World Journal of Medical and Surgical Case Reports

Case Report

Open Access

Polypoidal Ampullary Gangliocytic Paraganglioma: A Rare Case of Biliary Obstruction

Juinn Huar Kam, Alexander Yaw Fui Chung, Hock Foong Lui, Rafay Azhar

Hepatopancreatobiliary, Singapore General Hospital, Singhealth, Singapore.

Abstract Gangliocytic paraganglioma is a distinctively rare tumor that is mainly found in the second part of duodenum. It is a benign tumor which rarely recurs or metastasizes to regional lymph nodes. It can be sessile or pedunculated. It is usually in close proximity with the ampulla of vater, invasion of which may present with obstructive jaundice. Most of the gangliocytic paragangliomas are found incidentally during endoscopy or during investigation for gastrointestinal bleeding. Our patient is a 46 year old female and a hepatitis B carrier. Noted to have raised alkaline phosphatase on routine blood test. MRCP revealed dilated CBD with no ductal stone, pancreatic head mass or liver mass. ERCP showed a 4cm ampullary polyp with elongated mucosal stalk. Transduodenal ampullectomy with sphinteroplasty was performed. Histology and immunohistochemical staining confirmed tumor consistent with gangliocytic paraganglioma arises from the wall of ampulla, the tumor was excised completely with clear margin.

Introduction

Gangliocytic paraganglioma is a rare tumor that found mostly in the second part of duodenum. It is distinctively rare and only sporadic cases have been reported. Most were found incidentally or during investigation for upper gastrointestinal bleeding. We present a case of 46 year old lady with normal bilirubin and elevated alkaline phosphatase (ALP) due to a polypoidalampullary tumor.

Case Report

A 46 year old hepatitis B female carrier, noted to have raised alkaline phosphatase (ALP) but normal serum bilirubin in liver function test (LFT) during her routine follow up. She was otherwise asymptomatic with no abdominal pain or fever. She was not pale and declined history of melena. Clinical examination was unremarkable.

Ultrasound liver did not show any space occupying lesion but the common bile duct was prominent measuring 9mm. Magnetic resonance cholangio-pancreatography (MRCP) was performed which showed dilated CBD with normal pancreatic duct. No mass lesion at the head of pancreas or liver. A papillary grow was noted at the ampullary region of the duodenum. No lymph node metastasis was noted (Figure 1a, b).

Endoscopic evaluation revealed a 4 cm

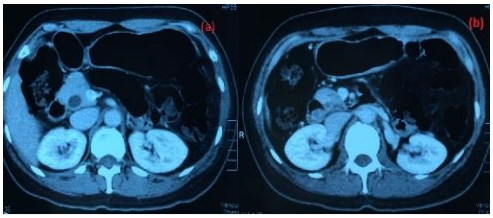

Figure 1: Computerized tomography scan a) and b) showing dilated CBD with ampullary tumor

Address for correspondence and reprint requests to:
Hepatopancreatobiliary, Singapore General Hospital, Singhealth, Singapore.
Email: kam.juinn.huar@sgh.com.sg

Submitted: Wednesday, January 7, 2015Accepted: Friday, February 13, 2015Published Friday, March 06, 2015

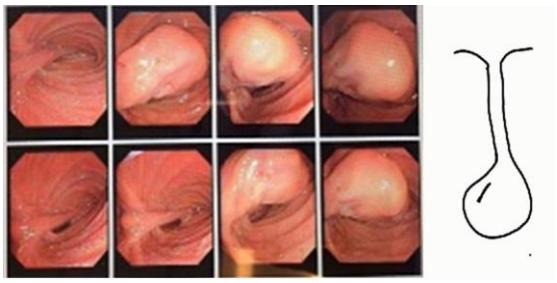

Figure 2: A) Endoscopic picture of polypoidal tumor with diagrammatic impression of mucosa stalk with slit representing exit for bilio-pancreatic duct B) line diagram

pedunculated ampullary tumor with thick mucosal stalk. There were surface ulcerations on the tumor with a slit like opening believed to be thebilio-pancreatico exit (Figure 2a, b). Endoscopic resection was considered but deemed unsuitable due to thick mucosal stalk.

The patient underwent transduodenal ampulectomy and sphinteroplasy. Dilated bile duct and normal caliber pancreatic duct were identified within the stalk after the transection of polypoidal tumor (Figure 3). Biliary and pancreatic cannulation revealed smooth bile and pancreatic juice flow (Figure4). Ducts to duodenal mucosa sphinteroplasty with septation were performed with PDS 5/0.

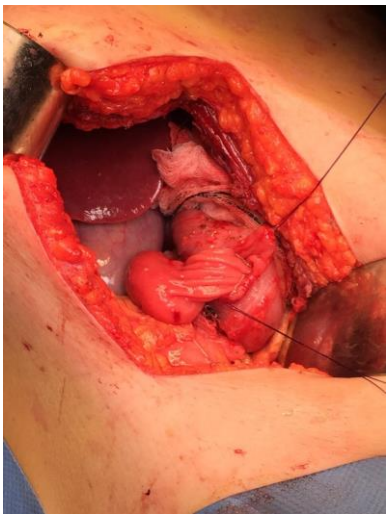

Figure 3: Broad mucosal stalk with palpable bilio-pancreatic duct

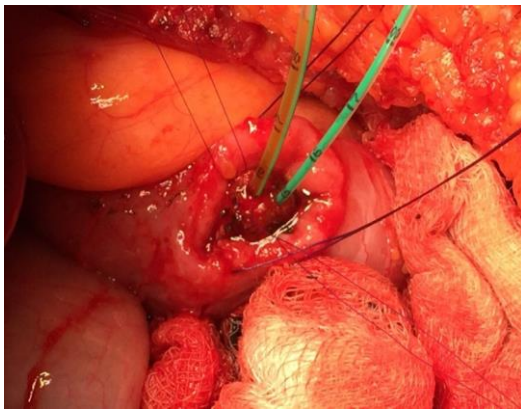

Figure 4: Post resection of tumor with septated bilio-pancreatic duct, the right superior showing cannulated bile duct with bilious fluid and the left inferior showing pancreatic duct with clear pancreatic juice.

Gross specimen revealed fleshy tumor at the ampulla (Figure 5). Histology showed an uncapsulated submucosal tumor located within the wall of the ampulla with tri-phasic morphology in varying proportions, comprising epithelioid cells arranged in nests, trabeculae and rosettes with abundant granular cytoplasm, surrounded by spindled cells with scattered ganglion cells showing abundant eosinophilic cytoplasm, large nuclei and prominent nucleoli. The epithelioid cells are positive for synaptophysin and neuron specific enolase and the spindled cells are S100 positive. Ki67 immunostain shows a low proliferation index (less than 1%) and c-kit (CD117) is negative. No definite amyloid deposition is noted. The lesion is 2cm from the resection margin and is completely excised. The morphological and immunohistochemical profile confirmed the diagnosis of gangliocytic paraganglioma.

Discussion

Gangliocytic paragangliomas (GP) are rare tumors that was first described by Dahl in 1957 as ganglioneuroma [1]. They present mostly in the periampullary region of the duodenum [2]. They are situated in the submucosa with mixture of epithelioid, ganglionic and spindle cells type in varying proportion [3]. They may be mistaken as GIST or schwannomas, which are

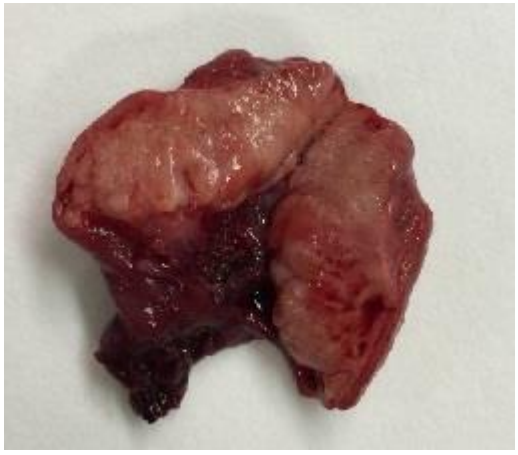

Figure 5: Specimen with mucosal stalk and bi-halved specimen

predominantly spindle-cell tumor but histologically and clinically distinct. Other differential diagnosis includes ganglioneuromas which are predominantly ganglion cells. Most patients are adults in their fifties with slight male to female preponderance (1.8:1). The origin and pathogenesis of GPs are not clear as they contain both endodermal (NET-like elements) and neural crest derivatives [4]. Some considered them neuroendocrine tumor as nest of epithelioid cells usually predominant and stained for synaptophysin and chromogranin. The ganglionic cells express synaptophysin and neurofilaments, and the spindle cells are positive for S100 protein [5]. It generally shows low proliferative index with Ki67 immunostaining and c-kit (CD117) is negative, which is the main differentiation from GIST, as with our case.

The association with Neurofibromatosis type 1 (NF1), an autosomal dominant disorder, has been reported in isolated cases, but vast majority of these cases are sporadic [6]. NF1 is known for its association with multiple benign and malignant tumors of nervous system and gastrointestinal tract. The most common gastrointestinal tumors occurring in patients with NF1 are GIST, neurofibromas and carcinoid tumors.

GPs are generally benign with rare occasion of recurrence or metastasis to regional lymph nodes. Regional lymph node metastasis was reported to occur in about 5-7% of the cases. Pleomorphism, increase proliferative index and regional lymph nodes involvement suggest potential malignancy [7].

Patients typically present with gastrointestinal bleeding with clinical manifestation of melena or anemia. Large tumor may present with upper abdominal pain and rarely gastric outlet obstruction. Obstructive jaundice is not common and is usually due to ampullary obstruction [8].

Endoscopic findings maybe of periampullary tumor, sessile or pedunculated, with or without ulceration. Due to its rarity, no consensus has been drawn regarding its treatment. A review of the presented cases in the literature has shown that local resection with clear margin, either endoscopic or open, may be sufficient for tumor without features suggestive of malignancy [9]. Endoscopic resection maybe feasible for tumor without deep invasion. Surgical resection is indicated for large sessile tumor or tumor with broad stalk as in our case. Transduodenal resection or ampulectomy with sphinteroplasty is usually sufficient if no sign of invasion or metastasis. Tumor with features suggestive of malignancy will require pancreaticoduodenectomy [10]. Patients should be followed up for recurrence.

Conclusions

Duodenal gangliocytic paraganglioma is a rare but important cause for biliary obstruction. Local wide excision with clear margin is adequate for tumor with no sign of invasion.

Conflict of Interest

We declare no conflict of interest

Authors' Contribution

JHK: Conceived the study, carried out the literature search, prepared the draft manuscript, design and edited the final manuscript

AYFC: Carried out the literature search and prepared the draft manuscript

HFL: Carried out the experiments and interpreted the results

RA: Carried out the experiments and interpreted the results

All authors read and approved the final manuscript.

Ethical Considerations

The written informed consent was obtained from the patient for publication of this case report.

References

1. Dahl EV, Waugh JM, Dahlin DC. Gastrointestinal ganglioneuromas; brief review with report of a duodenal ganglioneuroma. Am J Pathol 1957; 33: 953-65 [Pubmed].
2. Kwon J, Lee SU, Kang MJ, Jang JY, Kim SW. A case of gangliocytic paraganglioma in the ampulla of Vater. World J Surg Oncol 2010; 8:42 [Pubmed]
3. Perrone T, Sibley RK, Rosai J. Duodenal gangliocytic paraganglioma. An immunohistochemical and ultrastructural study and a hypothesis concerning its origin. Am J Surg Pathol 1985;9:31-41 [Pubmed].
4. Jarnagin WR. Blumgart's surgery of liver, biliary tract and pancreas Vol 1. 5th Edition, Saunders, an imprint of Elsevier Inc., Philadelphia 2012: 900.
5. Scheithauer BW, Nora FE, LeChago J, Wick MR, Crawford BG, Weiland LH, Carney JA. Duodenal gangliocytic paraganglioma. Clinicopathologic and immunocytochemical study of 11 cases. Am J Clin Pathol 1986; 86: 559-565[Pubmed].
6. Relles, D., Baek, J., Witkiewicz, A., and Yeo, C.J. Periampullary and duodenal neoplasms in neurofibromatosis type 1: two cases and an updated 20-year review of the literature yielding 76 cases. *J. Gastrointest. Surg.* 2010; 14: 1052–1061[Pubmed].
7. Okubo Y, Yokose T, Tuchiya M, Mituda A, Wakayama M, Hasegawa C, Sasai D, Nemoto T, Shibuya K. Duodenal gangliocytic paraganglioma showing lymph node metastasis: a rare case report. Diagn Pathol 2010; 5:27 [Pubmed].
8. Sathyamurthy A, Choudhary A, Ng D, Okponobi S, Diaz-Arias A, Grewal A, Hammoud GM. Obstructive jaundice due to rare periampullary tumor. World J Gastrointest Oncol. 2013; 5(10):195-7 [Pubmed].
9. Park SJ, Kim do H, Lim H, Lee JH, Choi KD, Song HJ, Lee GH, Jung HY, Kim JH, Park JY. Endoscopic resection as a possible radical treatment for duodenal gangliocytic paraganglioma: a report of four cases. Korean J Gastroenterol. 2014 Feb;63(2):114-9.[Pubmed]
10. Kemp CD, Russell RT, Sharp KW. Resection of benign duodenal neoplasms. Am Surg. 2007 Nov;73(11):1086-91.[pubmed]

World Journal of Medical and Surgical Case Reports

Published by Narain Publishers Pvt. Ltd. (NPPL)
The **Open Access** publishers of **peer reviewed** journals.
All articles are immediately published online on acceptance.
All articles published by NPPL are available **free** online