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Klatskin Tumor: A Rare Case with Review of Literature

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Abstract: ***Klatskin tumour is a very rare malignancy of the extrahepatic bile duct, the annual incidence being1:1,00,000. Because of its extremely rare incidence, the tumour is difficult to investigate. We present a case report of a 65 year old female patient who came to the outpatient department with chief complaints of abdominal pain, weight loss and later was diagnosed with Klatskin tumour on histopathological examination.***

Keywords: Adenocarcinoma, Bile duct, Cholangiocarcinoma, Klatskin tumour

1. Introduction

Klatskin tumour is a cholangiocarcinoma which arises at the confluence of right and left hepatic ducts at liver hilus. The annual incidence rate is 1 : 1,00,000[1]. The tumour is named after Gerald Klatskin, who first described it[2]. 25% of these cases are associated with biliary cystic diseases, like choledochal cysts or Caroli's disease, these being more invasive than the others[3].Klatskin tumour is a rare type of adenocarcinoma of the extra hepatic bile duct with a dismal prognosis. Until now, only a few cases pertaining to Klatskin tumour have been published.

We report a case of Klatskintumor in a 65 year old female patient to highlight its rare occurrence.

2. Case Report

A 65 year old female patient presented to the outpatient department with chief complaints of jaundice, pruritus, weight loss and generalised weakness since 1 month. The patient gave history of dark coloured urine and dull aching pain in the right upper quadrant of abdomen since a month. The patient had no prior history of chronic diseases like diabetes mellitus, hypertension, bronchial asthma or tuberculosis. Also, the patient was not a known case of Hepatitis B. Liver function tests were deranged with markedly elevated alkaline phosphatase levels of 407 IU/L and total bilirubin levels of 2.2 mg/dl. Imaging workup included Computed Tomography scan of the abdomen and pelvis which revealed ill definedheterogenously enhancing mass lesion in perihilar region arising from common hepatic duct with enhancement pattern and upstream dilatation of the biliary system. Periportal lymph nodes revealed metastasis.

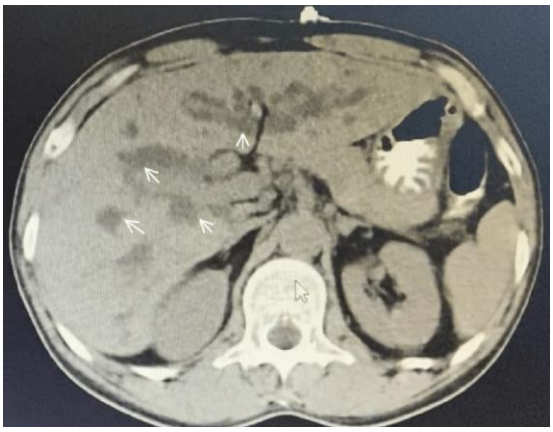

Figure 1: CT scan of the abdomen and pelvis

CT scan imaging revealedill defined heterogeneously enhancing mass lesion in perihilar region arising from common hepatic duct with enhancement pattern and upstream dilatation of biliary system.

Surgical exploration was done and the tumour was sent for histopathological examination.

Gross examination revealed an irregular, grey white to grey brown tumour totally measuring 1.8 x 1.0 x 0.5 cm.Cut section of the tumour was grey white to grey brown.

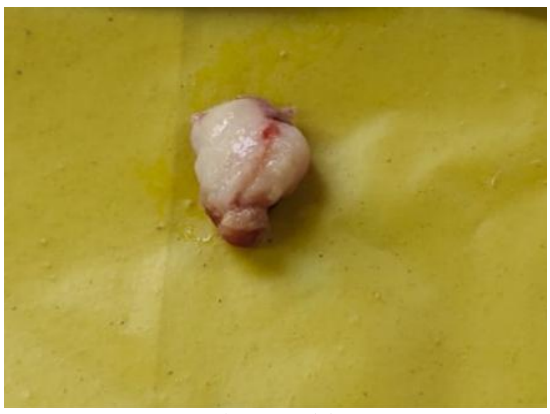

Figure 2 (a)

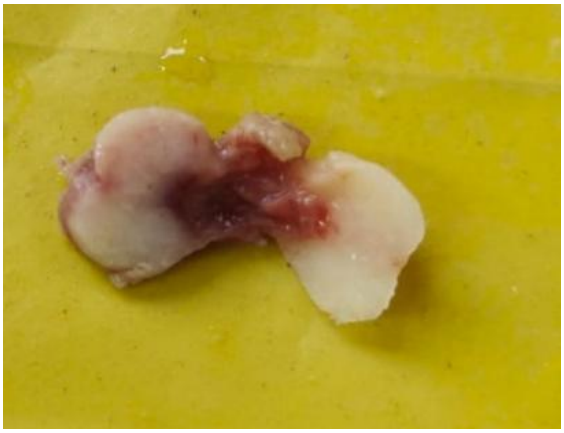

Figure 2 (b)

Figure (2a) and (2b): Gross examination revealed an irregular, grey white to grey brown tumour, cut section of the tumour tissue being grey white to grey brown.

Microscopy revealed moderately differentiated adenocarcinoma showing glandular pattern. Individual tumour cells were cuboidal to columnar with enlarged vesicular nuclei, prominent nucleoli and moderate amount of cytoplasm. Stroma revealed desmoplastic response and inflammation with areas of tumour necrosis.

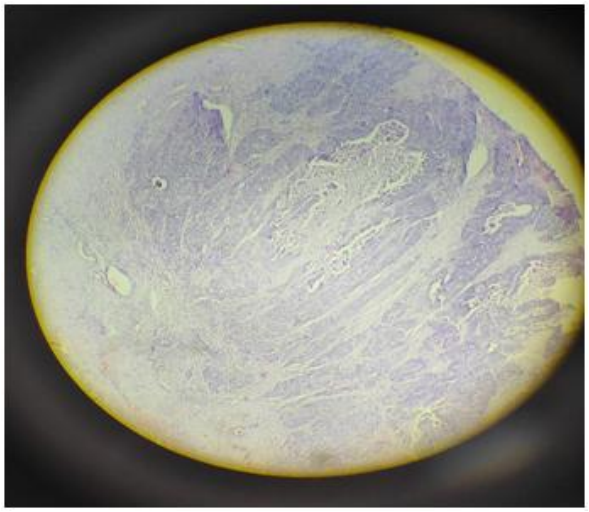

Figure 3 (a):100X H&E

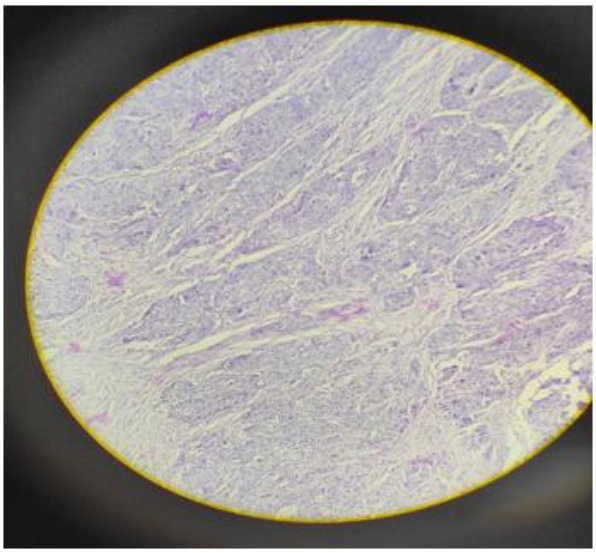

Figure 3 (b): 400X H&E

Figure 3a (100X H&E) and 3b (400X H&E) : Photomicrographs of Klatskin tumour revealing moderately differentiated adenocarcinomashowing glandular pattern, with individual tumour cells being cuboidal to columnar, having enlarged vesicularnuclei, prominent nucleoli and moderate amount of cytoplasm. Stroma shows desmoplastic response and inflammation with areas of tumor necrosis.

3. Discussion

Klatskin tumour is a very rare type of adenocarcinoma of the extrahepatic bile duct which arises from the intersection of the right and left lobe of the hepatic duct forming common bile duct[4]. Risk factors include alcoholism, Hepatitis B viral infection, primary sclerosing cholangitis, liver fluke infection, intrahepatic bile duct stones, Familial Polyposis Coli, chronic typhoid carrier states and thorotrast exposure[5]. Most of Klatskin tumours are sporadic, as seen in our case[5]. Most common clinical presentation of this malignancy includes painless, progressive jaundice with one third cases presenting with gall stones[6]. Imaging modalities assist in the initial diagnosis of these tumours. Klatskin tumours, on imaging, commonly present as ill defined masses with hilar necrotic lymph nodes and hepatic metastatic deposits. These tumours are also poorly differentiated on microscopy and spread along the duct and nerve sheath[7]. Klatskin tumours commonly present in age group above 60 years of age, which is similar to our case[8]. These tumours are small at the time of diagnosis, spread being local to the ampulla of Vater, colon, duodenum, gall bladder, liver, omentum, stomach and pancreas[9]. The diagnosis is poor, about 28-89% cases showing positive margins[9]. Laboratory investigations in these cases commonly show increased alkaline phosphatase levels and increased serum bilirubin levels. Favourable prognostic factors include low stage and papillary pattern whereas unfavourable prognostic factors comprise of high grade and high stage tumours, positive surgical margins, hilar tumours, presence of tumour necrosis and nuclear atypia[10]. Positive immmunostains include mucin, carcinoembryonic antigen (CEA), CK7, P-cadherin, CD_{24}, EGFR[11]. Differential diagnosis of Klatskin tumours are hepatocellular carcinoma, cholangiocarcinoma and metastatic carcinoma.

The only curative treatment is complete surgical resection with histologically negative margins. The median survival of inoperable patients is 6 to 12 months and the commonest cause of death is liver failure followed by sepsis[12]

4. Conclusion

Klatskin tumour is a rare malignant tumour with survival chances of the patient being abysmal. Histopathological examination is crucial for accurate diagnosis and optimal management.

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