PULMONARY PUZZLE

A rapidly growing lung mass with air crescent formation

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QUESTION

What is the diagnosis?

ANSWER

Air crescent formation within a mass-like lesion is most commonly associated with aspergillosis, usually in immunocompromised patients. However, this sign is occasionally seen in malignancy, vasculitis and lymphoproliferative disorders. The rapidity of progression was inconsistent with primary lung cancer and favoured an infective, lymphoproliferative or vasculitic process. The histological findings on biopsy led to a diagnosis of lymphomatoid granulomatosis (LG) grade 3.

LG is a rare, angiocentric and angiodestructive lymphoproliferative disease characterised by Epstein Barr virus (EBV)-infected B cells admixed with reactive T cells, often in association with an immunodeficient state. Key histological features are a mixed mononuclear cell infiltrate infiltrating vessels, varying numbers of CD20 B-lymphocytes positive for Epstein Barr virus and areas of dense lymphoid infiltration within the alveolar interstitium, and abundant fibrinous debris within the alveolar spaces. Within the infiltrate, an atypical CD20 lymphoid population was identified, lying in a denser CD3 T cell population.

Figure 1 Initial CT through the upper lobes showing an amorphous relatively low attenuation (near-fluid density) mass in the posterior segment of the right upper lobe. Sections through the lower lobes showed features consistent with non-specific interstitial pneumonia.
for EBV, present within a background of numerous CD3 T lymphocytes.4

LG is graded as 1–3 based on the proportion of large B cells, with morphological overlap in grades 2 and 3 with large B-cell lymphoma.5 Treatment for higher grades is with combination chemotherapy. Our patient received R-CHOP (rituximab, cyclophosphamide, doxorubicin, vincristine, prednisone) with good clinical response.

Key points illustrated by this case: (1) long-term immunosuppression should raise clinical suspicions for LG; (2) rapidity of disease progression can refine the differential diagnosis; (3) the air crescent sign is not specific to aspergillosis; (4) there can be sampling issues when needle biopsy is used, particularly for partially necrotic lesions.

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REFERENCES


Figure 2 High resolution CT two months later showing a marked increase in size of the mass-like area of consolidation and the development of an ‘air crescent sign’. In addition, there were some new small foci of peripheral consolidation.

Figure 3 (A) The biopsy shows necrosis and predominantly small lymphocytes and histiocytes. However, occasional atypical cells are also present (arrow), which (B) stain with CD20 ((A) H&E×400, (B) CD20×200). This figure is only reproduced in colour in the online version.