X-Ray Quiz

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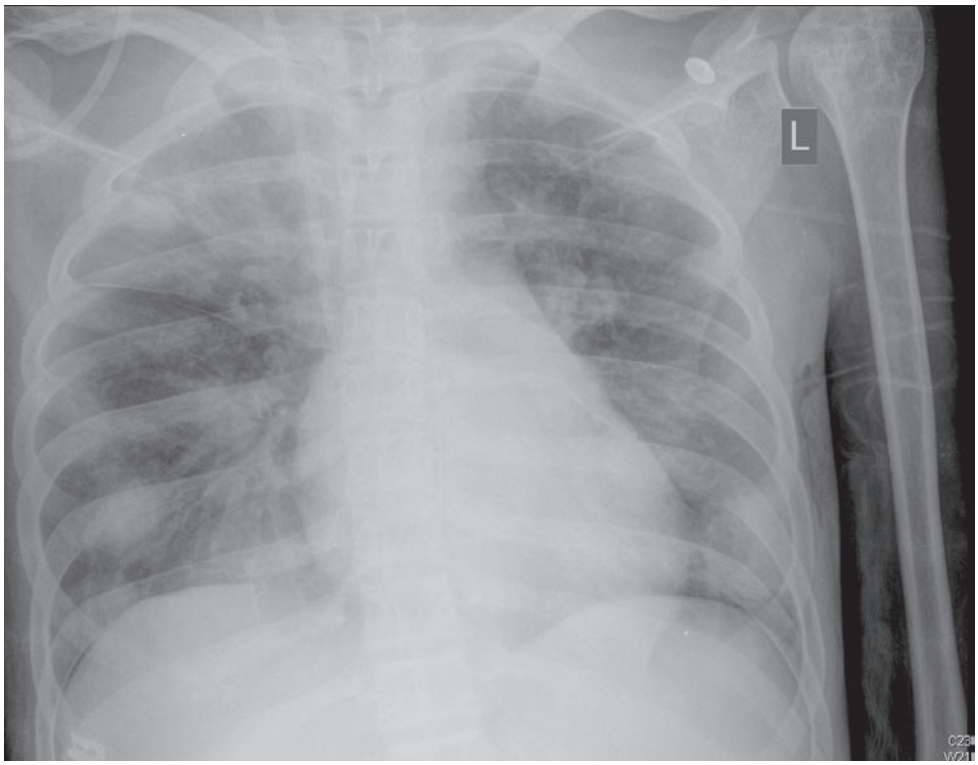
An 18-year-old boy undergoing chemotherapy for acute lymphoblastic leukaemia was admitted to the PICU for respiratory distress, septic shock with pancytopenia. He also had a blackish necrotic skin lesion over the scrotum. He was put on multiple potent antimicrobials including broad spectrum antibiotics, antiviral and antifungal (voriconazole) agents. However, he ran a rapid downhill course and this CXR was done at the time of deterioration.



Questions

What is the diagnosis?

A. Pyogenic abscesses
B. Pulmonary Aspergillosis
C. Pulmonary Mucormycosis
D. Pulmonary tuberculosis
E. Lung metastasis

(Answer on page 24)

Answers to X-ray Quiz on page 22

The correct answer is C (Pulmonary Mucormycosis)

This CXR reveals multiple round opacities over both lung fields. When this appears in immunocompromised patients, we should always consider infectious pneumonia especially caused by fungi. As our patient did not respond to voriconazole, pulmonary Mucormycosis is the most probable diagnosis rather than Aspergillosis. Pulmonary tuberculosis should always be kept at the back of our mind albeit associated skin lesion makes it less likely. Pyogenic lung abscess is unlikely as the CXR shows absence of typical air-fluid level. Lung metastasis from solid tumors is low on our list of differential diagnoses in the context of acute lymphoblastic leukaemia.

Unfortunately, a few days later, our patient developed profuse pulmonary haemorrhage and succumbed. Bronchoalveolar lavage (BAL) and scrotal wound swab both identified Absidia species, but the results were only available after the patient's death.

Absidia is a genus of fungi in the family Mucoraceae, which is commonly found in decaying organic substrates like bread, fruits and vegetables. Mucormycosis in healthy human is extremely rare but is frequently life threatening in immunocompromised hosts. Besides pulmonary manifestations, Mucormycosis can also cause cutaneous, sinus, cerebral, gastrointestinal and disseminated infections.

Signs and symptoms of pulmonary Mucormycosis are often non-specific. Patients usually present with persistent fever despite broad spectrum antimicrobials, non-productive cough, progressive dyspnea and pleuritic chest pain. Further, pulmonary haemorrhage is also reported in keeping with its angio-invasive characteristic. X-ray findings range from focal consolidation with non-specific infiltrates, cavitary lesions, diffuse opacities to large wedge-shaped infarcts. Upper lobar disease and bilateral lung involvement are common.

Our patient was too ill to undergo CT thorax, which otherwise might give us a hint to the causative agent of nodular pneumonia. The characteristics of CT features of pulmonary Mucormycosis are presence of multiple nodules (>=10) as well as the reverse halo sign (a focal round area of ground-glass attenuation surrounded by a ring consolidation), which differs from the typical halo sign in Aspergillosis. The other differentiated diagnoses such as pyogenic lung abscesses and pulmonary tuberculosis would display their characteristic CT features, typically air-fluid level and associated lymphadenopathy respectively. The definitive diagnosis can only be established by the microscopy, culture or histology of specimens obtained from BAL or direct aspiration. Nevertheless, concomitant polymicrobial infection is common in immunocompromised hosts, making early diagnosis of pulmonary Mucormycosis very challenging.

Mucormycosis carries a high mortality rate of 50-85%, patient usually die from massive haemoptysis or disseminated disease. Its prognosis is further impacted by the usually delayed diagnosis. Early microbiological diagnosis by either BAL or aspiration/biopsy, and pre-emptive treatment are crucial in improving the outcome. First line antifungal agent such as voriconazole is not effective. High dose amphotericin B and posaconazole are the drugs of choice.

Further references

1. Kontoyiannis DP, Lewis RE. Agents of mucormycosis and Entomophthoramycosis. In: Mandell GL, Bennett GE, Dolin R, eds. Mandell, Douglas and Bennett's Principles and Practice of Infectious Diseases. 7th ed. Philadelphia, PA: Churchill Livingstone; 2010:3257-69.
2. Kendig E, Wilmott R. Kendig and Chernick's Disorders of the Respiratory Tract in Children [e-book]. Philadelphia, PA: Elsevier/Saunders, 2012.