

# Clinical review of the carcinoid tumor of the cystic duct: A rare tumor at a rare site

Rasees Fawwaz Alotaibi, Yara Ali Alnashwan,  
Maryam Mohammed Alomar, Shaukat Ahmad Bojal,  
Mohammed Saad Alqahtani, Iftikhar Khan

## ABSTRACT

**Introduction:** Carcinoid tumor arising in the cystic duct is extremely rare. This is the 23rd case reported in English literature. **Case Report:** A case of carcinoid tumor in the cystic duct of a 68-year old lady is described that was found incidentally after laparoscopic cholecystectomy. **Conclusion:** Presentation, diagnosis, treatment and prognosis are discussed based on the review of the reported cases. It usually presents with biliary symptoms and the diagnosis is incidental. Radical resection and follow up are recommended because of its malignant potential.

**Keywords:** Carcinoid tumor, Cystic duct, Neuroendocrine tumor

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Rasees Fawwaz Alotaibi<sup>1</sup>, Yara Ali Alnashwan<sup>2</sup>,  
Maryam Mohammed Alomar<sup>3</sup>, Shaukat Ahmad Bojal<sup>4</sup>,  
Mohammed Saad Alqahtani<sup>5</sup>, Iftikhar Khan<sup>6</sup>

**Affiliations:** <sup>1</sup>MBBS, Radiology Resident, Medical Imaging Department, King Fahad Specialist Hospital, Dammam, Saudi Arabia; <sup>2</sup>MBBS, Medical Intern, University of Dammam, Dammam, Saudi Arabia; <sup>3</sup>MBBS, Medical Intern, University of Dammam, Dammam, Saudi Arabia; <sup>4</sup>MD, Hepatobiliary Surgeon, Section of Hepatobiliary Surgery, King Fahad Specialist Hospital, Dammam, Saudi Arabia; <sup>5</sup>MD, Transplant and Hepatobiliary Surgeon, Department of Transplant Surgery and Section of Hepatobiliary Surgery, King Fahad Specialist Hospital, Dammam, Saudi Arabia; <sup>6</sup>MD, Transplant Surgeon, Department of Transplant Surgery, King Fahad Specialist Hospital, Dammam, Saudi Arabia.

**Corresponding Author:** Dr. Iftikhar Khan, MOTC, KFSH, PO Box: 15215, MBC: 067, Dammam 31444, Saudi Arabia; Email: iftikhar.khan@kfs.med.sa

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## INTRODUCTION

Carcinoid tumor arising in the cystic duct is exceedingly rare. Over a period of 32 years, since 1984, when Goodman et al. [1] described the first case, only 22 cases of the cystic ductal carcinoid tumors have been reported. We present another similar case followed by review of literature. This is the first case reported from the Middle East.

## CASE REPORT

A 68-year-old female had undergone laparoscopic cholecystectomy for symptomatic gallstones in another hospital, was referred for incidentally discovered carcinoid tumor in the cystic duct. She was a known case of diabetes mellitus, hypertension and osteoporosis. Her symptom was post-prandial right upper quadrant pain for one year. At the time of her presentation to our hospital, she was asymptomatic with unremarkable examination. Her complete blood count, liver function tests and serum chromogranin A, serotonin and octreotide levels were

within normal range. Imaging of her abdomen and chest by computed tomography (CT), magnetic resonance (MRI) and positron emission tomography (PET) did not reveal any metastatic deposit.

Macroscopic review of the gallbladder revealed a nodular tumor in the cystic duct which was 0.7 cm in diameter. Microscopic examination showed nests, cords and trabeculae of small cells with granular chromatin which were positive for synaptophysin, chromogranin, and CD56 on immunohistochemistry (Figure 1). The tumor invaded the perineural and muscle tissue. This was consistent with well-differentiated carcinoid tumor.

The patient underwent resection of the cystic duct stump, liver segments 4B and 5 and clearance of hepatoduodenal lymph nodes. Appendectomy was also done because of a nodule in the mesoappendix which turned out to be benign fibrosis. No residual tumor was found on frozen and permanent sections. Her postoperative recovery was unremarkable. At 16th month follow-up, she is doing well with no evidence of recurrent disease on MRI and PET scans.

## DISCUSSION

Carcinoid tumors, arising from the enterochromaffin cells, are extremely rare and account for 0.49% of all malignancies [2]. Only 0.2–2.0% of carcinoid tumors are found in the extra-hepatic bile ducts [3]. Cystic duct carcinoid tumors represent 11–20% of all extra-hepatic bile duct carcinoid tumors [3], highlighting the rarity of this particular tumor at this particular site. Ours is the 23rd case reported in English literature (Table 1) [1, 2, 4–21].

Review of the reported cases demarcates the clinical presentation into two groups. Isolated cystic duct involvement usually presents with clinical picture similar to symptomatic gallstones. Gallstones were present in two-thirds of the reported cases. When main hepatic/bile duct gets involved then obstructive jaundice is the usual presentation. Endocrine symptoms are rare and only one patient, who had multiple carcinoid tumors at various locations as part of the multiple endocrine neoplasia type-I (MEN-I) syndrome, presented with Zollinger-

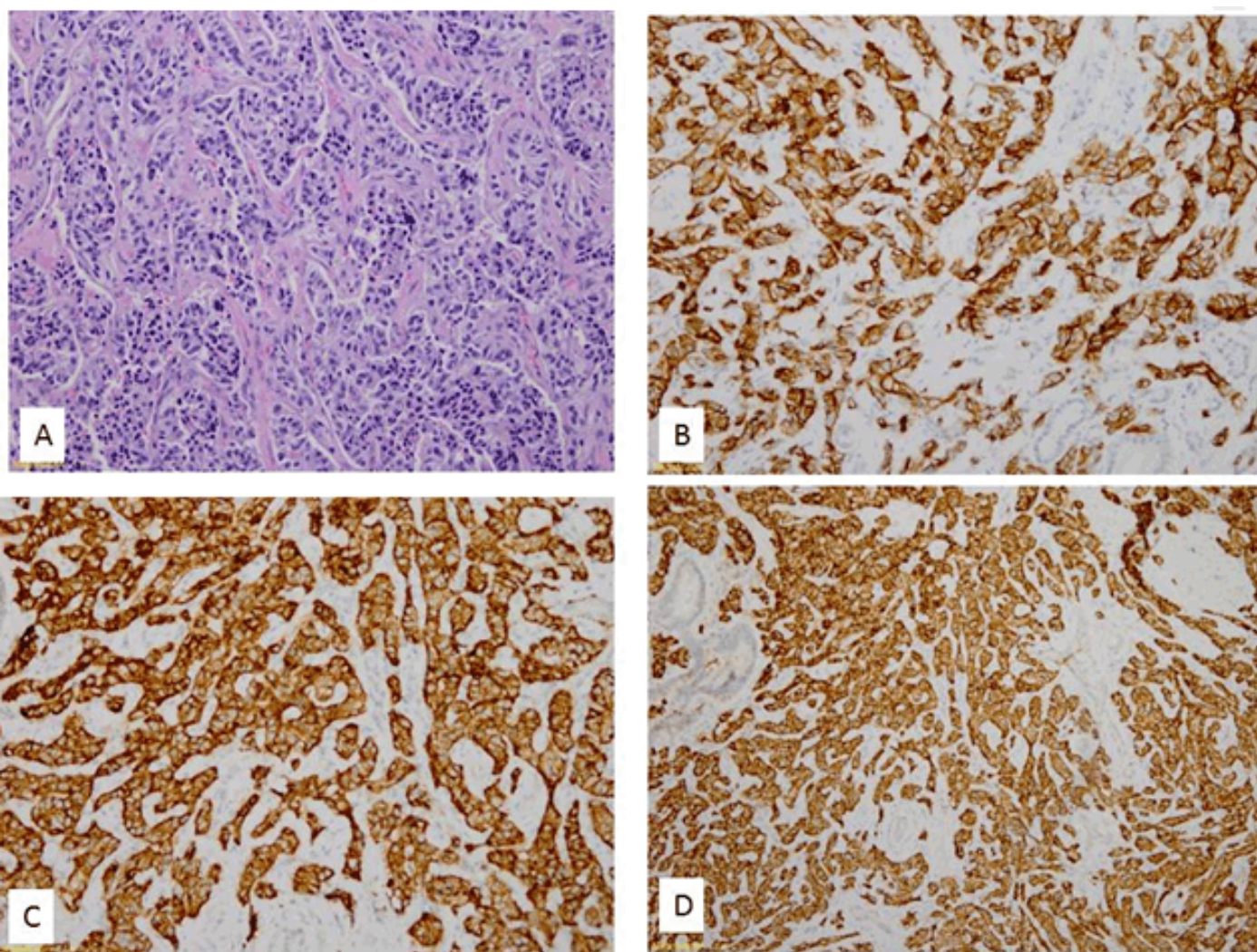


Figure 1: (A) Photomicrograph of the carcinoid tumor in the cystic duct, showing nests, cords and trabeculae of small cells (H&E stain, x200), (B) Positivity for synaptophysin, (C) Chromogranin, and (D) CD56.



Table 1: Cystic duct carcinoid cases

First Author/Year [Reference]	Sex	Age	Symptoms	Location	Gall-stones	Outcome
Goodman/1984 [1]	F	28	Epigastric, RUQ and mid-back pain	CD	No	Well 9 months
Nicolescu/1986 [4]	F	50	Biliary colic	CD	Yes	Not available
Chittal/1989 [5]	F	46	Biliary colic	CD	No	Well 3 years
Rugge/1992 [6]	F	64	Biliary colic Jaundice	CD-CHD junction	No	Well 12 months
Ishibashi/1995 [7]	F	77	RUQ pain, epigastric mass	CD(multiple) Gallbladder	No	Not available
Meyer/1997 [8]	F	56	Mild dyspepsia	CD	Yes	Well 4 years
Shah/1998 [9]	F	52	RUQ pain	CD	Yes	Not available
Oikawa/1998 [10]	M	70	Jaundice	CD-CHD junction	No	Died 6 months
Hermina/1999 [11]	M	69	RUQ pain	CD	Yes	Well 14 months
Aronsky/1999 [12]	F	64	Asymptomatic growing gall bladder polyp	CD	No	Well 47 months
	F	51	Acute cholecystitis	CD	Yes	Well 49 months
Hubert/2005 [13]	M	50	Epigastric pain	CD	No	Well 17 months
Stavridi/2007 [14]	F	49	RUQ pain, nausea, fever	CD	Yes	Well 12 months
Sethi/2007 [15]	M	51	Pedal edema, weight loss, abnormal LFT	CD-CHD junction	No	Well 22 months
Felekouras/2009 [16]	F	60	Jaundice	CD invading CHD	No	Liver metastasis at 12 months
Price/2009 [17]	F	55	ZES	CD-CHD junction	No	Well 2 years
Liew/2011 [2]	M	40	Biliary colic	CD	Yes	Well 12 months
Ioannidis/2012 [18]	F	41	Symptomatic gallstones	CD	Yes	Not available
Lim/2013 [19]	F	58	RUQ pain, nausea, vomiting, diarrhea	CD	Yes (sludge)	Well 10 months
Garland/2014 [20]	F	48	Epigastric/RUQ pain, nausea, vomiting, diarrhea	CD	No	Well 4 years
Hong/2015 [21]	F	58	Jaundice	CD	N/A	Not available
	M	51	Abdominal pain	CD	N/A	Not available
Present case	F	68	Symptomatic gallstones	CD	Yes	16 months

F female, M male, RUQ right upper quadrant, ZES Zollinger-Ellison syndrome, CD cystic duct, CHD common hepatic duct, N/A: not available.

Ellison syndrome [17]. Majority of the patients were in their fifth and sixth decades and were females. All cases with isolated cystic duct carcinoid tumor were diagnosed incidentally after cholecystectomy on histological examination. The yield of endoscopic retrograde cholangiopancreatography (ERCP) and biopsy in cases that presented with obstructive jaundice was low and only one patient was diagnosed correctly preoperatively [13].

One-third of the patients had evidence of metastasis. Synchronous spread to the regional lymph nodes was seen in four patients [1, 2, 11, 21]. Synchronous liver deposits were present in two patients [13, 17]. Metachronous liver metastasis occurred in two patients [10, 16]. It is important to note that the locations of liver metastasis were away from the gallbladder bed which requires re-assessment of the value of adding the resection of liver segments 4 and 5 in the primary clearance.

Complete surgical resection is the best treatment for the fit patient. Isolated cystic duct carcinoid tumor requires clearance of the cystic duct stump and the hepatoduodenal lymph nodes. This can be achieved with open or laparoscopic approaches [20]. Involvement of the main hepatic/bile ducts requires excision of the extra-hepatic bile ducts, clearance of the lymph nodes and reconstruction with hepaticojejunostomy. Liver metastasis can be treated by locoregional modalities such as radio frequency ablation, ethanol injection and chemoembolization. Hormone therapy with somatostatin analogues may be considered in frail patients and those with endocrine symptoms. Radiotherapy and chemotherapy should be reserved for surgically unfit, recurrent or metastatic disease.

During the reported follow-up that ranged between 6 and 49 months, one patient died at sixth month due to liver metastasis [10]. Due to their malignant potential, the patients ought to be followed-up by imaging.

## CONCLUSION

In conclusion, cystic duct carcinoid tumor is a rare tumor which presents either with biliary symptoms or obstructive jaundice. The preoperative diagnosis is difficult. Radical surgical resection is the best treatment. Follow-up is recommended because of relatively low but definite malignant potential.

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## Author Contributions

Rasees Fawwaz Alotaibi – Conception and design, Acquisition of data, Analysis and interpretation of data, Critical revision of the article, Final approval of the version to be published

Yara Ali Alnashwan – Conception and design, Acquisition of data, Analysis and interpretation of data, Critical revision of the article, Final approval of the version to be published

Maryam Mohammed Alomar – Conception and design, Acquisition of data, Analysis and interpretation of data, Critical revision of the article, Final approval of the version to be published

Shaukat Ahmad Bojal – Conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Critical revision of the article, Final approval of the version to be published.

Mohammed Saad Alqahtani – Conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Critical revision of the article, Final approval of the version to be published

Iftikhar Khan – Conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Critical revision of the article, Final approval of the version to be published

## Guarantor

The corresponding author is the guarantor of submission.

## Conflict of Interest

Authors declare no conflict of interest.

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