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GALL BLADDER PARAGANGLIOMA - A RARE AND PRECARIOUS DIAGNOSIS - A CASE REPORT AND REVIEW OF LITERATURE

Dr. Rohit Kapoor	M.S. General Surgery, Senior Clinical Fellow, Department Of General Surgery, , Barking Havering and Redbridge university hospitals, Romford London, United Kingdom
Dr. Safarudeen Sapharullah*	Post- graduate Resident, Department Of General Surgery, Maharishi Markandeshwar Institute of Medical Sciences & Research, Mullana, India *Corresponding Author
Dr. Vijay Shrawan Nijhawan	Prof. & HOD, Department of Pathology, Maharishi Markandeshwar Institute of Medical Sciences, Mullana-Ambala, Haryana
Dr. Vishesh Dhawan	Senior Resident, Department of Pathology, Maharishi Markandeshwar Institute of Medical Sciences, Mullana-Ambala, Haryana
Dr. Shubh Kirti	M.D.S. Periodontology, Associate Professor, Department of Periodontology, Maharishi Markandeshwar College of Dental Sciences & Research, Mullana, India

ABSTRACT

Introduction: Neuroendocrine neoplasm (NET) of the gallbladder is an extremely uncommon diagnosis. We present a case of a benign gallbladder paraganglioma which should be differentiated from malignant NET. **Case Presentation:** A 63-year-old female came to surgical clinic with complaints of recurrent biliary colic which aggravated on taking fatty food and relieved on medication. She was admitted in surgical ward and routine blood investigations and imaging work up was done. USG whole abdomen was consistent with cholelithiasis. She underwent elective laparoscopic cholecystectomy under General anaesthesia. Intra operative and post operative period were uneventful. Histopathological examination report revealed chronic cholecystitis with paraganglioma in the gall bladder **Discussion:** Neuroendocrine neoplasms of the gallbladder is a rare finding. Their management includes simple cholecystectomy and radical resections in advanced cases and chemotherapy. On the contrary gall bladder paraganglia is a benign entity which is managed with cholecystectomy. **Conclusion:** A neuroendocrine neoplasm of the gallbladder may closely resemble a benign paraganglion. If a NET is suspected, one should be aware of benign paraganglioma entity.

KEYWORDS : Paraganglioma, Neuroendocrine tumour, Benign

INTRODUCTION

A neuroendocrine neoplasm of the gallbladder is a rare diagnosis. Often there can be confusion between neoplastic tumour and benign paraganglia. After an elective cholecystectomy roughly 2.8 % of histopathology specimens turn out to be neoplastic and out of this only 2% are neuroendocrine neoplasm[1]. We present a patient who was diagnosed with gall bladder paraganglioma on histopathological analysis which is even more uncommon.. The aim of this case report is to make clinicians aware of this rare entity and differentiate between benign and malignant entity so that treatment can be offered accordingly.

Case Presentation:

A 63-year-old female came to surgical clinic with complaints of recurrent biliary colic which aggravated on taking fatty food and relieved on medication. She was admitted in surgical ward and routine blood investigations and imaging work up has been done. Ultrasound whole abdomen was consistent with cholelithiasis. She underwent elective laparoscopic cholecystectomy under GA. Intra operative and post operative period were uneventful. Histopathological examination report revealed chronic cholecystitis with paraganglioma in the gall bladder

DISCUSSION

Gall bladder cancer incidence is on the decline but it is still found [2]. The most common histopathological type of gall bladder cancer is adenocarcinoma followed by adeno-squamous carcinoma, squamous cell carcinoma, and neuroendocrine neoplasms [3]. A neuroendocrine neoplasm is an epithelial arising malignant tumour with cells differentiating into neuroendocrine lineage which is evident on morphology and immunohistochemistry. Often these tumours produce symptoms due to secretion of neuroendocrine hormones. A gall bladder NET is uncommon and found in 0.2 – 0.5 % of all neuroendocrine neoplasms, and just 2 % of all malignant gallbladder tumours [4-7].

The gall bladder NET are uncommon and difficult to diagnose. The diagnosis of a gallbladder NET is difficult due to its rarity and often has non-specific symptoms of right upper quadrant abdominal pain, jaundice and weight loss. This is most often a histopathological diagnosis after cholecystectomy. Of all NET only 1% present with a carcinoid syndrome or other paraneoplastic syndromes[8].

There is little known about primary gallbladder paragangliomas. They arise from the migration of the paraganglia of the hepatic plexus thus innervating the gallbladder through cystic plexus[9] which contains both parasympathetic and sympathetic fibres. These primary tumours are found to be non-functioning parasympathetic paragangliomas, hence are benign tumours. In these beningn incidental tumours simple cholecystectomy would suffice. In contrast, sympathetic paragangliomas are more prone for malignant transformation, therefore are resected after staging. The gall bladder also has sympathetic innervation so there are chances of malignant neoplasm although no case of malignant paraganglia has been reported. If the NET is suspected the hereditary syndromes should be investigated if multiple paragangliomas are present[10].

These paraganglioma of the gallbladder arises from chief and sustentacular cells which originate from the neural crest

cells[11]. The characteristic histopathologic findings include Zellballen nests and positive chromogranin and synaptophysin staining of tumour cells (Fig 1,2,3).

NETs of the gallbladder are rare form of gall bladder cancer and can be aggressive and of poor prognosis depending on subtype. Patients usually present with symptoms of benign gallstone disease. NETs are clinically indistinguishable from other types of gallbladder cancer. The radical resection with lymphadenectomy is recommended for the malignant tumour but a benign paraganglioma can be managed with simple cholecystectomy

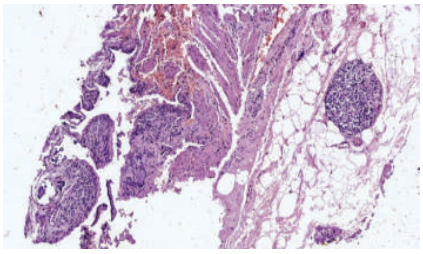

Figure 1. Low power photomicrograph showing patchy mucosal ulcerations with one well-demarcated, lobular structure, measuring 2 mm in diameter, in close proximity to small vessels within the subserosal connective tissue of the gallbladder wall (H and E, x100).

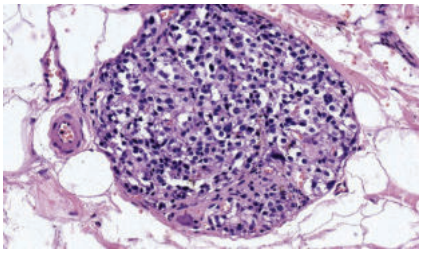

Figure 2. High power photomicrograph shows a well circumscribed lobular structure composed of nests of chief cells separated by thin bands of fibrovascular tissue ("zellballen" pattern). The cells have round dark nuclei, coarse chromatin ("salt and pepper") pattern and clear cytoplasm (H and E, x400).

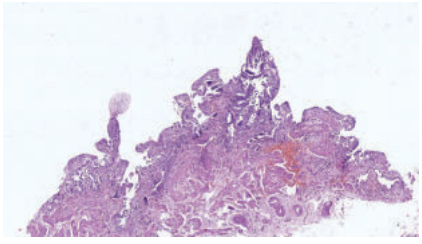

Figure 3. Section shows features of chronic cholecystitis in the form of patchy mucosal ulcerations, increase in lymphomononuclear infiltrate within the lamina propria, muscle hypertrophy and subserosal fibrosis (H and E, x40).

CONCLUSION

This was an incidental case of paraganglioma of gall bladder found after laparoscopic cholecystectomy for recurrent biliary colic. There are limited knowledge and case reports about the beningn paraganglioma which arise from parasympathetic fibres and can be managed with cholecystectomy. Although gall bladder has both sympathetic and parasympathetic fibres which can lead to malignant neoplasm but very less is known about this. Due to limited number of cases reported optimal follow up of these cases is unknown.

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