Case Report

iMedPub Journals
http://www.imedpub.com/

International Journal for Case Reports

2019
Vol.3 No.2:6

DOI: 10.36648/Case Report.3.2.6

Paraganglioma of the Gallbladder: An Extremely Rare Pathology

Humaira Haider Mahin*, Mina Sarofim and Mouhannad Jaber

Department of Surgery, Wollongong Hospital, University of New South Wales, Sydney, Australia

***Corresponding author:** Humaira Haider Mahin, MBBS, Department of Surgery, Wollongong Hospital, Tel: +61452464512; E-mail: mahinhaider@gmail.com

Citation: Mahin HH, Sarofim M, Jaber M (2019) Paraganglioma of the Gallbladder: An Extremely Rare Pathology. Int J Case Rep Vol: 3 No: 2:6

Received date: November 13, 2019; **Accepted date:** December 06, 2019**; Published date:** December 13, 2019

Abstract

We report an extremely rare case of gallbladder paraganglioma which was discovered incidentally, during a chest CT scan study of a 79-yr-old male for his underlying emphysema. He was asymptomatic with no abdominal pain, anorexia or jaundice. His abdominal CT scan showed a gallbladder mass which was intensely enhanced in the arterial phase. He underwent a laparoscopic cholecystectomy. His gallbladder histopathology concluded a completely resected non-chromaffin paraganglioma. The source of this gallbladder paraganglioma is likely from the paraganglionic cells along the vagi fibres which innervate the gallbladder. Chronic hypoxia is likely the risk factor for his paraganglioma. Paraganglioma should be considered in the differential diagnosis when investigating a gallbladder mass, particularly in patients with a history of chronic airway disease.

Keywords: Gallbladder; Paraganglioma; Neuroendocrine tumour; Extraadrenal; Chronic airway disease

Introduction

We present a 79 year old male who referred to the Upper GI unit of a tertiary hospital with a suspicious gallbladder wall mass, detected incidentally on a CT scan of his chest for his underlying emphysema.

He was asymptomatic with no upper abdominal pain, vomiting or jaundice. Physical examination was unremarkable as was full blood count and liver function tests.

Case Description

A dedicated abdominal ultrasound showed a 3.5 cm heterogeneous mass with increased vascularity in the fundus of gallbladder. Abdominal CT revealed a demonstrated the mass which intensely enhanced in the arterial phase **(Figure 1)**. There was no radiological lymphadenopathy or liver lesions.

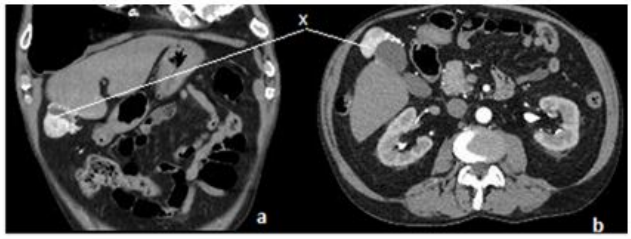

Figure 1: CT scan of the (a) Abdomen showing the coronal and (b) Axial view of the gallbladder tumor(X) which is intensely enhanced in the arterial phase.

This gentleman underwent an elective laparoscopic cholecystectomy. Intraoperatively, a 3 × 3 cm soft tissue mass was attached to the fundus of the gallbladder **(Figure 2)**, which was easily dissected away from the liver bed in the usual manner. Intra-operative cholangiogram was normal.

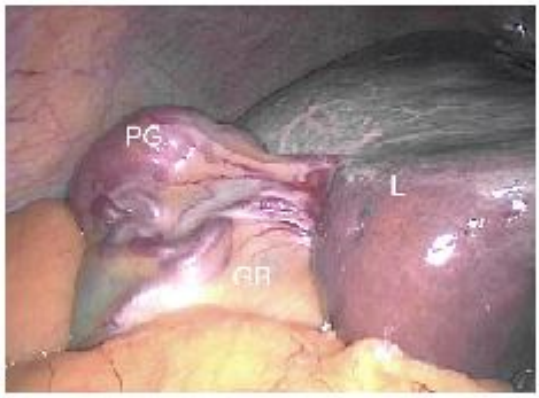

Figure 2: Intraoperative finding showing PG=Paraganglioma, GB=Gallbladder, L=liver.

Specimen histopathology concluded the lesion was a completely resected paraganglioma. The well circumscribed nodule arose from the interstitial tissues between the serosa and the wall of the gallbladder. Biliary mucosa showed no inflammation, dysplasia or metaplasia. The lesion was composed

of polygonal cells arranged in an irregular trabecular and nested pattern (known as Zellballen). The cells stained positively with synaptophysin and chromogranin. The sustentacular cells staining SOX-10 positive were arranged at the periphery of the poorly formed nests of chromatin and synaptophysin positive cell **(Figure 3)**. The lesion was negatively stained with CAM 5 suggesting a medullary chromaffin cell negative tumor [1].

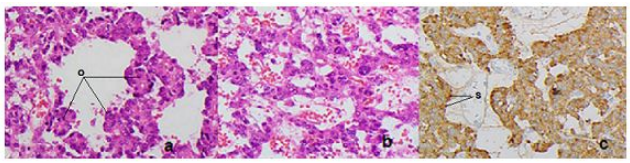

Figure 3: [a] Synaptophysin and chromogranin stained cells (o) with round to oval nuclei showing granular chromatin and eosinophilic cytoplasm [b] Cells are arranged in a trabecular and nested pattern (Zellballen) [c] SOX 10 positive sustentacular cells (s) are arranged at the periphery of the poorly formed nest of chromogranin and synaptophysin positive cells.

The patient made an uneventful recovery after being discharged on day one post-operative period. This case was discussed at a surgical oncology multi-disciplinary meeting and no further treatment was required.

Discussion

Paraganglia are clusters of neuroendocrine cells dispersed throughout the body, some connected with the sympathetic nervous system and others with the parasympathetic nervous system. The largest collection of these cells is found in the adrenal medulla where they give rise to pheochromocytomas. Paragangliomas develop in two general locations: [1]. Paravertebral paragangliomas which have sympathetic connections e.g. organs of Zuckerkandl and rarely bladder. These are chromaffin positive [2]. Paraganglioma related to the great vessels of the head and neck e.g. carotid bodies, ganglion nodosum of the vagus nerve and clusters located about the oral cavity, nose, nasopharynx, larynx and orbit. These are innervated by the parasympathetic nervous system and are referred to as non-chromaffin paragangliomas. These tumours infrequently release catecholamines, but because the neuroendocrine cells that make up these lesions sense oxygen and carbon dioxide tensions within adjacent vessels, the tumours are also sometimes referred to as chemodectomas [2]. The reported rate for paraganglioma is 2-8 cases per million per year [3], quite an infrequent pathology. Gallbladder paragangliomas are extremely rare, with less than 10 cases reported in the English literature.

In our case presented, the location and histopathology findings are in keeping with the latter group of paragangliomas. The intraabdominal distribution of paragangliomas typically involves the pancreas, duodenojejunal, periaortic, pericaval, bladder and sacral area [3]. The CT scan of abdomen and pelvis of our patient showed normal spleen, pancreas, suprarenal glands and kidneys. There was no retroperitoneal, mesenteric or pelvic lymphadenopathy demonstrated.

The origin of this gallbladder paraganglioma is likely from the paraganglionic cells along the vagi fibres which normally innervate the gallbladder [4].

Chronic hypoxia, for example secondary to chronic airway disease, is a recognized risk factor for paraganglioma development [5-7]. Apparently sporadic carotid body paragangliomas are more frequent in patients living at high altitudes and in the setting of chronic obstructive lung disease [5-7]. The prevalence of skull base and neck paraganglioma in high altitude areas is up to 1 in 10 in humans. For unclear reasons, most high-altitude paragangliomas (86% to 96 %) arise in females. The reason for the association between high altitude residence and paragangliomas of the carotid body is unclear.

This is the first reported paraganglioma in the gallbladder where the patient has a background of chronic hypoxia.

One case of gallbladder paraganglioma was associated with multiple endocrine neoplasia II syndromes (MEN II syndrome) [8]. Our patient did not have paroxysmal hypertension associated with episodic “classic triad” of tachycardia, sweating and headache. His blood levels of catecholamine were not raised. He did not have any positive family history of MEN II syndrome.

Considering a few factors like his presenting age, number of paraganglioma and his blood level of catecholamine, this was more likely a sporadic paraganglioma rather than hereditary paraganglioma.

There are few studies suggest MRI or Metaiodobenzylguanidine (MIBG) scintigraphy scans are ideal for suspected paragangliomas as they can identify multiple primary tumours and/or metastases. MIBG scintigraphy is a functional imaging technique which uses a norepinephrine analogue labelled with 123I or 132I. It specificity is 95%-100% and sensitivity is 85% [9]. Paragangliomas have low or intermediate signal intensity or T1 weighted images and high signal intensity on T2-weighted images. Classically, there are high and low-signal regions of the tumour described as “salt and pepper” appearance on T2-weighted images because of slow flow or haemorrhage and signal voids of tumour vessel [10].

Although these lesions may be discovered incidentally it is crucial to diagnose and treat biliary system paragangliomas due to their potential complications: obstructive jaundice, right upper quadrant abdominal pain or gastrointestinal bleeding [8].

Complete resection with laparoscopic cholecystectomy is the ideal management for gallbladder paraganglioma followed by discussion at multidisciplinary surgical oncology meetings as there are no defined management guidelines due to their rarity.

Conclusion

Gallbladder paraganglioma is an extremely rare pathology which was detected incidentally in this asymptomatic man with a history of chronic air way disease. Although infrequent, it should be considered in the differential diagnosis of any

hypervascular gallbladder mass particularly in patients with a history of chronic airway disease. From previously reported cases of paraganglioma of gallbladder and the outcome of multidisciplinary surgical oncology meeting, we believe cholecystectomy was the adequate treatment for this condition.

Disclosure statement

The authors declare no financial or non-financial conflicts of interest concerning the research, authorship and publication of this article.

References

1. Miller AD, Masek-Hammerman K, Dalecki K, Mansfield KG, Westmoreland SV(2009) Histologic and immunohistochemical characterization of Pheochromocytoma in six cotton-top tamarins (Saguinus oedipus). Vet Pathol 46: 1221-1229.
2. Cotran RS, Kumar V, Collins T (1999) Head and Neck (6th Edn) W.B. Saunders Company.
3. Erickson D, Kudva YC, Ebersold MJ, Thompson GB, Grant CS, et al. (2001) Benign paragangliomas: Clinical presentation and treatment outcomes in 236 Patients. J Clin Endocrinol Metab 86: 5210-5216.
4. Sclafani LM, Woodruff JM, Brennan MF (1990) Extraadrenal retroperitoneal paragangliomas: Natural history and response to treatment. Surgery 108: 1124-1129; Discussion 1129-1130.
5. Saldana MJ, Salem LE, Travezan R (1973) High altitude hypoxia and chemodectomas. Hum Pathol 4: 251-263.
6. Khan Q, Heath D, Smith P, Norboo T (1988) The histology of the carotid bodies in highlanders from Ladakh. Int J Biometeorol 32: 254-259.
7. Rodríguez-Cuevas S, López-Garza J, Labastida-Almendaro S (1998) Carotid body tumors in inhabitants of altitudes higher than 2000 meters above sea level. Head Neck 20: 374-378.
8. Mehra S, Chung-Park M (2005) Gallbladder paraganglioma: A case report with review of the literature. Arch Pathol Lab Med 129: 523-526.
9. Shapiro B, et al. (1995) The current status of Meta-iodobenzylguanidine and related agents for the diagnosis of neuro-endocrine tumors. Q J Nucl Med 39: 3-8. PubMed PMID: 9002740.
10. Lee KY (2006) Extraadrenal paragangliomas of the body: Imaging features. AJR Am J Roentgenol. 187: 492-504. PubMed PMID: 16861555.