Medical Image of the Month: Idiopathic Pulmonary Hemosiderosis

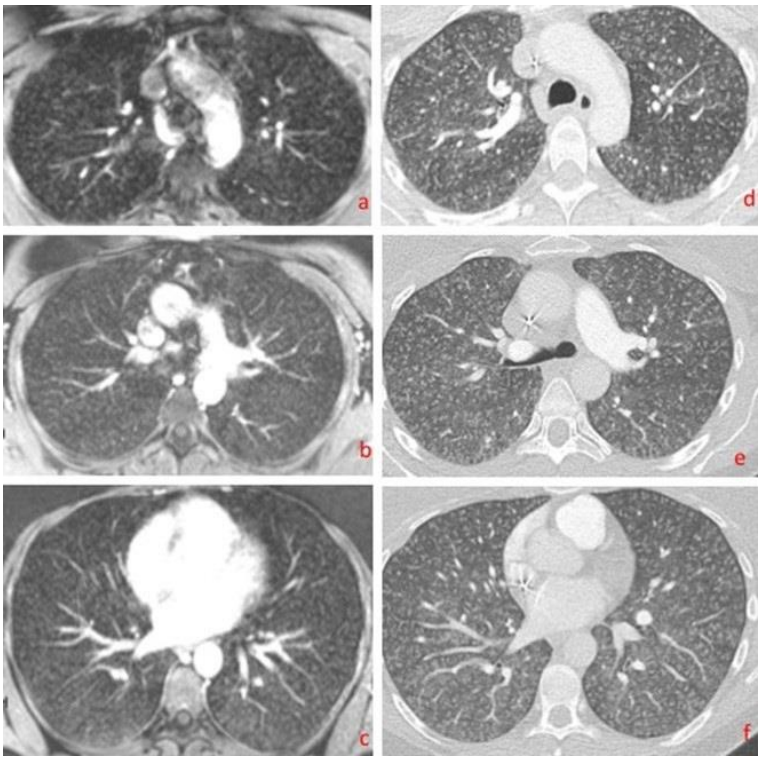

Figure 1. A-C: T1W 3D GRE post contrast multilevel axial images. D-F: CT axial images in a lung window (obtained three years before) both demonstrate innumerable centrilobular nodules consistent with the diagnosis of IPH.

The patient is a 36-year-old woman with a complex medical history including multiple venous thromboembolic events, miscarriages, heterozygous state for factor V Leiden deficiency, and Systemic Lupus Erythematosus. These images have been obtained during multiple admissions for shortness of breath during which she has been diagnosed with pulmonary embolism, anti-coagulation failure, pulmonary hypertension, and intracardiac right to left shunting. Images A-C are T1 weighted MRI axial sections showing centrilobular micronodules which are unchanged when compared to images D-F obtained during a CT scan of the chest three years prior. These findings are consistent with pulmonary hemosiderosis.

Idiopathic pulmonary hemosiderosis (IPH) is a rare condition that occurs with recurrent diffuse alveolar hemorrhage (1). Hemosiderin, a heme byproduct, gradually accumulates within the lung tissue, and can lead to fibrosis (2). IPH has a characteristic triad of hemoptysis, iron deficiency anemia, and pulmonary infiltrates on imaging (2) - although clinical presentation may be highly variable. The gold standard for diagnosis is lung biopsy, although bronchoalveolar lavage has 92% sensitivity of finding hemosiderin-laden macrophages in IPH (2). Classically, the disorder is found in children, but there have been more cases recorded in adults in recent years (3).

Radiographic findings: On chest x ray, areas of air-space consolidation or ground-glass opacities may be seen, usually with a perihilar or lower lung predominance. Consolidations typically clear within 3 days and are replaced by a reticular pattern (4). This may initially resolve but may progress to fibrosis after multiple occurrences - appearing as permanent reticulation or miliary stippling (1). On CT, the subacute phase demonstrates diffuse nodules and patchy areas of ground glass opacification. During an exacerbation, CT shows diffuse, homogenous areas of ground glass attenuation (4). On MRI, T1 images may show diffusely increased parenchymal signal intensity, whereas T2 images may show markedly reduced signal intensity due to the hemosiderin (4). The 3D gradient echo higher resolution MRI sequences in our patient, allowed for the recognition of the chronic micronodular pattern displayed.

Long term, low-dose, glucocorticoids are the main treatment for IPH, with immunosuppressants added on for severe cases. Tapering or reduction of glucocorticoids usually led to recurrence of pulmonary hemorrhage in patients (2). A large number of IPH cases coexist with Celiac disease (known as Lane-Hamilton Syndrome) and a gluten free diet may lead to remission (3).

On imaging, the differential diagnosis is broad, particularly if no remote imaging is available. In our patient's case, the micronodular pattern may be seen with miliary infections, hypersensitivity pneumonitis, some forms of bronchiolitis (particularly smoking related or inhalational diseases). Microangiopathies are also to be considered, such as capillary hemangiomatosis. IPH is a diagnosis of exclusion, and all other causes of diffuse alveolar hemorrhage must first be investigated, such as bronchiectasis, interstitial pneumonia, infections, connective tissue disease, coagulation disorders, systemic vasculitis, and/or anti-GBM disease (3).

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References

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