A case of disseminated bronchus-associated lymphoid tissue lymphoma masquerading as nonresolving pneumonia

Sir,

Bronchial mucosa-associated lymphoid tissue (MALT) lymphoma or bronchus-associated lymphoid tissue (BALT)-lymphoma (BALTOMA) is an uncommon, low-grade B-cell non-Hodgkin's lymphoma (NHL) of the bronchial MALT. The diagnosis is often missed since it has similar clinical and radiological presentation as that of a community-acquired pneumonia. A 60-year-old nonsmoker female was admitted with 6-month history of chronic nonproductive cough, gradually progressive breathlessness, and weight loss. She had a history of three hospitalizations prior to presentation for the above complaints, where initially she was diagnosed as a case of community-acquired pneumonia and discharged on antibiotics. When her symptoms worsened, she was re-evaluated and was started on fluconazole and ganciclovir, followed by antituberculosis therapy. On presentation to our center, she had a room air saturation of 88%. General physical examination revealed pallor, bilateral cervical lymphadenopathy, with firm nontender, nonmatted, mobile lymph nodes. Abdominal examination was unremarkable. On chest auscultation, there were bilateral crackles and bronchial breath sounds over the right base.

Laboratory studies revealed hemoglobin of 9.4 g/dl, white blood cells of 12,700/mm³ (polymorphs 60%, lymphocytes 38%, and monocytes 2%), and platelet count of 4.2 lakh/cumm. Peripheral smear was unremarkable. Liver and kidney function tests were within normal range.

Computed tomography (CT) scan of chest showed bilateral airspace consolidation with air bronchogram (right > left) and multiple randomly distributed nodules in both the lung fields [Figure 1] along with mediastinal lymphadenopathy, involving multiple lymph node stations. An ultrasound-guided fine needle aspiration (FNA) of the right upper cervical lymph node revealed only reactive lymphadenitis.

To establish the diagnosis and to ascertain the best site for tissue diagnosis, a positron emission tomography CT scan was done, which revealed a hyper metabolic irregular speculated mass lesion in the right lung in the upper middle and lower lobes with multiple metabolically active pleural and parenchymal nodules along with generalized lymphadenopathy with avid fluorodeoxyglucose uptake (cervical, axillary, mediastinal, abdominal, and pelvic).

The patient underwent fiber-optic bronchoscopy (FOB), endobronchial ultrasound-guided (EBUS) FNA of the mediastinal lymph nodes, and endobronchial biopsy (EBB). FOB revealed a bosselated, edematous-looking mucosa over the trachea–bronchial tree, especially over the right main bronchus, leading to narrowing of the right main and the upper lobe bronchus [Figure 2]. EBUS cell block showed aggregates of small lymphoid cells with scant cytoplasm raising the suspicion of a low-grade lymphoma. EBB demonstrated bronchial mucosa lymphoepithelial lesion – infiltration of the bronchial mucosa by small round lymphoid cells with scant cytoplasm classically seen in MALT lymphoma [Figure 3].

Immunohistochemical (IHC) of the EBB staining clinched the diagnosis of low-grade marginal zone lymphoma (MZL). (CD19, CD20, and CD79a were diffusely positive. CD3 was focally positive whereas CD5, CD10, and CD23 were negative. Ki-67 was 50%). Histopathology and IHC were consistent with extranodal low-grade MZL of MALT-BALTOMA. Bone marrow aspiration did not reveal any dysplasia or malignancy of any of the cell lines.

The patient was started on chemotherapy with chlorambucil and steroids. She responded dramatically in 3 weeks with significant clinical and radiological improvement. She has now completed 5 cycles of the same chemotherapy.
Primary pulmonary lymphoma (PPL) accounts for only 0.5–1% of all primary pulmonary malignancies and <1% of all NHL.[1] The most common histological variety of PPL is the MALT type arising from the BALTO. It is a low-grade B-cell extranodal lymphoma characterized by a proliferation of clonal marginal zone lymphocytes that invade epithelial structures and form characteristic lymphoepithelial lesions.[2-4]

Usually described in middle-aged patients, most of these tumors are clinically silent. Patients present with nonspecific complaints such as fever, anorexia, and weight loss. Radiology usually reveals unilateral airspace consolidation. However, bilateral diffuse reticulonodular opacities, atelectasis, pleural effusions, hilar, and mediastinal adenopathy, etc., have all been described.[5]

A combination of appropriate histopathology and IHC is necessary to ascertain the diagnosis. IHC demonstrates a B-cell phenotype (CD19, CD20, and CD79a positive) whereas CD5 and CD10 are negative as was in our case, thus ruling out follicular lymphoma, mantle cell lymphoma, and chronic lymphocytic leukemia.[5,6] As with other lymphomas, chemotherapy is the cornerstone of therapy for disseminated disease. Chlorambucil-based chemotherapy and conventional regimes such as cyclophosphamide, doxorubicin, vincristine, and prednisone with rituximab have been tried in these patients. The prognosis is usually favorable with prolonged progression-free survival despite disseminated disease.[5,6] Nongastric maltomas such as PPL are known to disseminate widely, as was the case in our patient.[6]

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Conflicts of interest
There are no conflicts of interest.

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