Universe
Scientific Publishing

Application of 640-Volume CT in Idiopathic Pulmonary Hemosiderosis in Adults

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Abstract: **Objective**: To have a deep understanding about idiopathic pulmonary hemosiderosis (IPH). **Methods**: Clinical and imaging data of 6 patients of IPH in adults were collected and analyzed. **Results**: Imaging findings must be combined with clinical laboratory examination to make a diagnosis of IPH. Transbronchial lung biopsy (TBLB) is the gold standard for evaluation of suspected patients. **Conclusion:** IPH is extremely rare amid adults, and imaging results on single chest CT are nonspecific. The longitudinal follow-up is critical for diagnosis and posttreatment evaluation.

Keywords: Idiopathic Pulmonary Hemosiderosis; Computed Tomography; Imaging Findings

Introduction

IPH is a rare disorder characterized by recurrent episodes of unexplained intra-alveolar hemorrhage, followed by accumulation of hemosiderin in macrophages. In selected populations, the prevalence of IPH alone is estimated to be 0.24 to 1.23 per [1]. IPH is most prevalent in children under the age of 10, although it is uncommon in adults, with a mean survival rate of approximately 2.5 years after diagnosis [2]. The most common clinical manifestations are chronic pulmonary symptoms (cough, dyspnea, or hemoptysis), iron deficiency anemia, and pulmonary infiltration [3]. However, one of the typical triads may be the leading symptoms.

1. Objective and methods

1.1 Objective

Data of 6 patients of IPH with biopsy-proven from February 2014-April 2022 were collected， chest CT plain scan was performed for each patient, including four males and two females, aged 23 – 65 years , the median age was 46 years. 4 patients had associated symptoms of cough, sputum, and hemoptysis symptoms, etc.

1.2 640-slice volume CT scan

Use of the 640-slice volume CT, all Patients were taken in the supine position, Arms on the head, Scanning ranges from the thoracic entrance to the bottom of the lung, Flat scanning at the end of deep inhalation; scanning collimator width is 64mm 0.6mm; Tube voltage: 120kV, Current: 35mAs; Spitch 0.55, Matrix 512*512, The reconstruction layer is 1mm thick, margin 1mm, The original imaging are transmitted to the post-processing workstation for post-processing technologies.

1.3 Imaging Diagnosis

Images were reviewed by 3 experienced radiologists (all with more than 15 years of diagnostic experience and associate chief physician), respectively, including lesion localization, morphology, size, density, margin,presence of adjacent pleural, hilum, mediastinum. The three people discuss to the consensus.

2. Results

2.1 CT findings

In the hilar, perihilar, and lower lobe regions, non-enhanced axial and coronal chest CT images revealed significant central ground-glass opacities and consolidation. The lung apices and the costophrenic sulci were spared (Fig 1a, b). After three days of antibiotic treatment, the lesions had progressed significantly (Fig 2). Based on the patient's symptoms, signs, and laboratory tests, idiopathic pulmonary hemosiderosis (IPH) was diagnosed, and the patient was given hormone and immunosuppressive therapy. The lesions were visibly absorbed after one week of treatment and vanished after two weeks (Fig 3 and Fig 4a, b).

3. Discussion

IPH is a rare disorder characterized by recurrent episodes of unexplained intra-alveolar hemorrhage, followed by accumulation of hemosiderin in macrophages. In selected populations, the prevalence of IPH alone is estimated to be 0.24 to 1.23 per [1]. IPH is most prevalent in children under the age of 10, although it is uncommon in adults, with a mean survival rate of approximately 2.5 years after diagnosis [2]. The most common clinical manifestations are chronic pulmonary symptoms (cough, dyspnea, or hemoptysis), iron deficiency anemia, and pulmonary infiltration [3]. However, one of the typical triads may be the leading symptoms.

The definite cause of IPH is still unknown, although, several hypotheses have been proposed for its explanation, including autoimmune, environmental, allergic, and genetic factors [3]. According to one study, patients with IPH are often linked with particular autoimmune illnesses, such as celiac disease, dermatitis herpetiformis, glomerulonephritis, and rheumatoid arthritis [4]. Based on its pathology, IPH is classified into three phases: acute phase, chronic phase, and sequela phase. In the acute phase, a large number of red blood cells are manifested and exudates in alveoli, alveolar edema, and alveolar septum thickening. In the chronic phase, the major symptoms include a large amount of hemosiderin being deposited in the alveolar interstitium, alveolar interstitial fibrous tissue proliferation, interlobular septum, and alveolar wall thickening. The development of extensive interstitial fibrosis within the lung is the most common symptom of the sequela phase. The pathogenic alterations influence the imaging findings. In the acute phase, the chest CT scan image shows extensive ground-glass opacities, which are mostly symmetrically distributed in the pulmonary hilum, middle, and lower lung regions; with the lung apices and costophrenic sulci being less affected. Ground-glass opacities indicate diffuse intra-alveolar hemorrhage when coexistence with consolidation indicates that the alveoli are full of blood. The interlobular septum can thicken due to the deposition of hemosiderin-containing macrophages in the interstitium [5]. The "crazy-paving" sign appears due to interlobular septum thickening coexistence with ground-glass opacities [6]. Recurrent pulmonary hemorrhage patients may develop pulmonary fibrosis, which includes thickening of the interlobular septum, reticulation, and small cystic foci. HRCT is an important tool for detecting diffuse tiny reticulation, fine nodules, and interlobular septum thickening. The patient studied in this report was in the acute phase.

Because of the non-specific imaging results of IPH, it must be combined with a clinical laboratory examination to distinguish it from other diffuse pulmonary diseases. For instance, pulmonary alveolar proteinosis, anti-neutrophil cytoplasmic autoantibodies (ANCA) are associated with systemic vasculitis and phylactic pneumonia. Ground-glass opacity, crazy-paving sign, and map-like alteration are all symptoms of IPH, although they're more common in pulmonary alveolar proteinosis [7]. Furthermore, the lesions of pulmonary alveolar proteinosis often involve the subpleural areas. In ANCA-associated systemic vasculitis, the serum ANCA is mostly positive and multiple organs can be involved, with the kidneys being the most frequently affected organs. The typical CT scan of phylactic pneumonia shows diffuse central lobular nodules, patchy attenuation or ground-glass opacities, and mosaic signs dominated in the upper and middle lobes [8]. It can be distinguished from IPH if the patient with a history of allergen exposure and the lymphocyte percentage in bronchoalveolar lavage fluid (BALF) solution is increased.

IPH is an exclusive diagnosis. The presence of hemosiderin-containing phagocytic cells in sputum, BALF, or gastric

aspiration liquid should be verified in patients suspected of IPH [9]. Other diseases such as infectious, cardiovascular, or immune-related diseases should be excluded in patients with pulmonary hemorrhage [10]. Lung biopsy is still the gold standard for the diagnosis of IPH.

IPH is typically treated with medications (glucocorticoid and immunosuppressant). Studies have shown that early administration of glucocorticoid can effectively delay pulmonary fibrosis [10]. In this study, hemoptysis recurred following glucocorticoid decrease, indicating that glucocorticoids should be lowered with caution during IPH therapy. In conclusion, IPH is rare among adults, and imaging results on single chest CT are not specific. The longitudinal follow-up would be very extremely beneficial in the diagnosis and assessment of efficacy. Early standardized therapy is essential; which would lead to a better outcome, fewer complications, and reduced sequela.

Disclosures of Conflicts of Interest: T.D. disclosed no relevant relationships. **MG.H.** disclosed no relevant relationships. **XL.C.** disclosed no relevant relationships. **Y.Z.** disclosed no relevant relationships.**BQ.X.** disclosed no relevant relationships.

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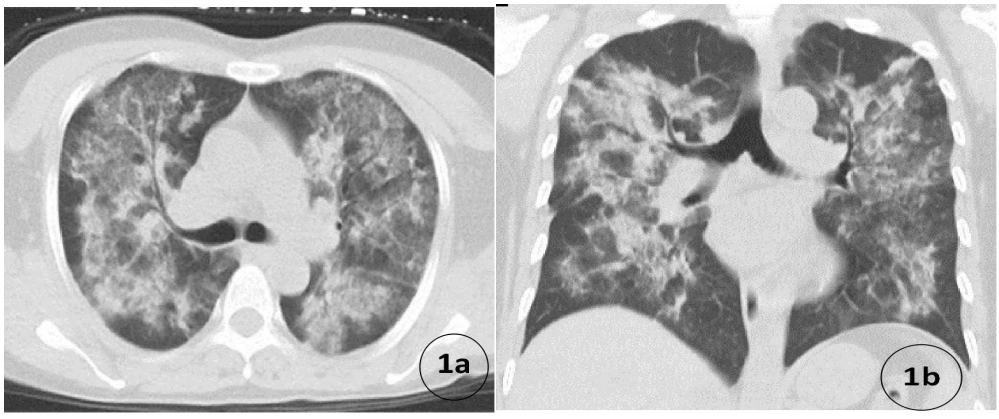


a. b.

Figure 1: (a) Axial and**(b)** coronal non-contrast composite computed tomography(CT) images of the chest shows diffuse ground-glass opacities and consolidation, within the hilar,perihilar,and lower lobe regions,the lung apices and the costophrenic sulciare spared.

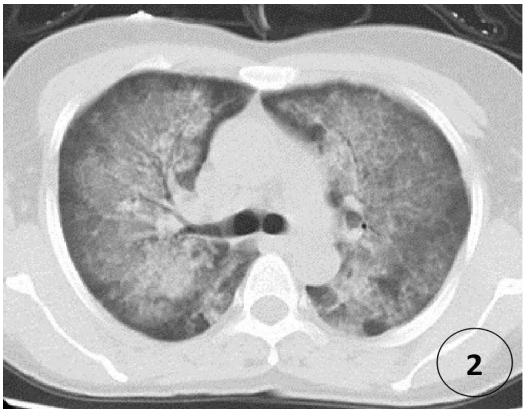


Figure2: Axial NECT chest images after three days treatment with antibiotic demonstrate the lesions are progressive,especially in bilateral subpleural regionsof lung apices.

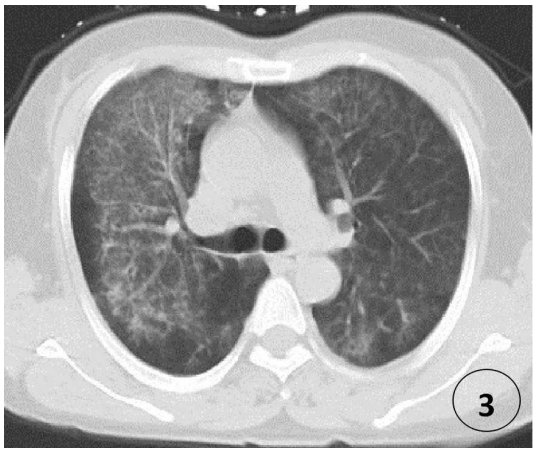
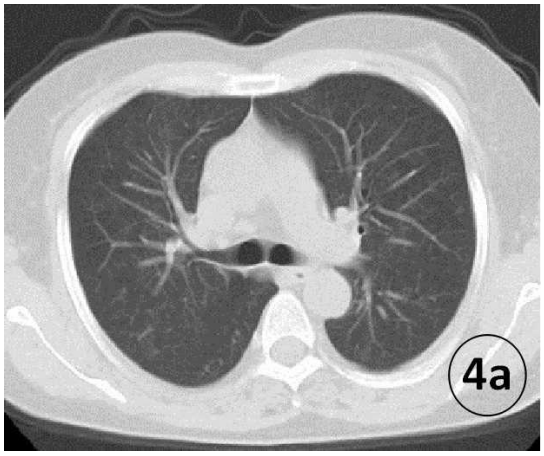


Figure 3: Axial unenhanced chest CT after one and two weeks' treatment with hormone and immunosuppressive show the lesions are obviously absorbed, and vanish.



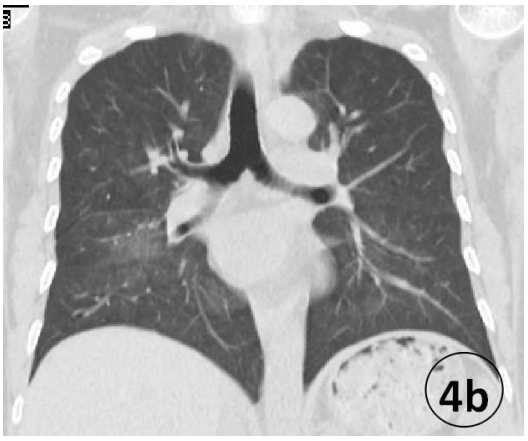


Figure 4: Axial and coronal unenhanced chest CT images after two weeks' treatment show the lesions totally absorb amid lung parenchyma.