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PULMONARY ALVELOLAR PROTEINOSIS : A case report

Radiodiagnosis

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ABSTRACT

Pulmonary alveolar proteinosis(PAP) is a rare diffuse interstitial lung disease with a prevalence of 1 per 10,00,000. It was first described in 1958 by Rosen et al. It is characterized by abnormal accumulation of pulmonary surfactant in the alveolar space, which in turn impairs gaseous exchange leading to a severe hypoxemia . Pulmonary surfactant is an insoluble proteinaceous material that is rich in lipids and stains positive with periodic acid–Schiff (PAS).

It is caused due to deficiency in granulocyte macrophage–colony stimulating factor (GM-CSF), as a result of the anti-GM-CSF antibody production, which ultimately leads to the accumulation of surfactant in the alveolar space.

It is usually present in young and middle-aged adults (20-50 years of age) . It is associated with smoking and has a male predilection (M:F of ~3:1)

Its clinical course is variable, ranging from complete remission to fatal respiratory failure.

On imaging, it is classically associated with" Crazy paving pattern"

on HRCT, although it is a rare cause of this non-specific finding.

Whole lung lavage was the gold standard therapy in PAP until the advent of GM-CSF.

We here report a case of 52 year old Indian male who presented to OPD with a history of long-standing cough with whitish sputum cough , progressive shortness of breath on exertion and multiple joint pain since 6 month

KEYWORDS

Pulmonary alveolar proteinosis,Crazy paving pattern Interstitial lung disease Whole lung lavage

CASE HISTORY

A 52 year old Indian man presented to OPD with a history of long-standing cough with whitish sputum, shortness of breath on exertion and multiple joint pain since 6 months. He had past medical history of Koch's. He smokes 20-30 cigarettes per week. Physical examination revealed a mildly dyspnoeic with normal vitals ; bibasilar fine inspiratory rales were present. Routine blood tests were normal.

Chest radiograph revealed bilateral alveolar infiltrates in butterfly distribution, sparing the costophrenic angles. Patient was then advised HRCT (plain + contrast)

HRCT showed extensive areas of ground glass opacities with interspersed interlobular septal thickening involving bilateral lung parenchyma predominantly in lower lobes with sparing of thin peripheral subpleural rim of parenchyma.

Multiple calcified nodules were noted in bilateral lower lobes and right middle lobe.

Extensive pleural calcifications was noted along the diaphragmatic and pericardial surface and also along the fissures.

Fibro calcific changes with adjacent pleural thickening with calcification was noted involving apical segment of right upper lobe and apicoposterior segment of left upper lobe – This findings were sequalae of old infective etiology(i.e) Kochs.

Owing to clinical profile and imaging findings a provisional diagnosis of Pulmonary alveolar proteinosis was made.

The patient was then advised bronchoscopy for confirmation of our diagnosis.

Fiberoptic bronchoscopy revealed alveoli filled with lipoproteina ceous material that stains positively with periodic acid—Schiff (PAS) staining.

A therapeutic whole lung lavage (WLL) was carried out, resulting is an immediate improvement in symptoms as well as in gas exchange, thus confirmed our diagnosis.

DISCUSSION:

Pulmonary alveolar proteinosis is a rare interstitial lung disease characterized by dense accumulation of phosphor-lipoproteinaceous material within the alveoli, however lung interstitium is preserved. It was first described in 1958 by Rosen et al and is believed to involve defective surfactant homeostasis. The clinical presentation is nonspecific .The most common symptoms are cough and dyspnoea (usually on exertion) and sometimes low grade fever.

Physical examination may reveal cyanosis, clubbing and bilateral fine rales .

Diagnosis was traditionally found on open lung biopsy. But now a days ,Whole lung lavage(WLL) is accepted as the most effective form The three main forms of PAP are: Autoimmune or primary, secondary, and congenital.It is caused due to deficiency in granulocyte macrophage–colony stimulating factor (GM-CSF), as a result of the anti-GM-CSF antibody production, which ultimately leads to the accumulation of surfactant in the alveolar spaces

Chest radiographs may vary in appearance. It can show bilateral perihilar nodular infiltrates, with areas of consolidation. Sometimes they coalesce predominantly on the lung bases, poorly defined mosaic-like and ground glass with "butterfly wings" pattern, mimicking atypical pneumonia or massive pulmonary edema. Interstitial disease can also be seen, in longstanding cases.

HRCT reveals diffuse ground glass opacities with septal, intra and interlobar thickening; a pattern known as "crazy paving", which is characteristic but not pathognomonic, as it may be seen in various other conditions too.

Diagnosis is confirmed by transbronchial fiberoptic bronchoscopy guided biopsy,.. Moreover, when a bronchoalveolar lavage is

performed, milk-like foamy opaque fluid is pathognomonic The most accepted treatment is whole lung lavage, with goal of extracting the proteinaceous material occupying the airway. There are various other alternative treatments such as subcutaneous or inhaled granulocyte-macrophage colony-stimulating factor (GM-CSF), plasmapheresis and rituximab . Usually these treatments are administered when there is lack of response to whole lung lavage or the patient does not tolerate the conventional treatment.

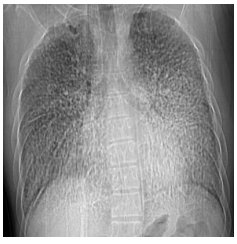
DIFFERENTIAL DIGNOSIS-

The radiologic differential diagnosis of crazy-paving includes:

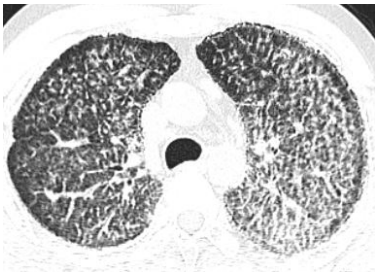
- pulmonary edema,
- pneumonia,
- alveolar hemorrhage,
- diffuse alveolar damage,
- lymphangitic carcinomatosis

Others-

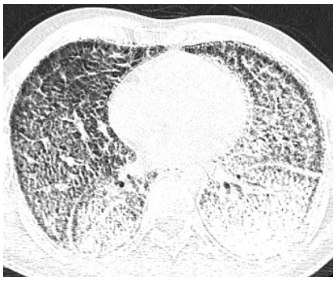
- Pneumocystis jirovecii and Mycoplasma sp pulmonary infections,
- Lipoid pneumonia,
- Drug-related hypersensitivity reaction



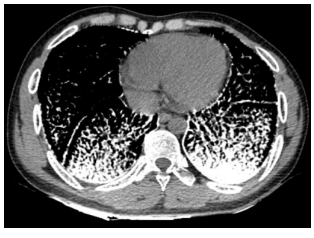
Chest radiograph PA view showing bilateral alveolar infiltrates in butterfly distribution, with sparing the costophrenic angles.



HRCT thorax (Plain) lung window, bilateral upper lobes showing extensive areas of ground glass opacities with interspersed interlobular septal thickening involving bilateral lung parenchyma with sparing of thin peripheral subpleural rim of parenchyma.



HRCT thorax (Plain) lung window, showing extensive areas of ground glass opacities with interspersed interlobular septal thickening involving bilateral lung parenchyma predominantly in lower lobes .



HRCT (plain) mediastinal window showing multiple calcified nodules in bilateral lower lobes and extensive pleural calcifications along the diaphragmatic and pericardial surface and also along the fissures.

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