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Case Report

PROXIMAL FEMORAL METASTASIS WITH PATHOLOGICAL FRACTURE IN CHOLANGIOCARCINOMA: CASE REPORT

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A B S T R A C T

Tumors of the biliary tract are rare, and their survival prognosis is short since diagnosis is often made at advanced stages of the disease. Diagnosis remains difficult because symptoms are often unspecific. Cholangiocarcinoma most commonly advances locally and regionally by invading the lymph nodes. In rare cases, it has been noted that cholangiocarcinoma can metastasize to bone, with a preponderance for the axial skeleton. Herein, we describe what we believe to be the first clinical report of a proximal femoral bone metastasis with multiple vertebral and pelvic metastasis with pathological fractures of neck of femur, D5, L4 and L5 vertebra from metastatic cholangiocarcinoma. We present a case of middle aged patient with a severe and pro¬gressive pain due to pathologic fractures of femoral neck, D5, L4 and L5 vertebra. The medical investigations revealed the presence of an extrahepatic cholangio-carcinoma.

INTRODUCTION

Cholangiocarcinoma (CCA) is a rare malignant neoplasm of biliary tract epithelium in the intra- and extrahepatic bile ducts, which accounts for less than 2% of all human malignancies.[1] It originates from the liver (intrahepatic), at the confluence of the right and left hepatic ducts (hilar) or in the extrahepatic bile ducts. After hepatocellular carcinoma it is the second most common primary hepatic malignancy.[2] Over the past three decades, the worldwide incidence of and mortality from cholangiocarcinoma have steadily increased.[3] These cancers are locally advanced at presentation, having high mortality rate.[4] Cholangiocarcinoma most commonly advances locally and regionally by invading the lymph nodes.[5] In rare cases, it has been noted that cholangiocarcinoma can metastasize to bone, with a preponderance for the axial skeleton.[5] Only a few case reports have described long bone metastasis.[6,7,13,14] There are several established risk factors for CCA, including parasitic infections, primary sclerosing cholangitis, biliary-duct cysts, hepatolithiasis, and toxins. Other less-established potential risk factors include inflammatory bowel disease, hepatitis C virus, hepatitis B virus, cirrhosis, diabetes, obesity, alcohol drinking, tobacco smoking, and host genetic polymorphisms. Greater than 90% of bile duct cancers are well-differentiated and mucin-producing adenocarcinomas. Surgical resection has been the mainstay of curative treatment for CCA.

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Unfortunately, many patients present with unresectable tumors. This cancer frequently metastasizes to lungs, adrenal glands, brain, lymphatic system and the axial skeleton.[8] Palliative treatment including bypass surgery, endoscopic or percutaneous stenting, photodynamic therapy, intraluminal brachytherapy, external radiation and systemic therapy, remain the gold standard of treatment for these patients.[9] Cholangiocarcinoma has been shown to be resistant to com-mon chemotherapy. For which numerous drugs have been tested alone and in combination. The response rate has been unacceptably low. Gemcitabine has been suggested as an alternative for patients with unresectable CCA.[10] In the present report, we describe the first clinical report of proximal femoral bone metastasis and multiple vertebral and pelvicmetastasis from metastatic extra hepatic cholangiocarcinoma.

Case report

A 35 year old female patient presented with progressive increasing pain in left hip, middle back and progressive paraplegia without bowel and bladder involvement for last 10 days following trivial trauma. She also had fatigue, anorexia with significant weight loss. On radiographic evaluation she was found to have fracture neck of femur left side (Fig. 1). MRI evaluation of spine suggested of compression collapse fracture of D5 vertebra with associated epidural soft tissue component causing severe cord compression and early myelopathy changes along with compression fracture of L4 and L5 vertebra and altered marrow signal changes in sacrum and bilateral iliac bones (Fig. 2). A provisional diagnosis of

pathological fracture of neck of left femur with multiple vertebral fractures with secondary metastatic deposits was made and patient was further evaluated. Complete blood count revealed anemia. LFT (liver function test) revealed bilirubin 4.2, AST (aspartate transaminase) 76, ALT (alanine transaminase) 52, ALP (alkaline phosphatase) 1280. KFT (kidney function tests) was within normal limits. Contrast enhanced computed tomography (CECT) chest, abdomen and pelvis revealed luminal obstruction and enhancement within proximal and midpoint of common bile duct, diffuse gall bladder thickening with multiple heterogeneously enhancing lesions in the liver (Fig. 3). Ultrasound guided biopsy of the liver deposits revealed malignant tumor cells arranged in vague glandular pattern consistent with metastatic deposits from cholangiocarcinoma (Fig. 4). 99m technetium bone scan revealed increased uptake in left parietal bone, left scapula, D5-D10, L4-L5 vertebra, left ischium, left acetabulum, bilateral sacroiliac joints and left proximal femur (Fig. 5). A diagnosis of metastatic extrahepaticcholangiocarcinoma was made. Patient general condition deteriorated over short period of time as the patient came in advanced stage. She was unfit for any kind of orthopedic surgery for bony fixation. She was given 3 cycles of chemotherapy (4 gm zoledronate and 6 mg dexamethasone, 1650 gm gemcitabine and 1000mg carboplatin). There was modest decrease in pain but patient general condition deteriorated and she succumbed to her illness after 10 days of her third chemotherapy cycle.

DISCUSSION

Tumors of biliary tract are rare. The incidence has been reported to be 1 to 2 per 100,000.[4] Cholangiocarcinoma (CCA) is a lethal cancer of biliary epithelium. Cholangiocarcinoma arises from the malignant proliferation of cholangiocytes, the epithelial cells lining the biliary tree. They are characterized by a bad prognosis and poor response to current therapies. CCA may emerge at any portion of the biliary tree. CCA is currently classified as intrahepatic (IH-CCA) or extrahepatic (EH-CCA), the second-order bile ducts acting as the separation point.[11] The EH-CCA is comprised of the perihilar form (Klatskin tumor) and distal form where the separation point being posed at the level of the cystic duct. Intrahepatic cholangiocarcinomas initially present as mass-forming lesions, and obstructive symptoms are rare.In contrast EH-CCAs usually present with signs and symptoms of biliary obstruction (ie, painless jaundice, pruritus, pale stools, and dark urine). The major pattern of metastasis of cholangiocarcinomas is early lymphatic spread. Unfortunately, many patients present with unresectable tumors, the majority of whom die within a year of diagnosis. Weight loss and abdominal pain, when present, are usually manifestations of advanced, unresectable disease. Metastasis is frequent and occurs to many organs but less frequent to the bone. Most patients with bone metastasis suffer from pain resulting from structural damage, periosteal irritation, and nerve entrapment. Because of continuous, severe and progressive nature of metastatic bone pain, it is difficult to treat. However recent trial confirmed the benefits of zoledronic acid on pain and quality of life in elderly patients with bone metastasis from solid tumors.[12] The development of metastatic lesion of long bone is rare and only three case reports of humerus and one case report of fibula is there in medline search as per our knowledge.[6,7,13,14] However this is the first report that describes a case of cholangiocarcinoma that features with the severe pain due to the pathologic fracture of neck of femur with fracture of D5, L4 and L5 vertebra with paraplegia without bowel and bladder involvement. This report confirms that the possibility of long bone metastasis should be considered in patients with a history of cholangiocarcinoma.

In conclusion, cholangiocarcinoma is an aggressive carcinoma that, although rare, has the ability to metastasize to bone. Although more commonly seen in central locations, it can very rarely metastasize to long bone with multiple vertebra and pelvis and it should be considered in patients who present with hip pain and a liver lesion, or a history of cholangiocarcinoma.

Figures

Fig 1 Plane Antero-posterior radiograph of pelvis with both hips revealed pathological fracture of neck of left femur

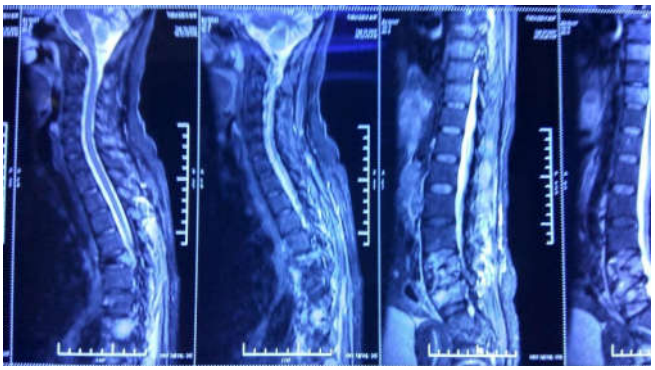

Fig 2 MRI(magnetic resonance imaging) scan of spine revealed compression collapse fracture of D5 vertebra, L4 and L5 vertebra and altered marrow signal changes in sacrum and bilateral iliac bones

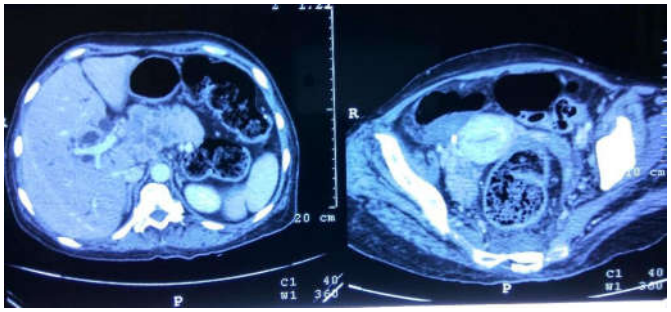

Fig 3 Abdominal Computed tomography (CT) scan revealed several metastatic lesion in the liver parenchyma

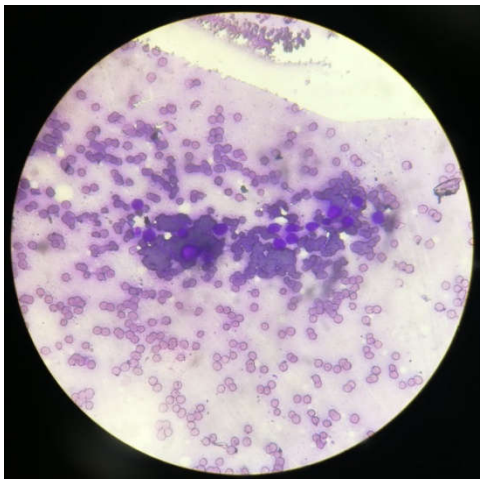

Fig 4 Photomicrograph showing clusters of tumor cells arranged in vague glandular pattern, large round to polygonal tumor cells with scant cytoplasm and few scattered hepatocytes in background (hematoxylin and eosin stain)

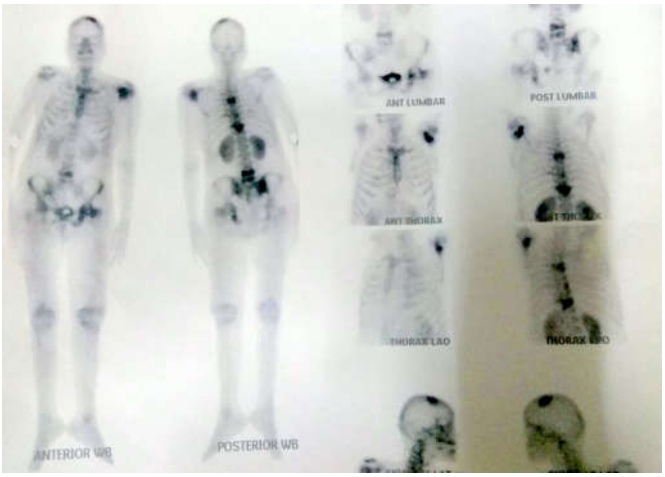

Fig 5 99m technetium bone scan revealed increased uptake in left parietal bone, left scapula, D5-D10 ,L4-L5 vertebra, left ischium, left acetabulum, bilateral sacroiliac joints and left proximal femur

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