Gallbladder neurofibroma presenting as chronic epigastric pain - Case report and review of the literature

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Abstract
Context: Benign nonepithelial neoplasms of the gallbladder are unusual. The majority of gallbladder neurofibromas are found incidentally in the gallbladder specimens following cholecystectomy. There have been only few reports in the literature describing this rare entity. In this study we report a case of gallbladder neurofibroma presenting as chronic epigastric pain in a young patient. Case Report: A thirty two year old otherwise healthy man presented to our clinic with chronic epigastric pain symptom after eating. Physical examination, laboratory and radiologic workups were unremarkable for signs of biliary tract diseases. Past medical and surgical histories were significant only for neurofibromatosis type I. Due to persistent symptomatology, the patient was taken to the operating room for a diagnostic laparoscopy followed by laparoscopic cholecystectomy. Open conversion was necessitated because of the presence of a gallbladder mass preventing safe anatomic dissection. Surgical pathology revealed plexiform neurofibroma with noninflamed gallbladder. The postoperative course was unremarkable and the patient was pain free at 3 weeks postoperatively. Conclusions: Benign neoplasms such as gallbladder neurofibroma should be included in the differential diagnosis for chronic epigastric pain symptom in a young otherwise healthy patient with neurofibromatosis. Diagnostic laparoscopy should be considered in an individual presenting with this condition.

Keywords: Neurofibroma, gallbladder, benign tumors.

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Introduction
Neurofibroma is a benign and slowly growing tumor arising in a nerve structure and composed primarily of Schwann cells. This benign tumor can occur as a solitary lesion or more commonly is associated with neurofibromatosis type I (NF-1). While most neurofibromas commonly occur as superficial skin or subcutaneous lesions, neurofibromas of abdominal viscera are seen infrequently and when they occur in this location, they are usually associated with NF-1 [1].

Despite cholecystectomy having been the most commonly performed operation by general surgeons for many decades, there are only a few reports describing gallbladder neurofibroma in the clinical literature.

Case Report
A 32-year-old man presented to our clinic with a one-year history of epigastric pain and dyspepsia related to meals. Past medical/surgical/social histories were significant only for NF-1. Physical examination, liver function tests, abdominal ultrasonography and hepatobiliary iminodiacetic acid (HIDA) scan were otherwise unremarkable for signs of biliary tract pathology. Due to the unrelenting symptoms, the decision was made to proceed with diagnostic laparoscopy and possible
cholecystectomy. Upon exploration of the right upper quadrant, the gallbladder was found to have markedly thickened wall with a dense soft tissue bundle along its medial aspect extending down to the hepatocystic triangle. Due to the difficulty in achieving a critical view of the cystic duct and artery, the operation was converted to an open cholecystectomy. The resected specimen measured 8 x 5 cm with a soft tissue mass protruding from the gallbladder wall (Figs. 1 and 2).

![Fig. 1](image1.jpg)
**Fig. 1** Intact gallbladder specimen following the operation. Neurofibroma extended along the inferior wall of the gallbladder from the cystic duct to the fundus, protruding extraluminally.

![Fig. 2](image2.jpg)
**Fig. 2** Opened gallbladder specimen showing mural nodules and normal mucosa. Gallbladder appeared to be normal without signs of inflammation. Silk sutures were used to ligate the cystic duct prior to division.

![Fig. 3](image3.jpg)
**Fig. 3** Histologic appearance of gallbladder neurofibroma. Photomicrograph shows diffuse neurofibroma cells with short fusiform and round shapes within fine fibrillary collagen matrix.

Surgical pathology revealed a plexiform neurofibroma with noninflamed gallbladder tissues. Photomicrograph showed diffuse neurofibroma cells with short fusiform and round shapes within fine fibrillary collagen matrix (Fig. 3). The postoperative course was unremarkable and at three-week followup, the patient had complete resolution of his epigastric symptoms.

**Discussion**
Benign tumors of the gallbladder are rare and lesions of neural origin are even more unusual. Such benign growths include paragangliomas that presumably arise from paraganglia of the gallbladder, neuromas of the cystic duct remnant, or granular cell myoblastomas. Neurofibromas are formed by a combined proliferation of all components of peripheral nerve with Schwann’s cell usually being the predominant element. On histopathology, they appear as spindle-shaped cells positively stained with immunoperoxidase technique.

The incidence of hepatobiliary neurofibromas is lower than that of other digestive organs. In the gastrointestinal tract, neurofibromas are most commonly located in the ileum, followed by the jejunum, duodenum and stomach [2]. In a case described by Hochberg et al., the most common sites affected were the jejunum and stomach [3]. Gastrointestinal involvement of von Recklinghausen’s disease essentially occurs in three forms: (a) hyperplasia of the submucosa and myenteric plexus with mucosal ganglioneuromatosis, (b) gastrointestinal stromal tumor showing varying degrees of neural and smooth muscle differentiation, (c) somatostatin-rich carcinoid of the duodenum with psammoma bodies and which may be associated with pheochromocytoma [3-5].

Ultrasoundographic imaging often demonstrates either localized thickening of the gallbladder wall or broad based elevation of the mucosa. Riopelle has described superficial or mucosal, and deep or adventitial neurofibromatosis [6]. Grossly, the appearance of a gallbladder neurofibroma can be very similar to that of a coexisting cholesterol polyp.

Based on the literature review presented in Table 1, most neurofibromas are found incidentally following cholecystectomy for symptomatic cholelithiasis or chronic cholecystitis. Gender distribution is equal with mean age of 57.9 (range=32-77). Approximately 50% of patients were found to have associated gallstones. Neurofibromatosis mainly occurred in the body of gallbladder and presented as a mural nodule that can protrude either intra or extraluminally. With further technological advances in diagnostic imaging, these lesions will likely be identified in increasing frequency. Cholecystectomy is the current standard of treatment.

**Conclusion**
Benign neoplasms such as gallbladder neurofibromas
should be included in the differential diagnosis for chronic epigastric pain symptom in patients with neurofibromatosis. Diagnostic laparoscopy followed by cholecystectomy is recommended for this condition.

Table 1 Previously published reports on gallbladder neurofibroma

| Authors            | Age | Sex  | Location | Size (cm) | Gross Pathologic Appearance         | Cholelithiasis | NF-1 |
|--------------------|-----|------|----------|-----------|-------------------------------------|----------------|------|
| Aisner SC [7]      | 44  | Male | Neck     | 0.3       | Mural Nodule                        | Yes            | No   |
| Albores-Saavedra J [8] | 44  | Female | Body   | 0.7 x 0.5 | Papillary polypl                    | No             | No   |
| Arbab AA [9]       | 77  | Female | Neck     | 0.4       | Mural Nodule                        | Yes            | No   |
| Christensen AH [10] | 70  | Male | Body     | 1         | Mural Nodule                        | Yes            | No   |
| Eggleston JF [11]  | 65  | Female | Body     | 1         | Mural Nodule, protruding intraluminally | Yes            | No   |
| Elhag AM [12]      | 70  | Female | Body     | 0.5       | Mural Nodule                        | Yes            | No   |
| Fuller CE [4]      | 61  | Male | Fundus   | 5.2 x 2.8 | Luminal Mass                        | No             | No   |
| Current Case       | 32  | Male | Body     | 5 x 3     | Mural Nodule, protruding extraluminally | No             | Yes  |

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