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Neuroendocrine Papillary Carcinoma of Gallbladder with Hepatic Space Occupying Lesion – A Rare Case Report in Chattogram, Bangladesh

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Abstract:

Introduction:

Neuroendocrine tumors, commonly originating from neuroendocrine cells of different organs, specifically in gastrointestinal tract (GIT), lungs and thyroid. However, the tumors which originates from gall bladder are not only very rare, but also very aggressive as a neoplasm.

Case presentation:

A patient named Mrs. Khodeza aged 35 years old, with a complain of abdominal pain was diagnosed as neuroendocrine papillary carcinoma of gall bladder by magnetic resonance cholangiopancreatography (MRCP) along with histopathological exclusion. Patient received 4 cycles of Cisplatin and Etoposide followed by radiotherapy.

Conclusion:

Treatment with Cisplatin and Etoposide has demonstrated significant clinical response with satisfactory remission of the disease. In addition, adverse effects are minimal. At present, patient is leading a healthy life.

Keywords: Neuroendocrine, papillary, carcinoma, gallbladder, hepatic, lesion

BACKGROUND

Biliary tract cancers are one of the most life-threatening cancers which arises from gallbladder and intrahepatic and extrahepatic (hilar and distal common bile duct). Among other cancers of biliary tract, gall bladder cancer is the most common cancer. Elderly aged persons are in high risk of gall bladder cancer (GBC). Moreover, it is evident that, women have two to sixth time chance of affecting with GBC comparing to men [1].

In early stage of GBC, as there is no specific symptom, the diagnosis of the disease is done in an advance stage. non-

specific abdominal pain, weight loss, anorexia, and obstructive jaundice (which is a sign of advanced disease) are the common symptoms [2]. Chole-lithiasis (especially mixed gall stone), chronic infections of the gallbladder, obesity, sedimentary life style, hepato-biliary anomalies and environmental exposure to specific chemicals are the associated predisposing factors of GBC.

Bacterial degradation of bile and prolong chronic inflammation exaggerates the whole carcinogenic process.

Laparotomy is the treatment of choice for all the cases GBC or suspected GBC. Simple cholecystectomy is the treatment of choice in case of early gall bladder cancer. Radical cholecystectomy along with partial hepatectomy and regional lymphadenectomy is the management option for advanced advanced-stage carcinomas without distant metastases [3].

On the other hand, being the third cause of cancer related death, the prevalence of hepatocellular carcinoma (HCC) is expected to be increased in upcoming decades. Although treatment decision depends of hepatocellular carcinoma (HCC) staging, most major staging of HCC is followed by hepatic resection. Moreover, the vital route which develops hepatic metastasis, arises from gall bladder carcinoma as well as is entrained the portal tracts after direct hepatic invasion [4].

Furthermore, a simple cholecystectomy has evidence of 100% cure rate for tumors confined to muscular layer of gallbladder majority of stage II patients are failed with the treatment of simple cholecystectomy. Extended cholecystectomy, including resection of hepatic segments IV b and V, and an extensive lymph node dissection of the porta hepatis, posterior pancreaticoduodenal, and inter-aortocaval lymph nodes for stage II gallbladder cancer has 90-100% of 3- years survival rate [5]. Hepatic invasion more than 20mm is managed by extensive hepatic resection [6].

Of all the type of GBC, neuroendocrine gallbladder carcinoma is highly aggressive and rare one. It originates from neuroendocrine cells and secreting peptides as its neurotransmitter. Carcinoid or typical carcinoid (low malignancy), atypical carcinoid (moderate malignancy) and small cell carcinoma (high malignancy) are the three pathological classification of neuroendocrine gallbladder carcinoma. In spite of having poor prognosis, the most effective and choice of treatment of neuroendocrine gallbladder carcinoma is radical resection followed by local liver resection and lymph node clearance [7].

CASE STUDY

A patient named Mrs. Khodeza aged 35 years old, Bangladeshi female, visited to a surgeon in the year of 2022 with the complain of severe abdominal pain as well as pale stools and dark urine. Physician advised her to do CBC, urine for R/M/E, and magnetic resonance cholangiopancreatography (MRCP). On MRCP examination, hepatic space occupying lesion (SOL) was found and might suggest focal nodular hyperplasia, The mass partially encasing the gall bladder and compressing the proximal common bile duct, resulting in mild intrahepatic biliary obstruction. The surgeon underwent with obstructive hepatic resection along with cholecystectomy. Patient was stable after operation and suggested to do histopathological examination.

After that, she was referred to an oncologist. The oncologist suggested her to do immunohistochemistry of resected tissue for histopathological confirmation. On histopathological report, intra-cholecystic papillary neoplasm along with high grade dysplasia with neuroendocrine carcinoma was diagnosed, followed by liver tissue involvement by tumor. Before starting the treatment, the doctor advised her some routine tests including CBC, serum creatinine and serum bilirubin. All the findings were found normal.

TREATMENT

Patient was advised to give Cisplatin 100mg with Etoposide 400mg followed by radiotherapy. The patient received her first chemotherapy on 15/12/2022. In the whole treatment journey, patient received 4 cycles of chemotherapy. The patient received her last chemotherapy on 7/01/2023. During treatment with these regimens, no major side effect was recorded.

PATIENT OUTCOME

Patient was stable during the whole treatment period. No major side effect due to drugs was found. After radiotherapy, CT scan report showed complete response of this disease.

DISCUSSION

Neuroendocrine carcinoma of the gall bladder is such an uncommon and aggressive disease along with an unsatisfactory prognosis.

In the year of 2013, a 70 years old female patient with the complaint of right upper abdominal pain, was diagnosed with gallbladder mixed neuroendocrine neoplasm similar to our case. After identifying gallbladder mass by Contrast-enhanced computed tomography (CT) scanning and magnetic resonance imaging (MRI), open cholecystectomy was advised. Two years later, hepatic metastasis along with obstructive jaundice was found. Patient was suggested to do endoscopic retrograde

cholangiopancreatography (ERCP). A stricture in the middle extrahepatic bile duct was found and further managed with biliary stenting. On histopathological finding, about 65% large cell neuroendocrine carcinoma (LCNEC) and 35% moderately differentiated papillary adenocarcinoma were found. Unfortunately, three months later, the patient died due to systemic organ failure [8].

In another similar study, a 66 years old female patient with no significant symptoms like fever or abdominal pain, underwent abdominal ultrasonogram followed by Contrast-enhanced computed tomography (CT) and magnetic resonance imaging (MRI) to exclude a mass in the fundus of the gallbladder. This suspected gallbladder cancer patient was advised to do cholecystectomy with regional lymph nodes dissection. adenocarcinoma and neuroendocrine components in the tumor were confirmed by histopathological examination. After operation, the physician started chemotherapy for 5 months. However, a local recurrency occurred after 14 months of operation [9].

Overall, the recurrence of neuroendocrine papillary carcinoma of gall bladder is very frequent according to these cases the prognosis is not so satisfactory.

CONCLUSION

The patient is at present leading a normal and healthy life. The adverse effect of cisplatin and etoposide followed by radiation was minimum in this case. The prognosis was satisfactory. At present patient is under regular follow up.

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