CASE REPORT

Large retroperitoneal paraganglioma concurrent with periampullary adenocarcinoma

Seyed Mohammadreza Hakimian, Azar Naimi[1], Seyed Mohammadhasan Emami[2], Golnar Rozatii[3], Vahid Goharian[4]
Surgeon, Fellowship in Oncosurgery at Poursina Institution, [1]Pathologist at Poursina Institution, [2]Gastroenterologist, Professor Associated at Isfahan University, Works at Poursina Institution, [3]Internist at Poursina Institution, [4]Surgeon, Fellowship in Thoracic Surgery at Poursina Institution

Paragangliomas are tumors that originate from extra-adrenal medullary neural crest derivatives. They are rarely located in retroperitoneal space. These tumors are often discovered incidentally during imaging studies performed for other reasons. Periampullary cancers include adenocarcinomas arising from the pancreas, ampulla of Vater, duodenum or distal common bile duct. The exact site of origin of periampullary tumors is often difficult to ascertain pre-operatively. We report the case of a patient who had a retroperitoneal non-functional paraganglioma, concurrent with periampullary adenocarcinoma. An 81-year-old woman was admitted with progressive abdominal fullness. There was an upper paramedian, left sided, large, palpable mass on the physical examination. Laboratory investigations showed an increase in liver enzyme levels. On abdominal computed tomography the patient found to have a large retroperitoneal mass and dilation in biliary tract, which was confirmed by magnetic resonance cholangiopancreatography. She had a tumoral papi in Endoscopic Retrograde cholangiopancreatography. Which biopsy revealed adenocarcinoma. She underwent surgery for excision of abdominal mass and pancreaticoduodenectomy. And pathologic study showed paraganglioma. This is the first ever reported case of concurrent paraganglioma and periampullary adenocarcinoma.

Key words: Paraganglioma, periampullary carcinoma, retroperitoneal mass

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INTRODUCTION

Retroperitoneal paragangliomas are rare and easily misdiagnosed. These tumors are often discovered incidentally during imaging studies performed for other reasons. Paragangliomas are tumors that originate from extra-adrenal medullary neural crest derivatives. Most of them are located in the head and neck, but can be found in various body sites, including the thoracic cavity, abdomen, pelvis, and bladder.[1]

Periampullary cancers include adenocarcinomas arising from the pancreas, ampulla of Vater, duodenum or distal common bile duct. The exact site of origin of periampullary tumors is often difficult to ascertain preoperatively.[2]

We report the case of a patient who had a retroperitoneal paraganglioma manifested as an abdominal mass with concurrent periampullary carcinoma; it is the first report of such a concurrency.

CASE REPORT

An 81-year-old woman with fit physiologic condition, was admitted to our institution in Isfahan, Iran, in 2012, because she experienced progressive abdominal fullness and dull epigastric pain for a few months. Her past medical history was unremarkable. Her blood pressure and heart rate were within normal limits and liver enzymes including serum glutamic oxaloacetic transaminase (SGOT), serum glutamic-pyruvic transaminase (SGPT), alkaline phosphatase and gamma glutamyl transferase showed increased values. Bilirubin level, Carcinoembryonic antigen and cancer antigen 19-9 levels were within normal limits.

In the abdominal examination a large mass on upper paramedian, left side was touched. A computed tomography (CT) scan of the abdomen demonstrated a large retroperitoneal solid-cystic mass and significant dilation of gallbladder, intra and extra hepatic and pancreatic ducts [Figure 1].

Magnetic resonance cholangiopancreatography of the patient showed large solid-cystic tumor anterior to left psoas muscle and left kidney, inferior to pancreatic tail. It showed either, complete obstruction of main distal pancreatic duct and distal Common Bile Duct (CBD) [Figure 2].

Address for correspondence: Dr. A. Naimi, Department of Pathology, Poursina Research Institution, Poursina Research Institution, Isfahan, Iran.
E-mail: azar.naimi@yahoo.com
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The patient underwent endoscopic retrograde cholangiopancreatography (ERCP), which revealed a tumoral papilla (2 cm × 2 cm × 1 cm) and biopsy was taken. Microscopic examination of papi biopsy showed adenocarcinoma.

She was planned for surgery with frozen section examination of abdominal mass. At the time of surgery, an encapsulated, hypervascular mass (approximately 13 cm × 10 cm × 4 cm) was observed in her retroperitoneal area with the central degeneration and hemorrhage and was subsequently excised [Figure 3].

Pathological analysis of the mass on frozen section showed a cellular lesion with probability of malignancy, without any sign of metastatic carcinoma.

Pancreaticoduodenectomy was considered for management of her periampullary mass. In permanent histologic sections, the abdominal mass revealed an encapsulated tumor; the tumor cells were arranged in sheets, large nests and organoid patterns, and contained mild pleomorphic nuclei with low mitotic activity [Figure 4]. The neoplasm was histologically benign, with no evidence of invasion, compatible with a paraganglion. The histologic evaluation of peiampullary tumor revealed an adenocarcinoma in stage IIB. After three months from patient's surgery, her treatment has been completed and patient seems to be healthy.

DISCUSSION

Pheochromocytomas and extra-adrenal paragangliomas are very rare tumors that arise from the neural crest tissue. These tumors are functional in more than half of cases, and patients commonly present with symptoms, such as palpitation, headache, and sweating that are related to excess catecholamine secretion.[1] About 70% of sympathetic paragangliomas are intra-abdominal, usually found in the perinephric and paraaortic spaces. The remaining 30% are located in the thorax. Malignant retroperitoneal paragangliomas range from 30% to 50%.[3] Paragangliomas metastasize approximately in 20-42% of the cases.[4] Our case was a non-functional retroperitoneal paraganglioma.

The non-secretory type most commonly presents as an abdominal pain or mass;[5] a large proportion of these

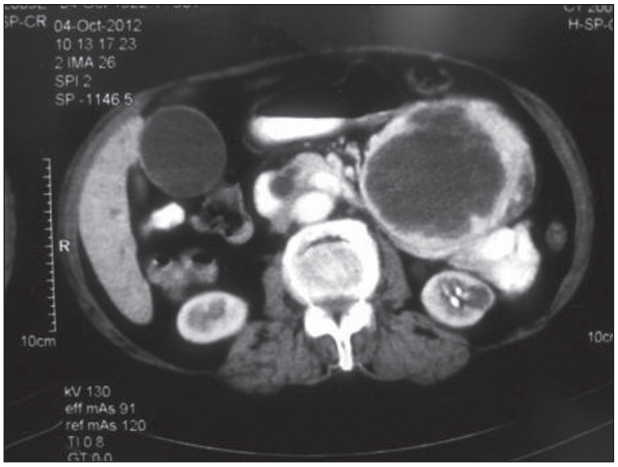

Figure 1: Abdominal CT revealed large retroperitoneal mass

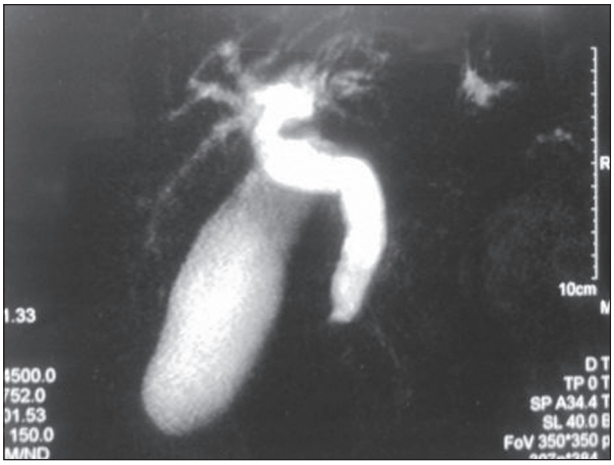

Figure 2: MRCP revealed biliary tract dilation

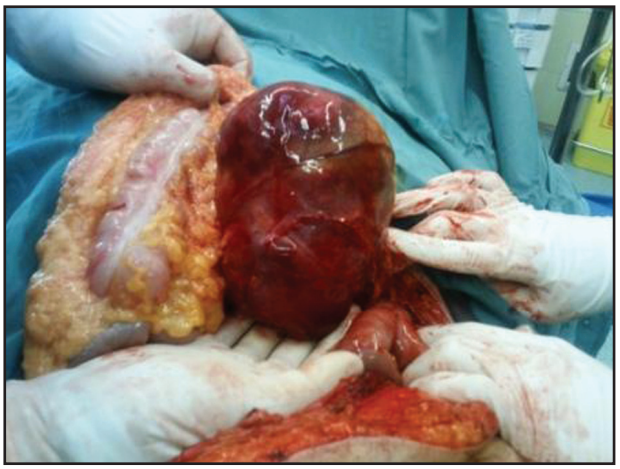

Figure 3: Large abdominal mass excised

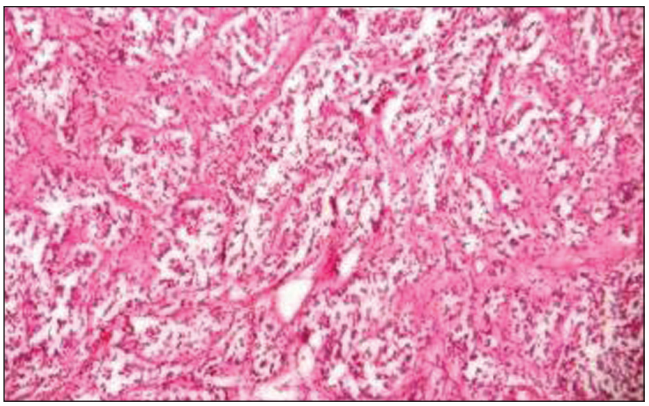

Figure 4: Paraganglioma histology (H&E, ×100)

tumors are incidentally discovered in normotensive patients during imaging evaluation for other reasons.[6]

They are commonly located in the para-aortic region and they may be confused with other retroperitoneal tumors, especially pancreatic tumors. Magnetic Resonance Imaging (MRI) is more sensitive than CT in detecting extra-adrenal tumors. Scintigraphy with 123-I labeled Meta-Iodo-Benzyl-GuanidineMIBG offers superior specificity than CT and MRI imaging.[4]

Besides the use of imaging studies for diagnosing paragangliomas, assessment of the plasma normetanephrine level or of the metanephrine and catecholamine levels via 24 h urine collection can help physicians make the actual diagnosis. In our patient's case, diagnosing the paraganglioma before surgery was not clear; so, we did not analyze the relevant serum marker. Complete surgical excision is the treatment of choice for extra-adrenal paragangliomas.[1]

A definitive diagnosis can be made only by histology and there are no histologic criteria to distinguish between benign and malignant tumors. Only the appearance of distant metastases, metastases at a site where paraganglionic tissue is not usually found or local recurrence can confirm that the tumor is cancerous. However, total excision is the basis of curative treatment because these tumors are potentially malignant.[7]

The precise origin of a periampullary adenocarcinoma is often difficult to determine even with standardized histopathologic evaluation, particularly if the tumor is large and involves more than one potential site of origin.[3,8] Tumor destruction of normal periampullary anatomy, and presence of epithelial dysplasia in more than a single periampullary compartment, occurs frequently.[9] However, in our case, according to early detection, the precise site of the neoplasm was clear.

Surgeons in treating patients with these tumors have favored an aggressive method of resection to benefit these patients with better prognosis. Although the perioperative outcomes for these tumors are similar, the long-term survival has traditionally varied.[8,10,11] It is unknown why outcome varies for adenocarcinomas arising from anatomic sites in such close proximity.[2]

Of the 242 patients with resected periampullary adenocarcinoma, 149 (62%) were pancreatic primaries, 46 (19%) arose in the ampulla, 30 (12%) were in distal bile duct, and 17 (7%) were duodenal cancers. The tumor-specific 5 year actual survival rates were 15% for pancreatic, 39% for ampullary, 27% for distal bile duct, and 59% for duodenal cancers. Among patients with periampullary adenocarcinoma treated by pancreaticoduodenectomy, those with duodenal adenocarcinoma are most likely to survive long-term.[8]

Developments in the broad field of molecular genetics may allow for earlier detection of periampullary neoplasms, perhaps using gene-based diagnostic testing of easily accessible clinical specimens such as blood, duodenal juice or stool. Furthermore, progress in adjuvant therapies, maybe incorporating new combinations of chemotherapy, radiation therapy, and immunotherapy, would result in improvements in disease-free and overall survival rates.[8]

In conclusion, Concurrent duodenal cancer and a retroperitoneal paragangliomas is an in experienced event in the clinic and pathologic fields and it makes pre-operative diagnosis of such masses difficult or even impossible, especially when the paraganglioma is non-functional.

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