choice. Functional tools are used when the anatomical tools fail to detect PanNETs despite a strong clinical suspicion. When there is a suspicion of a high-grade PanNET, ^{18}F-fluorodeoxyglucose is preferred for tumor detection because the somatostatin receptor expression of these tumors is low. Radiologically, classical PanNETs are solitary 1–5 cm sized well-circumscribed solid lesions. These lesions show avid enhancement in arterial and venous phase CT because of rich intralesional vascularity.[3] On MRI, most functioning PanNETs are hypointense on T1W and hyperintense on T2W images and show intense and early enhancement on dynamic T1W sequence after contrast injection.

Various atypical patterns of PanNETs have been described in the literature. Atypical patterns include pure cystic variety in 10% cases (usually seen in multiple endocrine neoplasia type 1), and complex solid-cystic and calcified variety in less than 5% cases.[4] When cystic, these lesions may show intense thick peripheral arterial enhancing rim. According to recent studies, up to 41.5% of PanNETs may not show arterial hyperenhancment and these hypovascular PanNETs may be difficult to differentiate from hypovascular pancreatic ductal adenocarcinomas.[5–7] The presence of well-defined margins, the lack of upstream pancreatic atrophy or ductal dilatation, and progressive and persistent enhancement in the portal and delayed phase are the imaging clues to differentiate these atypical tumors from pancreatic adenocarcinomas that show ill-defined margins, pancreatic atrophy, upstream ductal dilatation, and persistent hypoenhancement in all phases or gradual delayed enhancement.[7] Rarely, PanNET may present as diffuse infiltrative variety in which the entire pancreatic tissue is enlarged and replaced by calcifications and cysts.[8]

PanNET with a central scar has not been previously reported in the literature. The differential diagnoses of atypical PanNETs include serous cystadenoma, focal mass-forming pancreatitis, solid serous adenoma, metastases from renal cell carcinoma, acinar cell carcinoma, solid pseudopapillary tumor, intraductal papillary mucinous neoplasm with mural nodule, and pancreatoblastoma in children.[9,10] According to previously published case reports, solid serous adenoma is less than 3 cm in size, can have a solid-like appearance on imaging although it belongs to the serous cystadenoma group, has well-defined borders, and shows strong arterial phase enhancement.[11] Preoperatively, we mislabeled this tumor as solid serous adenoma because of the central scar. The hypothesis behind this atypical enhancement pattern as seen in our case could be because of the presence of higher fibrotic and less cellular component within the tumor. Radiological diagnosis of solid serous adenoma is difficult because it cannot be distinguished from other solid tumors because of its radiologic characteristics which are similar to those of a solid tumor and do not distinguish it as a cystic tumor. Radiologic images such as those of CT and MRI are not diagnostic, and even endoscopic ultrasound fine-needle aspiration can fail to differentiate solid serous adenoma from a neuroendocrine tumor.

DISCLOSURES

Author contributions: B. Sureka prepared the manuscript and is the article guarantor. V. Varshney conceptualized the manuscript. P. Elhence and J. Bharti analyzed the data and reviewed the manuscript. T. Yadav and PK Garg edited the manuscript. PS Khera edited the manuscript and reviewed the literature.

Financial disclosure: None to report.

Informed consent was obtained for this case report.

Received August 10, 2018; Accepted January 11, 2019

REFERENCES

1. Metz DC, Jensen RT. Gastrointestinal neuroendocrine tumors: Pancreatic endocrine tumors. *Gastroenterology*. 2008;135:1469–92.
2. Dromain C, Déandréis D, Scoazec JY, et al. Imaging of neuroendocrine tumors of the pancreas. *Diagn Interv Imaging*. 2016;97:1241–57.
3. Lee NJ, Hruban RH, Fishman EK. Pancreatic neuroendocrine tumor: Review of heterogeneous spectrum of CT appearance. *Abdom Radiol (NY)*. 2018;43:3025–34.
4. Gallotti A, Johnston RP, Bonaffini PA, et al. Incidental neuroendocrinetumors of the pancreas: MDCT findings and features of malignancy. *AJR Am J Roentgenol*. 2013;200:355–62.
5. Manfredi R, Bonatti M, Mantovani W, et al. Non-hyperfunctioning neuroendocrine tumours of the pancreas: MR imaging appearance and correlation with their biological behaviour. *Eur Radiol*. 2013;23:3029–39.
6. Humphrey PE, Alessandrino F, Bellizzi AM, Mortele KJ. Non-hyperfunctioning pancreatic endocrine tumors: Multimodality imaging features with histopathological correlation. *Abdom Imaging*. 2015;40:2398–410.
7. Jeon SK, Lee JM, Joo I, et al. Nonhypervascular pancreatic neuroendocrine tumors: Differential diagnosis from pancreatic ductal adenocarcinomas at

MR imaging-retrospective cross-sectional study. *Radiology*. 2017;284: 77–87.
8. Singh R, Calhoun S, Shin M, Katz R. Pancreatic neuroendocrine tumor with atypical radiologic presentation. *Radiol Case Rep*. 2015;3:162.
9. Raman SP, Hruban RH, Cameron JL, Wolfgang CL, Fishman EK. Pancreatic imaging mimics: Part 2, pancreatic neuroendocrine tumors and their mimics. *AJR Am J Roentgenol*. 2012;199:309–18.
10. Javadi S, Menias CO, Korivi BR, et al. Pancreatic calcifications and calcified pancreatic masses: Pattern recognition approach on CT. *AJR Am J Roentgenol*. 2017 ;209:77–87.
11. Katsourakis A, Dimitriou I, Noussios G, Chatzis I, Chatzitheoclitos E. Solid serous adenoma of the pancreas: A case report and review of the literature. *Case Rep Surg*. 2016;2016:3730249.