Inflammatory pseudotumor of the lung with complete resolution

Sir,

Inflammatory pseudotumor (IPT) is a mass-forming lesion that commonly involves lung, orbit, and liver.[1] A simple classification has been reported which divided the spectrum of IPT into nonneoplastic versus neoplastic variants, the latter including inflammatory myofibroblastic tumor (IMT).[2] Clinical diagnosis is challenging because of diagnostic dilemma and may encompass malignant lesions, such as lung carcinoma and metastatic tumor.[3] Literature available on diagnosis and treatment varied from minimally invasive biopsies to invasive thoracotomy specimens, and the treatment options varied from the conservative approach to treatment with anti-inflammatory drugs, antibiotics, and/or steroids to the surgical resection of the mass.[4]

A 35-year-old nonsmoker male presented with low-grade fever and a recent history of hemoptysis for 5–6 days. There was no other significant history; acute-phase reactant IgA was increased, with a value of 2620 mg/dl (90–450 mg/dl). The rest of the hematological and biochemical parameters were within normal limits. Examination of bronchoalveolar lavage fluid did not show any evidence of infection, including tuberculosis and fungal infections. Computed tomography (CT) chest showed a heterogeneously enhancing mass lesion measuring 3.6 cm × 2.5 cm with irregular margins seen in the posterior-basal segment of the right lower lobe. Vessels were seen traversing through the mass lesion, and the lesion was abutting the right paravertebral soft tissue at the D11 level along with few tiny nodules [Figure 1]. The CT-guided biopsy from the lung mass lesion showed a tumor composed of spindle cells to stellate cells arranged in short fascicles and whorls in a collagenous background. These cells displayed mildly anisomorphic oblong-to-spindle-shaped nuclei, with bland chromatin, inconspicuous nucleoli, and moderate cytoplasm. These cells were accompanied by lymphoid aggregates, and dense inflammatory cell infiltrates chiefly composed of plasma cells, lymphocytes, few eosinophils, and neutrophils. No mitosis or necrosis was seen. Obliterative phlebitis was also observed. On immunohistochemistry tumors, cells are positive for vimentin, smooth muscle actin, desmin (focal) and are negative for anaplastic lymphoma kinase. Increased IgG4-positive plasma cells were seen (>20/hpf) [Figure 2]. After 3 months of follow-up, the patient remained asymptomatic, recovered well, and showed no evidence of recurrence.

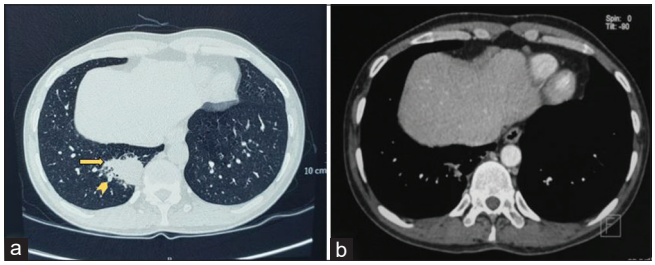

Figure 1: (a) Pretreatment computed tomography of the chest revealing a heterogeneously enhancing mass (arrow) with multiple calcific nodules surrounding the lesion (arrow-head). (b) Posttreatment follow-up computed tomography chest after 1 month

IMT of the lung has conjointly allotted terminology as IPT, plasma cell granuloma, histiocytoma, and fibroxanthoma are rare, and the incidence reported is to be 0.04% of all tumors of the lung.[5] It can happen in all age groups, with a slight predominance in those younger than 40 years; however, there is no sex predilection.[1]

Majority of cases are asymptomatic or present with nonspecific symptoms, such as cough, hemoptysis, shortness of breath, chest pain, and fever, and are thus discovered incidentally on imaging.[6] A review article by Faraj *et al.* has elaborated on the pathogenesis of IPT

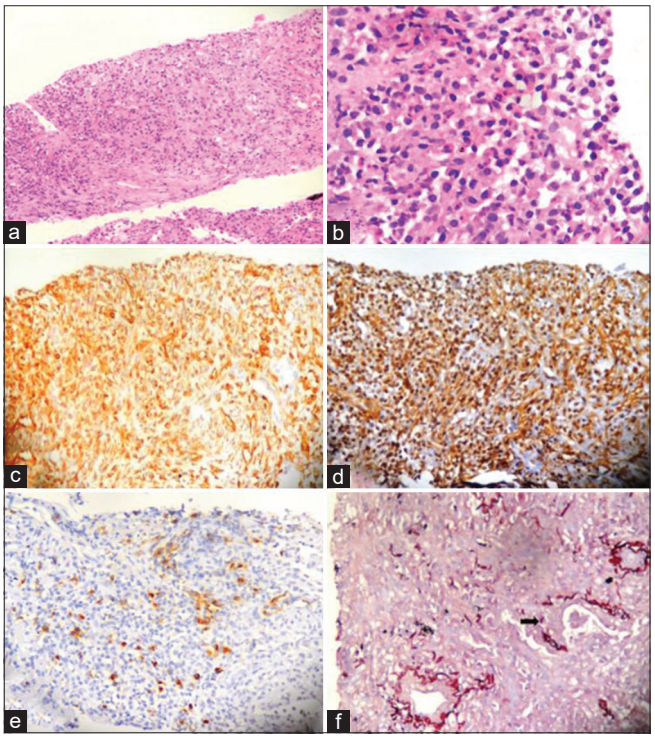

Figure 2: Lung biopsy showing fibrosis and a mixed inflammatory infiltrate with a predominance of lymphocytes and plasma cells (a and b, H and E, ×20 and × 40). Immunohistochemistry reveals positive immunoreactivity for vimentin (c, ×20), smooth muscle actin (d, ×20), increased IgG4-positive plasma cells (e, ×20). Obliterative phlebitis is also noted (f, Orcein stain, ×40)

as an exaggerated inflammatory process and concluded the possible etiologies being infectious agents, autoimmune reactions, and systemic inflammatory response syndrome.[7] Radiological findings in most of the patients embrace well-circumscribed solitary peripheral lung nodules with a predilection for the lower lobe. The remainder of the lesions may present as multiple pulmonary nodules and endobronchial lesions. Calcifications and lymphadenopathy are seen in 15% and 7% of cases, respectively.[8] Recently, IPT has been classified pathologically into fibrohistiocytic and lymphoplasmacytic subtypes. Fibrohistiocytic IPT is portrayed by xanthogranulomatous inflammation, multinucleated giant cells, and neutrophilic infiltration. However, lymphoplasmacytic IPT showed diffuse lymphoplasmacytic infiltration, prominent eosinophilic infiltration, and more commonly obliterative phlebitis as compared to fibrohistiocytic type.[9]

Immunohistochemical investigations could also be beneficial in distinguishing IMT from tumors with similar histopathologies, such as spindle cell carcinomas, lymphomas, and inflammatory sarcomas. IMT profiles are typically immunoreactive to vimentin (99%), SMA (92%), desmin (69%) and are negative for S100 protein, cytokeratin, and CD68 (25%).[10] A significant amount of IgG4-positive plasma cells (>20/hpf in core biopsy or >50/hpf in the surgically resected specimen) are seen in lymphoplasmacytic type.[11]

Surgical resection is the treatment of choice not only to exclude malignancy but also to attain cure. Early and complete resection of the IMT remains the best treatment option.[12] By making an accurate diagnosis, unnecessary thoracotomy can be avoided and reserved for complicated cases. In such cases, antibiotic/or corticosteroid treatment could also be curative, and surgical resection is avoided. The patient can be reassured about a favorable outcome and good prognosis.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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