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A Descriptive Study On Cholangiocarcinoma

Gastroenterology

KEYWORDS: cholangiocarcinoma, bile duct cancer, jaundice

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ABSTRACT

Cholangiocarcinoma is a grave malignancy with 85-90% patients presenting to healthcare at advanced stage of disease. Best treatment for this condition is surgery and without surgery median survival is only 5-8 months. The most important prognostic factor is complete resection and absence of lymph node metastasis - both of which require early detection. In our study there were total 39(66%) hilar, 13(22%) distal and 7(12%) intrahepatic cholangiocarcinomas. 20 (34%) patients had distant metastasis at presentation itself. Only 3(5%) patients underwent curative resection and rest of the patients mainly underwent only palliative interventions. This is an important concern as the only chance for cure in cholangiocarcinoma is surgery and hence more studies are urgently required to focus on early detection and better treatment of this condition.

Introduction

Cholangiocarcinoma is a grave malignancy with 85-90% patients presenting to healthcare at advanced stage of disease. Best treatment for this condition is surgery and without surgery median survival is only 5-8 months. The most important prognostic factor is complete resection and absence of lymph node metastasis - both of which require early detection. The three types are hilar, intrahepatic and distal.

Aims of the study

1. To study the pattern of cholangiocarcinoma cases presenting to a teritary care centre in South India.
2. Comparing the features of hilar, intrahepatic and distal cholangiocarcinoma.
3. To optimize the preoperative diagnosis and management of cholangiocarcinoma.

Methodology

The medical records of 59 patients with a diagnosis of cholangiocarcinoma admitted in Medical College Hospital Trivandrum during 2011 to 2015 were collected and studied.

Results

Of the 59 patients 30 were females and 29 were male and the mean age was 62 years. 21(36%) patients presented with pain and 44(75%) patients had jaundice at presentation. 35(59%) patients had asthenic symptoms- loss of appetite and loss of weight. 11(19%) patients had co existing chronic liver disease and 5(9%) patients had co existing gall stone disease.

CA 19-9 was elevated in 53(90%) patients. All patients took USG abdomen and 52 (88%) patients took CECT abdomen. USG mainly showed biliary obstruction and CECT abdomen was more of a diagnostic investigation.

There were total 39(66%) hilar, 13(22%) distal and 7(12%) intrahepatic cholangiocarcinomas. 20 (34%) patients had distant metastasis at presentation itself. Male sex was found to be protective for metastasis (P value 0.033). Ascitis was significantly associated with metastasis at presentation (P value 0.038).

There was no major difference in clinical features when comparing the three types. Anemia (P value 0.006) and hypoalbuminemia (P value 0.042) were significantly associated with Hilar type. Only (7%) patients underwent curative resection and rest of the patients mainly underwent only palliative interventions like percutaneous transhepatic biliary drainage in 16(27%) patients and endoscopic stenting in 6(10%) patients.

All the four patients who underwent curative surgery were having hilar cholangiocarcinoma with three patients having Bismuth Corlette type IIIb and one patient with type IIIa. All patients underwent hepatectomy for clearance of the tumor. In histopathology all patients had well differentiated adenocarcinoma and all were of sclerosing type. Only 2 paients had R0 resection.

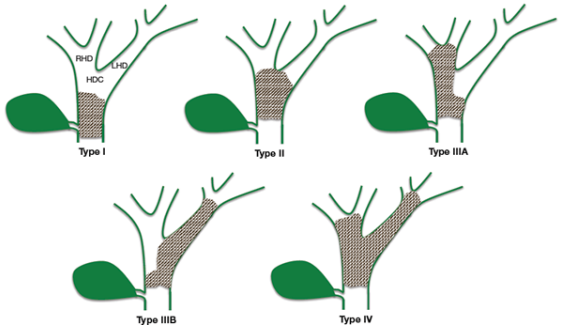

Figure 1- Bismuth Corlette Classification of cholangiocarcinoma

Features	Hilar(39)	Distal(13)	Intrahepatic(7)
JAUNDICE	28(72%)	12(92%)	4(57%)
PAIN	14(36%)	3(23%)	4(57%)
ANEMIA	22(56%)	3(23%)	0
HYPOALBUMINEMIA	17(44%)	12(92%)	4(57%)
CEA	12(31%)	2(15%)	1(14%)
METASTASIS	13(33%)	6(46%)	1(14%)
SURGERY	4(10%)	0	0

Table 1- comparison of the three types of cholangiocarcinoma

Discussion

Cholangiocarcinomas are malignant tumors arising from the epithelial lining of the biliary tract. They are rare tumors but their incidence is increasing with cholangiocarcinoma accounting for 1-2% of new carcinomas detected[1].

They are classified as extrahepatic cholangiocarcinoma and intrahepatic or peripheral cholangiocarcinoma. Extrahepatic cholangiocarcinoma includes hilar cholangiocarcinoma or

Klatskin's tumor and distal cholangiocarcinoma.

The etiologic factors identified are primary sclerosing cholangitis, congenital biliary cystic disease, hepatolithiasis, parasites like chlonorchis, and various teratogens like thorotrast, asbestos, nitrosamines and genetic causes[2.]

Patients often have non specific complaints and extrahepatic cholangiocarcinoma usually presents with jaundice and intrahepatic type most commonly presents with right hypochondrial pain. Asthenic symptoms are very common. Patients rarely presents with cholangitis unless prior biliary intubation was done[3.]

There are 3 pathologic types- sclerosing(most common), nodular and papillary type. The disease tends to extend via submucosal route with associated perineural invasion. 2 pathological factors that influence the prognosis highly are complete R0 resection and absence of lymph node metastasis.

The AJCC TNM staging is based on pathology and it doesn't correlate with survival. The Bismuth Corlette classification is anatomically oriented and it doesn't say potential for resection. The Jarnagin's staging for hilar cholangiocarcinoma is based on imaging and it correlates well with resectability and survival.

The investigations employed are LFT, CA 19-9 which is elevated in 80% of cases, USG abdomen as the first imaging, CECT abdomen, MRCP and EUS. Tissue diagnosis prior to surgery is unnecessary. Bile cytology and brushings are unreliable[4.]

Treatment depends on site of disease and extent. The only hope for long term survival is surgery. Only 10-15% cases are suitable for surgery. Resectability rate increases with more distal the location of the tumor. The aim of surgery is to achieve negative resection margin and to restore biliary enteric continuity. Surgery may include hepatectomy for clearance. Distal tumors may require pancreaticoduodenectomy. Preoperative biliary drainage maybe beneficial. Adjuvant chemotherapy or radiotherapy has no proven survival benefit but are employed if there is high risk of recurrence as in positive margins[5.]

In unresectable cases palliation may be necessary for pain, jaundice or duodenal obstruction. Surgical palliation doesn't prolong survival or decrease complications. Palliative options are biliary endoprosthesis, endoscopic or percutaneous biliary drainage, hepatojejunostomy or choledochojejunostomy and percutaneous destruction of celiac plexus for uncontrolled pain.

Since the disease gets detected late the outcome is generally poor with unresected median survival of only 5-8 months and median survival of 18 months after resection.

Conclusion

Most patients in our series presented at an advanced stage of disease and curative resection could be offered only for 7% of the patients which is very less compared to previous studies where 10-15% patients were found to be suitable for surgery. This is an important concern as the only chance for cure in cholangiocarcinoma is surgery and hence more studies are urgently required to focus on early detection and better treatment of this condition.

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