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A rare case of paraganglioma of the cystic duct

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ABSTRACT

INTRODUCTION: Biliary system paragangliomas are rare neuroendocrine tumors of embryonic neural crest origin. The majority is asymptomatic and incidentally found due to gallbladder functional disorders. Herein, we present a non-functional, 2.25 mm focus in the cystic duct, which to our knowledge, is the first reported paraganglioma of the cystic duct.
PRESENTATION OF CASE: The patient presented to the Emergency Department complaining of a sudden-onset, right upper abdominal and epigastric pain. Ultrasound and Computed Tomography were both consistent with signs of early cholecystitis. Laparoscopic cholecystectomy was performed without major complications. In addition to cholelithiasis and chronic cholecystitis, pathological examination reported a neuroendocrine proliferation in the cystic duct measuring 2.25 mm favoring paraganglioma. Incidentally, the patient is unique in that they were also found to have an adrenal nodule and a normocalcemic primary hyperparathyroidism that raised suspicion for an underlying endocrinopathy. Nevertheless, genetic testing was negative.
DISCUSSION: Extensive literature review demonstrates only nine cases of gallbladder paraganglioma, and three cases of hepatic ducts paraganglioma, but no cases of paraganglioma occurring at the cystic duct. Although a gene mutation and syndrome was not identified in the patient, the fact that an adrenal nodule and normocalcemic primary hyperparathyroidism were present, suggests that a complete hormonal workup should be obtained in these patients.
CONCLUSION: It is important to realize that biliary system paragangliomas, although rare, may occur. As they have an association with multiple endocrine neoplasia syndrome, a thorough endocrine investigation should be made.

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1. Introduction

Paragangliomas are rare neuroendocrine tumors of neural crest origin that arise from chromaffin cells. Paragangliomas can be potentially found anywhere along the paravertebral axis from their predominant location at the base of the skull and neck to the pelvis [1]. Pheochromocytoma and carotid body tumor are the two most common types of paragangliomas, which occur in the adrenal medulla and at the bifurcation of the carotid artery, respectively; however, remaining paragangliomas are usually retroperitoneal in origin, and found in the sympathetic or parasympathetic ganglia [1,2]. Most paragangliomas are asymptomatic and present as a painless mass [1]. While all can potentially secrete hormones such as catecholamines due to their origin from chromaffin cells, only a small percentage of cases are clinically significant and evoke systemic symptoms [1]. Biliary system paragangliomas are predominantly seen in females in the fifth to sixth decade of life [3]. These tumors are typically discovered incidentally during gallbladder or unrelated surgery, or secondary to complications such as obstructive jaundice, right upper quadrant pain, and gastrointestinal bleeding [3]. Herein, we present a non-functional, 2.25 mm focus in the cystic duct, which to our knowledge, is the first reported paraganglioma of the cystic duct. This work has been reported in line with the SCARE criteria [4].

2. Presentation of case

A patient with a past medical history of atrial fibrillation, hypertension, and hyperlipidemia and no past surgical history walked into the Emergency Department of an academic institute complaining of a sudden-onset, sharp, right upper abdominal and epigastric pain radiating to the back. Family history was negative and social history did not include any tobacco, alcohol, or drug misuse. On physical examination, the patient was tender in the right upper quadrant. On imaging, a right upper quad-

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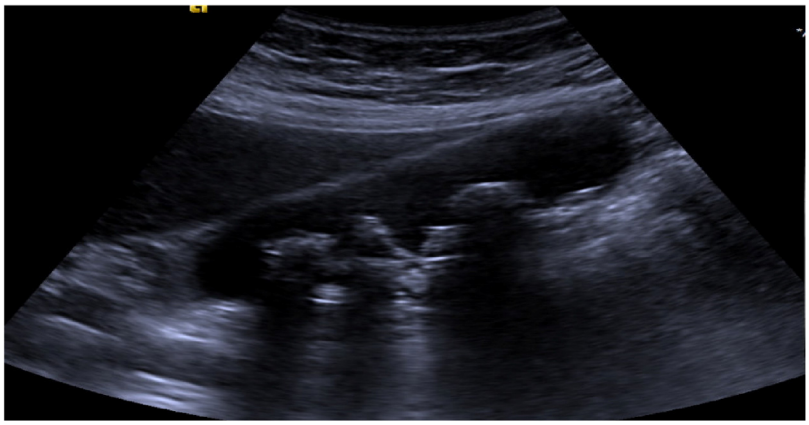

Fig. 1. Ultrasound mass showed signs of early cholecystitis.

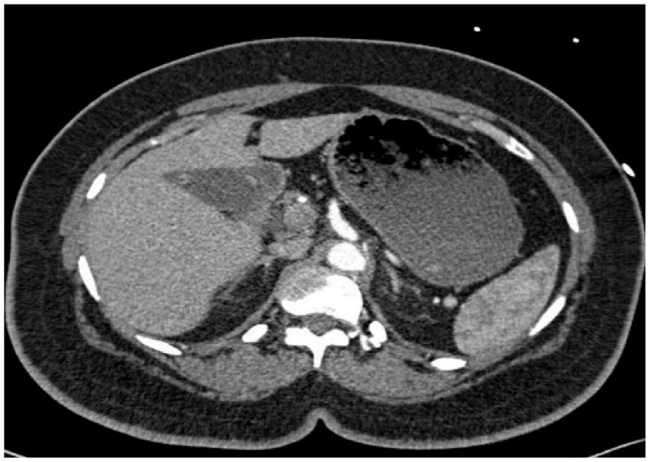

Fig. 2. CT scan at the time of original presentation of the patient.

rant ultrasound showed signs of early cholecystitis (Fig. 1) and computed tomography of the abdomen showed gallstones and distended liver bile ducts, distended gallbladder with wall thickening, edema, and a mild surrounding inflammation consistent with the ultrasound findings (Fig. 2). Anti-coagulation was held and the patient underwent an uncomplicated laparoscopic cholecystectomy two days after presentation by the general surgery team. Intra-operatively, an inflamed appearing gallbladder was noted. The patient was discharged on the second postoperative day and recovered uneventfully.

On pathology, routine hematoxylin and eosin-stained sections revealed a relatively well-circumscribed 2.25 mm lesion adjacent to the cystic duct. The lesion was composed of nests of monomorphic cells containing pale-to-eosinophilic foamy cytoplasm. The nuclei were round, with granular chromatin and lacked significant mitotic activity. No necrosis was identified. On immunohistochemical studies, the specimen showed diffuse positivity for synaptophysin and focal positivity for chromogranin, supporting neuroendocrine differentiation. SOX-10 highlighted a few cells at the periphery of the nests, consistent with sustentacular cells. In all, the morphologic and immunophenotypic findings were most consistent with an incidental paraganglioma (Fig. 3a–c).

Based on this pathology report, in the follow-up appointment a few weeks later, the patient was referred to an endocrine surgeon. The patient had blood and urine work-up including a plasma catecholamine panel showing epinephrine levels of 20 pg/mL (10–200), norepinephrine levels of 492 pg/mL (80–520), and dopamine of <20 pg/mL (0–20), all within normal levels. The patient also had

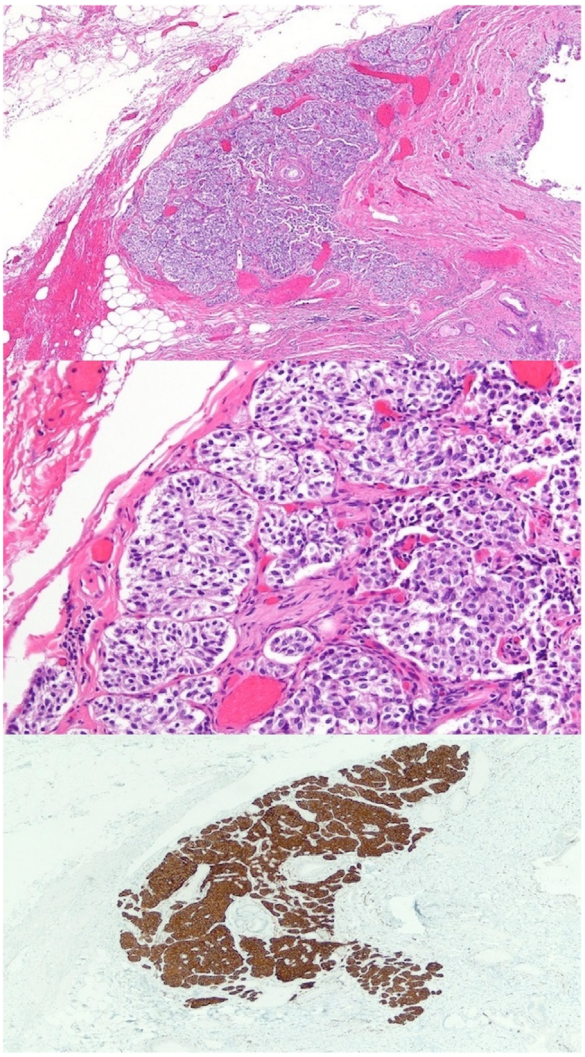

Fig. 3. (a) Low power photomicrograph of the lesion in the center of the image. Part of the cystic duct lumen is seen at the right edge of the micrograph (hematoxylin and eosin stain, original magnification ×40). (b) Medium power photomicrograph demonstrating the characteristic nested growth pattern of small, monomorphic cells that contain pale-to-eosinophilic foamy cytoplasm (hematoxylin and eosin stain, original magnification ×200). (c) Low power photomicrograph revealing diffuse and strong immunopositivity for synaptophysin (original magnification ×40), supporting the morphologic impression of neuroendocrine differentiation.

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