Pulmonary inflammatory pseudotumor with jaundice and anemia: A case report

Sir,

Inflammatory pseudotumor (IPT) of the lung is an uncommon entity and only a few studies are available till date although Brunn first described it way back in 1939. In fact, it is still not clear whether IPT is an uncontrolled inflammatory process or a true neoplasm.[1] Here, we describe a case of pulmonary IPT, which had an unusual presentation contrary to many of the previously reported cases.

An 18-year nonsmoker male presented with low grade fever since 4 months, along with left-sided chest pain since 3 months and nonproductive cough since 2 months. There was no history of hemoptysis, breathlessness, wheeze, or weight loss. He had been treated outside for over the last two months with repeated courses of antibiotics and received three units of blood transfusion for anemia of uncertain etiology. However, he denied any history of bleeding from the natural orifices.

On examination, he had icterus and pallor without any clubbing, edema, or lymphadenopathy. There was diminished breath sound over the left infrascapular area with dull percussion note, without any mediastinal shift or adventitious sound. He also had mild hepatomegaly without any tenderness.

In his chest x-ray [posterior-anterior (PA) view], a nonhomogenous left perihilar mass was noted [Figure 1]. Complete hemogram revealed microcytic, hypochromic anemia with hemoglobin 7 gm/dl, mildly elevated total leukocyte count (TLC) (13000/mm^3), and raised erythrocyte sedimentation rate (ESR) (100 mm at first hour) with normal differential leukocyte count (DLC) and platelet count. Liver function test (LFT) showed conjugated hyperbilirubinemia (total bilirubin 3.4 mg/dL and direct bilirubin 1.9 mg/dL) with raised alkaline phosphatase (ALP) (960 IU/L) but normal transaminases. This derangement of LFT was similar to another test performed 2 months back. However, other routine biochemical tests were within the normal range.

He was evaluated for pyrexia of unknown origin and malaria dual antigen, widal test, urine routine examination and culture, blood culture, and human immunodeficiency virus (HIV) were all negative. Regarding jaundice, the viral markers were negative and ultrasonography of the abdomen did not reveal any gallstone or extraluminal biliary obstruction except mild hepatosplenomegaly. With regard to anemia, direct Coombs test was negative and bone marrow examination revealed only hypercellular marrow without any evidence of hematological malignancy. But serum iron profile could not be performed as he received multiple blood transfusions within the last 2 months. Sputum for acid-fast bacilli (AFB) was negative; Mantoux test showed an induration of only 4 mm; electrocardiogram (ECG) and two-dimensional (2D)-echocardiogram were normal apart from trivial mitral regurgitation.

Subsequent contrast-enhanced computed tomography (CECT) of the thorax disclosed a large well-defined lobulated mass in the lower lobe of the left lung with extensive internal calcification [Figure 2]. But no mediastinal lymphadenopathy or pleural effusion was noted. A computed tomography (CT)-guided trucut biopsy from the mass showed the presence of interlacing fascicles of spindle-shaped fibroblastic and myofibroblastic cells having eosinophilic cytoplasm, oval nuclei, and inconspicuous nucleoli without cytological atypia or mitosis; there was heavy infiltration with inflammatory cells, mainly lymphocytes, plasma cells, and histiocytes [Figure 3]. Therefore, the possibility of IPT was considered and a cardiothoracic surgical opinion was sought.

Prior to surgery, preoperative fiberoptic bronchoscopy was performed, which was within normal limits except for slightly narrow left lower lobe bronchus due to extraluminal compression. Later, the left lower lobectomy was performed and histopathological examination of the resected mass confirmed presence of IPT. Immunohistochemistry from the mass further strengthened the diagnosis with positivity for vimentin and CD68, along with CD34 negativity. The patient was discharged from the hospital with advice for regular follow-up. On subsequent visit after 1 month, he was symptomatically better; the jaundice had subsided, ALP had normalized, hemoglobin had improved to 10.7 g/dL, and there was no clinical or radiological sign of recurrence over next 3 months.

Although pulmonary IPT is rare, it is the commonest isolated primary lung tumor in children. Their peak prevalence is in the second decade of life. This tumor mostly involves the lung and orbit but it has been reported to occur in different parts of the body including the liver, spleen, colon, mesentery, thymus, and central nervous system (CNS). Etiology and pathogenesis of this tumor is still uncertain and the natural course of the disease

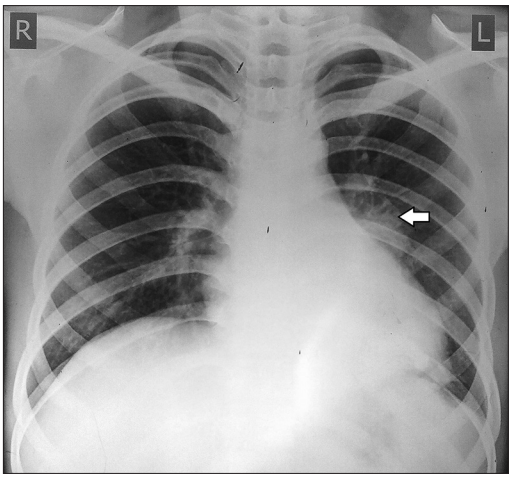

Figure 1: Chest x-ray (PA) showing a mass in the left hilar region (indicated by arrow)

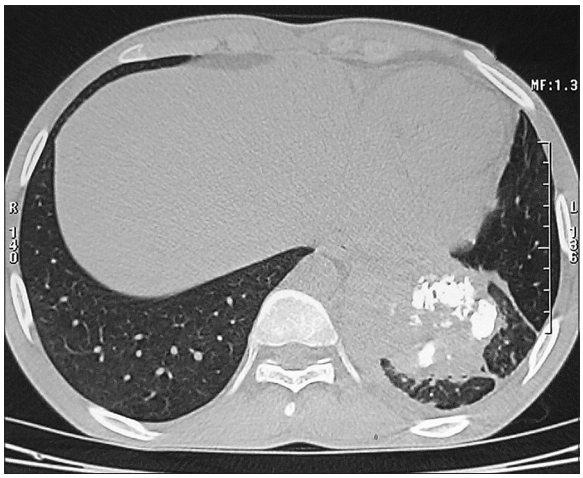

Figure 2: CT of the thorax (lung window) showing a left lower lobe lobulated mass with internal calcification

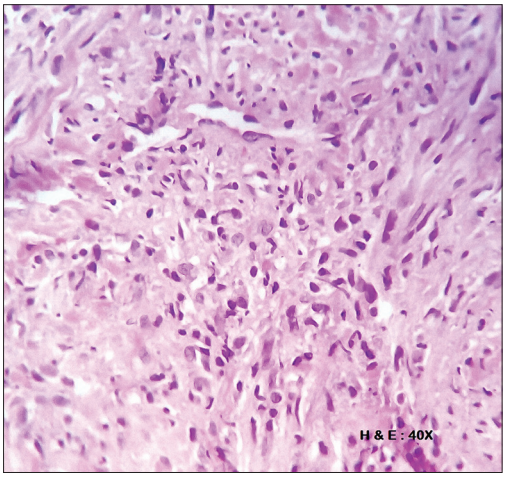

Figure 3: Histopathology showing prominent infiltration of lymphocytes and plasma cells among the spindle-shaped fibroblastic cells in the lesion (H and E stain, ×40 magnification)

may vary from spontaneous regression to sarcomatous transformation.

IPT is often asymptomatic and gets identified incidentally. Only 44% of the cases were symptomatic as per an Italian series with cough, chest pain, and fever being the common clinical presentations.[1] However, in another series from Mayo clinic, 78% of the cases had respiratory symptoms with cough being the commonest presentation.[2] A similar symptomatic presentation was found in our case too.

But in contrast to other cases of pulmonary IPT, our patient had anemia, disproportionately raised ESR, and jaundice in association. Very few case reports have mentioned anemia in cases of IPT. In a Korean review of 28 cases of pulmonary IPT, hematological picture was described in 15 cases, out of which only three patients had anemia (i.e., 20% cases) and 53.3% cases had normal blood picture.[3] Only one case in that series had a raised ESR. Another case report from Turkey described microcytic hypochromic anemia with pulmonary IPT in an 8-year-old boy and surgical resection of the lung mass improved that anemia, which complemented the present case.[4]

Almost all of the IPT cases with hyperbilirubinemia had involvement of hepatobiliary tree or pancreas and caused obstructive jaundice by mechanical compression. But in our case, no mechanical obstruction was detected and the reason behind jaundice and elevated ALP was unclear. However, one old case report of IPT of the thymus (1986) described elevated ALP (525 mU/mL) without any identifiable mechanical cause and that enzyme level returned to normalcy following tumor resection. Another case of maxillary sinus IPT in a 2-year-old girl also had raised ALP (541 IU/L) although no hepatobiliary obstruction was described in that report.[5]

Transthoracic fine-needle aspiration is often nondiagnostic and the surgically resected specimen is required for appropriate diagnosis. Immunohistochemistry is often needed to differentiate IPT from entities such as sarcoma, lymphoma, malignant fibrous histiocytoma, and malignant plasmacytoma.[1] IPT is universally negative for CD34 and universally positive for vimentin and it is usually positive for smooth muscle actin, CD68, and epithelial membrane antigen, which corroborated with our data.[1]

Complete resection with pulmonary reservation remains the treatment of choice. Radiation and corticosteroid therapy has been successfully tried in a few cases. Prognosis is excellent after resection with minimal chance of recurrence.

In conclusion, although a rare entity, possibility of IPT should be kept in mind while dealing with tumor-like lesion in the lung in young patients. Several systemic features of this disease subside spontaneously following tumor removal and they do not require specific management. Although complete resection is the key to prevent recurrence, the patient should undergo a prolonged and strict follow-up to detect relapse at the earliest, if any.

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Conflicts of interest
There are no conflicts of interest.

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