Original article

Pulmonary MALT lymphoma: imaging findings in 24 cases

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Received: 4 October 1999; Revised: 24 February 2000; Accepted: 18 April 2000

Abstract. The aim of this study was to describe the imaging features of pulmonary mucosa-associated lymphoid tissue (MALT) lymphoma. The chest radiographs (n = 18) and CT scans (n = 17) of 24 patients (18 men and 6 women) aged 27–78 years (mean = 56 years), with a known diagnosis of pulmonary MALT lymphoma, were retrospectively reviewed by two radiologists and the imaging findings are described. Six of the 24 patients had a history of an autoimmune disorder and 1 patient had acquired immune deficiency syndrome. Multiple pulmonary lesions were identified in 19 of 24 patients (79%) and solitary lesions in 4 of 24 patients (17%). Diffuse pulmonary infiltration was present in 1 patient. Lesions included masses or mass-like areas of consolidation (n = 21) and pulmonary nodules (n = 18). Associated findings were air bronchograms, airway dilatation, a positive angiogram sign and a halo of ground-glass shadowing at lesion margins. Peribronchovascular thickening was also observed, as were hilar or mediastinal lymph node enlargement and pleural effusions or thickening. Although rare, the diagnosis of pulmonary MALT lymphoma should be considered in patients with the imaging features described, particularly when in association with an indolent clinical course or a history of autoimmune disease.

Key words: Lung neoplasms – Lymphoma

Introduction

Pulmonary B-cell non-Hodgkin’s lymphomas, although rare, are now well recognised as primary lung neoplasms. They have now previously been classified according to nodal classification systems, [1, 2, 3, 4] but are currently thought to be primary extranodal lymphomas, mainly of low grade, which arise from bronchial mucosa-associated lymphoid tissue (MALT) [5, 6, 7, 8]. These are analogous to extranodal MALT lymphomas, which were first described in the stomach [9] and are now known to arise at numerous other mucosal sites [10, 11, 12, 13]. MALT lymphomas are now classified in the revised European American system as extranodal, marginal zone, B-cell lymphomas [14]. Most lung lesions that were previously termed “pseudolymphomas” [15] are now also thought to represent pulmonary MALT lymphomas. The radiographic features of pulmonary involvement by other types of lymphoma have been previously described [16, 17, 18, 19], although the radiological appearances of pulmonary MALT lymphomas are less well documented [20, 21, 22, 23, 24, 25] with no large series reporting the findings on CT.

The purpose of this article is to describe the radiological appearances of pulmonary MALT lymphomas in a series of 24 histologically diagnosed cases.

Materials and methods

Patients

Review of pathological records at the Royal Brompton Hospital from 1985 to 1998 identified 40 patients with pulmonary MALT lymphoma. Thoracic imaging was available in 25 of these patients, one of whom had a history of a previous orbital high-grade non-Hodgkin’s lymphoma and was excluded from this study. The remaining 24 patients presented with pulmonary lesions and comprised 18 men and 6 women, aged 27–78 years (mean age 56 years). The case notes and radiological investigations of these 24 patients were retrospectively reviewed. Case notes were examined for a history of autoimmune disorder, co-existing chest disease, previous lymphoma, evidence of extra-thoracic lymphoma, acquired immune deficiency syndrome (AIDS) and the method employed in obtaining a diagnostic specimen of lung tissue for each patient.

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Seventeen of the 24 patients had a history of respiratory symptoms such as cough or dyspnoea at presentation, 2 patients complained of chest pain and 6 patients were asymptomatic. A history of autoimmune disorder was recorded in 6 of the 24 (25%) patients: rheumatoid (n = 4); Sjogren’s syndrome (n = 1); mixed connective tissue disorder (n = 1); and 1 patient had cryptogenic fibrosing alveolitis without an associated connective tissue disorder. One patient was HIV positive with a history of recurrent pulmonary infections including a single episode of Pneumocystis carinii pneumonia, and there was a history of treated nodal Hodgkin’s disease in 1 patient who presented with an asymptomatic pulmonary mass 10 years after mantle radiotherapy. Three patients were known current cigarette smokers, 8 were non-smokers, 8 were previous smokers who had stopped between 10–38 years previously (mean = 21 years) and a smoking history was unavailable in the remaining 5 patients.

The mean duration from the onset of symptoms or identification of an asymptomatic lung lesion, to the histological diagnosis being made, was 14 months (range = 2 weeks to 14 years). Delays in diagnosis of greater than 3 months occurred in 11 of the 24 (46%) patients due to a variety of factors. These included: non-diagnostic biopsy specimens; patients declining biopsy or surgery; patients being medically unfit for thoracic intervention; and the slow, insidious progression of several lesions such that the possibility of malignant disease was not considered.

At diagnosis, none of the 24 patients had evidence of nodal or extranodal lymphoma outside the thorax, on clinical examination (n = 24), abdominal CT (n = 18), abdominal ultrasound (n = 3) or at subsequent post-mortem examination (n = 3). Bone marrow involvement was demonstrated in 1 patient, but no other extrathoracic disease was identified clinically, radiologically or at subsequent post-mortem examination.

Pathological analysis
Pathological analysis was performed independently by two pathologists (A.G.N. and A.C.W.) on specimens of lung tissue obtained at biopsy, surgery or post-mortem examination. Specimens were obtained for histological analysis as follows: bronchoscopy biopsy (n = 3); percutaneous cutting needle biopsy (n = 2); thoracoscopic or open-lung biopsy (n = 7); surgical resection (n = 9); and post-mortem examination (n = 3). Prior to definitive biopsy or resection, numerous non-diagnostic biopsies had been obtained: percutaneous (n = 7); transbronchial (n = 7); thoracoscopic or open-lung biopsy (n = 2); and mediastinoscopy (n = 2).

All 24 cases were considered to be pulmonary B-cell non-Hodgkin’s lymphomas of MALT origin by both of the reviewing pathologists. The diagnosis was based on a combination of typical pathological appearances including characteristic cell cytology, pattern of infiltration, demonstration of lymphoepithelial lesions and a B-cell immunophenotype. Evidence of clonality was assessed using immunohistochemistry to look for light chain restriction and polymerase chain reaction (PCR) analysis to look for rearrangements in the immunoglobulin heavy chain gene.

In the absence of an established method of grading pulmonary B-cell lymphomas the principles of the updated Kiel classification were used to subdivide cases into low- and high-grade lesions, mirroring the approach used to classify intestinal lymphomas [26]. Tumours were considered to be low-grade lymphomas of MALT origin when they were composed of a monomorphic proliferation of small malignant centrocyte-like, lymphocyte-like or monocytoïd cells with colonisation of germinal centres and the development of lymphoepithelial lesions. Tumours were considered to be high grade when comprised predominantly of blast cells, with the appearances being more diffusely infiltrative and destructive with evidence of necrosis.

Thirteen of the 24 cases were classified as low grade, 10 cases as high grade and one case as high grade with focal areas of low-grade tumour, which for the purpose of analysis was considered high grade. Four of the ten high-grade tumours were partly necrotic on histological examination. Lymph node sampling had been performed at surgery in 15 patients, of whom 3 of 15 (20%) were node positive.

A small synchronous T1 N0 adenocarcinoma was identified in 1 patient. Three patients with high-grade lymphoma also had a histological diagnosis of pulmonary fibrosis, one in association with rheumatoid arthritis and one in association with a mixed connective tissue disorder. Mycobacterium tuberculosis was isolated from the lung resection specimen of 1 patient who had a history of previously treated pulmonary tuberculosis.

CT technique
The CT examinations were performed on one of three machines: Imatron 150L electron beam scanner (Imatron, San Francisco, Calif.; n = 13); Elscint 2002 (Elscint, Haifa, Israel; n = 3); or Siemens Somaton (Siemens Medical Systems, Erlangen, Germany; n = 1). The CT scans were obtained with 1.5-mm sections at 10-mm intervals (n = 2), 3-mm sections at 10-mm intervals (n = 3), contiguous 6-mm sections (n = 9), contiguous 8-mm sections (n = 1) and contiguous 10 mm sections (n = 3). Both 3- and 10-mm-collimation sections were available for 1 patient and intravenous contrast medium was administered in 9 cases. The 1.5- and 3-mm sections were reconstructed using a high-spatial-frequency algorithm.

Image analysis
An independent, retrospective review of the available chest imaging for each patient was performed by two radiologists (L.J.K. and S.P.G.P.). Where there was disagreement on the radiological findings a consensus opinion was obtained. Plain chest radiographs were