and have a predilection for areas of weakness in the chest wall. These areas occur anteriorly from the costochondral junction to the sternum because of the absence of external intercostal muscles and posteriorly from the costal angle to the vertebra as a result of the absence of internal intercostal muscles.4

The diagnosis can be made with a palpable defect in the thoracic wall through which a reducible soft tissue mass appears. The contents can be ascertained by observing that the containing lung varies in size paradoxically with respiration and is increased by Valsalva maneuver.2 An increase in hernia size with inspiration and a decrease with expiration occur when there is a diaphragmatic injury with prolapse of abdominal viscera into the thorax and out through the chest wall.2 The chest radiograph may reveal divergent ribs with bowel gas shadows beyond the confines of the abdominal cavity. CT scan confirms the diagnosis.5

Surgery can be performed by an incision over the hernial sac, which can be extended as a thoracoabdominal incision if adhesions are present. Diaphragmatic injuries can be repaired by primary closure or by a prosthetic mesh. Intercostal muscles are usually attenuated and need reinforcement by a prosthetic mesh.

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Acute myeloid leukemia presenting as obstructive jaundice

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We report a 32-year-old man with acute myeloid leukemia presenting as obstructive jaundice. Imaging revealed dilated common bile duct with abrupt narrowing at the lower end, distended gall bladder, and dilated intrahepatic biliary radicles. In addition he had a mass lesion in the urinary bladder. On evaluation he was found to have the eosinophilic variant of M4 subtype acute myeloid leukemia. He expired before chemotherapy could be instituted. [Indian J Gastroenterol 2006;25:93-94]

Jaundice in acute myeloid leukemia (AML) is most often the result of treatment-related hepatocellular damage. We report a patient with acute myeloid leukemia who presented with obstructive jaundice because of common bile duct obstruction.

A 32-year-old man was referred with history of progressively increasing yellow discoloration of eyes and urine for 2 months and generalized pruritus. He also complained of anorexia and had lost 8 Kg over this period. There was no history of fever, abdominal pain, hematemesis or melena. He had no previous blood transfusion, surgery or significant medical illness. He used to smoke and had been a social drinker but had discontinued both for nearly 5 years. General examination revealed deep icterus and shiny nails, but no pallor or nodes. Abdominal examination revealed a firm palpable gall bladder. There were no stigmata of chronic liver disease.

Investigations: hemoglobin 11.9 g/dL, platelets 289,000/μL, WBC count 10,200/μL (neutrophils 65%, lymphocytes 13%, monocytes 20%, eosinophils 2%, basophils 1%), ESR 76 mm in 1st hour, serum bilirubin 24.9 mg/dL (direct 14.7), AST 52 U/L, ALT 60 U/L, alkaline phosphatase 434 IU/L (normal 39-117), serum protein 6.7 g/dL (albumin 3.3). CT scan done at the referring hospital showed hepatomegaly, distended gall bladder, dilatation of intrahepatic biliary radicles, and dilated common bile duct with abrupt tapering at distal end suggestive of malignant stricture. In addition he had a smooth filling defect in the right posterolateral wall of the urinary bladder. Ultrasonography done on admission confirmed the above findings, and suggested a hilar filling defect that had not been seen in the CT scan. Peripheral blood smear examination revealed myeloid blast cells. Bone marrow aspiration and trephine biopsy revealed more than 75% peroxidase-positive myeloid blasts with large number of eosinophils, confirming a diagnosis of eosinophilic variant of M4 acute myeloid leukemia.

White blood cell counts began to rise progressively during the hospital stay. Therapeutic endoscopic retrograde cholangiography was attempted but was unsuccessful. Percutaneous cholangiogram (Fig) showed isolation of right anterior and posterior and left systems by a stricturing lesion and narrowing of common hepatic duct. Biliary drainage was achieved by an 8.5 Fr pigtail catheter placed at the confluence into the right anterior duct. However the patient’s general condition continued to deteriorate and he expired before definitive chemotherapy could be instituted.
The most common cause of jaundice in AML is drug-induced hepatocellular damage. A late event may be transfusion-related viral hepatitis. AML presenting as obstructive jaundice is rare. Chloromas or granulocytic sarcomas (GS) are masses of leukemic cells that may form deposits in different organs. A more appropriate nomenclature proposed is extramedullary myeloid tumors (EMMT). There are case reports of chloromas in various sites like the GI tract, ovary, uterus, breast, testis, cranial or spinal dura, orbit, lung, mediastinum, prostate, and other organs. However there are very few reports of chloromas in the common bile duct especially with jaundice as the presenting feature. Cholestatic hepatitis due to hepatic sinusoidal infiltration as the presenting feature of AML has also been reported. Chloromas 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, immunocytochemistry, electron microscopy, and molecular studies are all useful tools in making a definitive diagnosis. There appears to be some association with chromosomal abnormalities like t (8; 21) and inv 6. A proposed diagnostic panel includes chloroacetate-esterase, myeloperoxidase, lysozyme, and CD43, together with other B- and T-lineage markers, in particular CD79a and CD3, for confirming the diagnosis of chloromas.

When isolated granulocytic sarcomas occur without evidence of AML, radiotherapy or surgery may be indicated. There is no evidence that treatment of an isolated granulocytic sarcoma with systemic chemotherapy will prevent later occurrence of typical AML. The presence of chloromas in the setting of AML is usually a sign of disease acceleration.

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Ivemark syndrome in association with congenital septum transversum defect and pancreatic divisum
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A four-month-old female baby presented with cyanosis and respiratory distress. A provisional diagnosis of congenital posterolateral diaphragmatic hernia was made but on exploration there was a defect in the septum transversum along with features of Ivemark syndrome – asplenia with visceral heterotaxia, malrotation and pancreatic divisum – an association not yet reported in literature. The child did well after operative correction of the hernia. Echocardiography showed situs inversus with dextrocardia with double outlet right ventricle, atrial septal defect, ventricular septal defect, patent ductus arteriosus and pulmonary stenosis. [Indian J Gastroenterol 2006;25:94-96]

The characteristic association of asplenia with visceral heterotaxia is traditionally named after the Swedish pediatrician, Ivemark. He described the implications of splenic agenesis on the pathogenesis of heart malformations in childhood. The condition has been described by terms like cardioesplenic syndrome, syndrome of visceral symmetry or heterotaxy syndrome.

The most common congenital defect in diaphragm is through the pleuroperitoneal membrane (left > right).