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Case Report

Rare cause of biliary obstruction from myeloid sarcoma has better prognosis, if timely diagnosed: case report with literature review

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ABSTRACT

Extramedullary myeloid sarcoma (EMMS) involving the biliary tract is extremely rare. We describe, a case of a 40-year-old gentleman who presented with obstructive jaundice and features of malignant biliary stricture on ERCP and MRCP. Histopathology revealed myeloblasts, while peripheral blood and bone marrow did not reveal any evidence of leukemia.

Keywords: Extramedullary myeloid sarcoma, Biliary stricture, Obstructive jaundice

INTRODUCTION

EMMS also known as chloroma, a rare extramedullary tumor composed of immature cells of the myelomonocytic series which can occur anywhere in the body.[1] Myeloid Sarcoma (MS) may occur de novo or concurrently with Acute Myeloid Leukemia (AML) or a myeloproliferative disorder (MPD).[1] MS is reported in 2-8% of patients with AML, and can even occur as initial manifestation of relapse in a patient treated for AML, or after allogenic stem cell transplant.[1,2] Both sexes are equally affected with a mean age of 48 years (range 1-81).[1] MS often involves the skin, lymph nodes and bones.[1] Uncommon sites of MS occurrence reported are in the brain, orbit, perineum, GI tract, vagina, female gynecological tract, prostate, lung mediastinum, urinary bladder and other organs. MS involving biliary tract is also a rare occurrence.[3-11] A large proportion of EMMS are initially misdiagnosed.[12] EMMS involving gallbladder is very rare. It is very important to distinguish a case of gallbladder myeloid sarcoma from carcinoma, as the diagnosis of former is challenging and can lead to delay in management. MS is frequently mistaken for non-Hodgkin's lymphoma (NHL), small round cell tumor (neuroblastoma, rhabdomyosarcoma, Ewing sarcoma/ PNET, medulloblastoma) and undifferentiated carcinoma. The diagnosis is missed in about 50% of cases when Immunohistochemistry (IHC) is not used.[13]

MS may precede, accompany or complicate the course of AML. They may pose a diagnostic dilemma when detected in the absence of typical manifestations of AML. Histology, touch imprint cytology, cytochemistry, IHC, electron microscopy and molecular studies are all useful tools in making a definitive diagnosis. Most EMMS left untreated, will progress to AML and those initially treated with chemotherapy show longer disease-free intervals and overall survival.[13]

Herein, we present a case of myeloid sarcoma occurring in the gallbladder with the involvement of common hepatic duct (CHD) and confluence causing obstructive jaundice. On initial evaluation, it appeared as malignant biliary stricture and the final diagnosis could be arrived at only after surgery and IHC analysis.

CASE REPORT

A 40-year-old gentleman presented with obstructive jaundice of one month duration. His hematological profile revealed hemoglobin of 10.6 g%, Total Leucocyte Count-25,190 cells/mm3. Peripheral smear suggestive of neutrophilic leukocytosis with toxic granules and normochromic red blood cells with adequate platelets. Biochemical parameters showed a total bilirubin of 5.5 mg/dl with direct component being 2.8 mg/dl, ALP-367 IU/l, GGT-103 IU/l, serum albumin-1.4 gm/dl. Ultrasound abdomen revealed hepatomegaly with multiple cholangiolytic abscesses. Patient further underwent ERCP with findings of CHD stricture, dilated to 12F and stented (Figure 1). Brush cytology taken, revealed no malignant cells. After 4 weeks, when reviewed with CT scan abdomen, it revealed multiple cholangiolytic abscess in resolution in the right lobe of liver. MRCP revealed CBD stent in right hepatic duct, 6mm smooth stricture of CHD with mild circumferential thickening, multiple small abscesses and no significant enhancement/mass lesion; no evidence of ascites or lymphadenopathy (Figure 2).

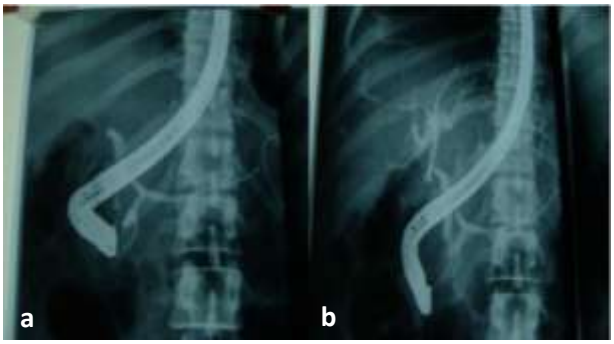

Figure 1: ERCP showing (a) tight CHD stricture and (b) status post stenting.

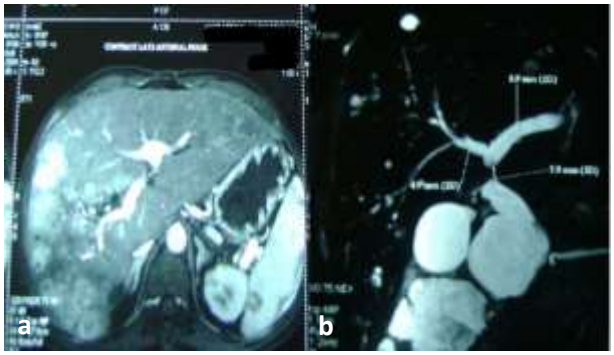

Figure 2: MRCP showing (a) multiple small ill-defined T2 hyper dense parenchymal lesions with intense peripheral rim enhancement in late arterial phase suggesting abscesses rather than secondaries, (b) stricture involving CBD with mild mural thickening but no significant enhancement/mass lesion.

Patient underwent laparotomy for malignant biliary stricture. Intra operatively, friable growth in the neck of gallbladder invading mid CBD and hilum, distended gallbladder with mucocele and cholangiolytic abscess in right liver lobe (Figure 3-4). Frozen section of gallbladder and liver lesion were suggestive of lymphoma with plasma cell rich infiltrate. Patient had and uneventful post-operative recovery and was followed up to 12 months post operatively. Histopathology revealed a monomorphous and discohesive infiltration of tumour cells in submucosal layer of gall bladder and along the bile duct wall. Tumour cells were of medium size with large nuclei showing dispersed chromatin, and one or more distinct but not prominent nucleoli. The cytoplasm was deeply eosinophilic and granular reminiscent of metamyelocytes, and indicative of myeloid differentiation.

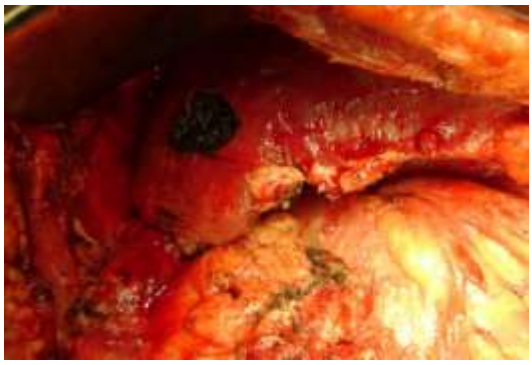

Figure 3: Intra-operative picture showing liver surface with multiple cholangiolytic abscesses lesions mimicking secondaries; an excised lesion site taken for frozen section.

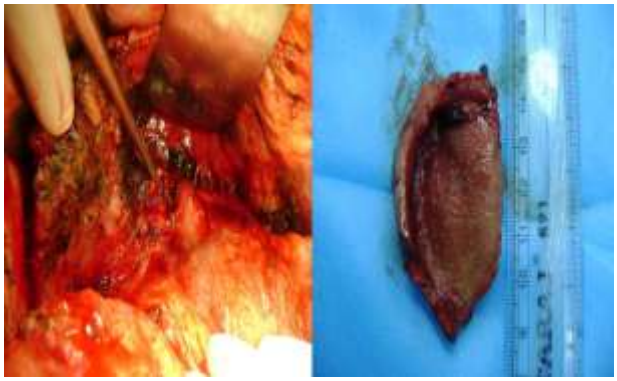

Figure 4: Intra-operative picture showing gall bladder neck mass encroaching bile duct and resected specimen.

IHC showed positively for (Myeloid lineage) – LCA+, MPO+, BCL-2+, MIB-1+, but negative for (B/T cell lineage derived lymphoma esp. Burkitt's ruled out by) – CD-20, CD-3, CD-33. Epithelial derived MALT ruled out by – CD-138, CYCLIN-D1 (Figure 5). The diagnosis of EMMS was made. Patient underwent bone marrow aspiration and trephine biopsy, which showed normal trilineage hemopoiesis with normal blast count and was planned for adjuvant chemotherapy (Figure 6).

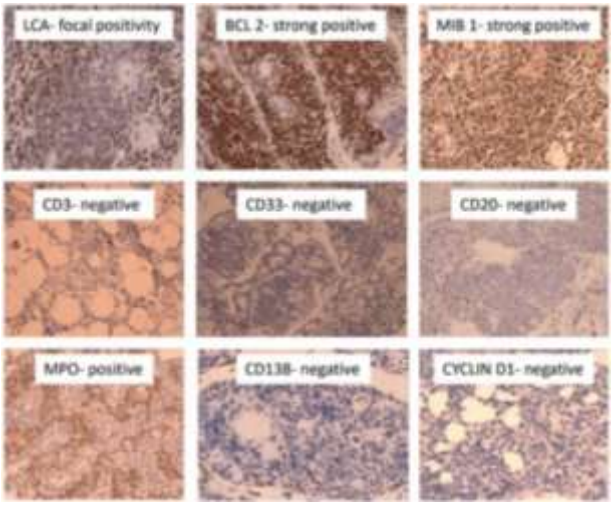

Figure 5: Immune-histochemical markers with – Myeloid lineage - LCA+ , MPO+ , BCL-2+ , MIB-1+ B/T cell lineage derived lymphoma esp. Burkitt's ruled out by – CD-20, CD-3, CD-33, epithelial derived MALT ruled out by – CD-138, CYCLIN-D1.

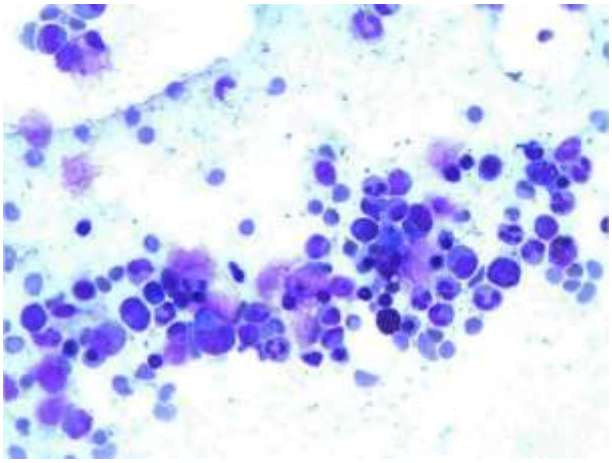

Figure 6: Bone marrow aspiration smear with normal elements.

DISCUSSION

EMMS is a rare neoplasm wherein extramedullary proliferation of myeloblasts or immature myeloid cells form tumor masses and disrupt the normal architecture of the tissue.[11] MS usually occurs in one of the following three clinical settings. The first with previous or current AML, second as a sign of blast transformation of CML or as some other MPD and third with no other clinical evidence of hematologic disease at the time of diagnosis. Our patient, had no hematological abnormalities. Diagnosis was never possible to do preoperatively as clinical suspicion for malignant biliary stricture of Klatskin's type was encroaching on to CHD and confluence. This final diagnosis was possible after surgery because of availability of tissue for IHC markers.

MS is described in association with a variety of chromosomal abnormalities in particular, t (8; 21) (q22; q22) and inv (16).[15] An IHC panel including CD43, MPO, CD117, CD68 (or CD163), CD3 and CD20 can successfully identify the majority of MS in formalin-fixed, paraffin-embedded tissue specimens.[1,15]

In our case, B-cell and T-cell lymphomas were excluded by negative stains for CD20, CD79a and CD3; Burkitt lymphoma excluded by negative immunoreactivity for B-cell antigens. We could not start chemotherapy until tissue diagnosis made; which required laparotomy and IHC. This was shown that if diagnosis and treatment (chemotherapy) started early, can avoid a major surgery.[16]

The correct diagnosis of MS is often delayed because of a high misdiagnosis rate.[12] Previous studies have indicated that the biologic behavior is dramatic irrespective of presentation, age, sex, phenotype and cytogenetics.[15] Untreated isolated MS ultimately transforms to AML typically over a 10-to-12-month period. MS should be treated as AML, with systemic chemotherapy. Disease progression and prognosis is same as of that of AML and median survival is approximately 22 months, with worse outcomes more common in elderly patients.[17]

CONCLUSION

A diagnosis of biliary MS is difficult to make by radiographic imaging. It may be misdiagnosed as cholangitis, cholecystitis or malignancy (as in our case). Biliary MS reported previously, and the current case shows that it commonly presents with extrinsic bile duct compression. Therefore, biliary MS should be included in the differential diagnosis of obstructive jaundice and its diagnostic workup should include IHC.

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