Case 14941

Eurorad

Idiopathic pulmonary haemosiderosis

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ISSN: 1563-4086
Section: Chest imaging
Area of Interest: Lung
Procedure: Diagnostic procedure
Procedure: Biopsy
Imaging Technique: CT-High Resolution
Imaging Technique: Experimental
Special Focus: Haemorrhage Tissue characterisation
Pathology Case Type: Clinical Cases
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Patient: 6 years, female

Clinical History:

A 6-year-old African American female patient presented with a 3-week history of cough unresponsive to treatment with antibiotics. The patient was born prematurely at 28 weeks and was below the 5th percentile for height and weight at presentation. She has a history of gastro-esophageal reflux disease, aspiration, and recurrent colds.

Imaging Findings:

A frontal chest radiograph (Fig.1) demonstrates bilateral middle and lower lung predominant airspace opacities. Non-contrasted axial CT-images (Fig. 2) obtained a few days later demonstrated multifocal ground glass opacities with denser areas of airspace consolidation scattered throughout the bilateral lungs.

Discussion:

Background:
Idiopathic Pulmonary Haemosiderosis (IPH) is a rare condition in which the patient suffers from recurrent episodes of diffuse alveolar haemorrhage of unknown aetiology [1]. The pulmonary system responds to haemorrhage via activation of alveolar macrophages. These specialised macrophages phagocytose erythrocytes at a rate six times slower than systemic macrophages [2]. This reduced ability to metabolise the haemorrhaged blood ultimately leads to a build-up of haemosiderin [2].

Clinical Perspective:
The pulmonary haemorrhage of IPH gives rise to the symptoms commonly seen in this condition. The clinicalcourse is defined by unpredictable alternations between acute and chronic phases. In the acute phase, alveolar haemorrhage presents as dyspnoea, cough and haemoptysis and is sometimes referred to as an IPH exacerbation. Patients in the chronic phase have similar but slowly resolving symptoms [1].

This patient suffered from all symptoms except for haemoptysis. Imaging is ordered as a non-invasive means of discovering whether the symptom profile is derived from bleeding within the lung tissue [3]. Serology is used in order to exclude autoimmunity, vasculitides, and other potential causes [1]. Pathology is used to confirm the diagnosis of IPH [1].

Imaging Perspective:
Thoracic radiographs are ordered to evaluate for lung opacities that may indicate pulmonary haemorrhage [3]. A thoracic CT is ordered to better characterise and localise the suspected bleed [3]. CT shows ground glass opacities often predominating in the lower lobes [3]. Imaging is paramount to reaching the diagnosis of IPH because it rules out causes of haemorrhage apparent on imaging and allows the surgeon to target the correct pathologic sites for bronchoalveolar lavage (BAL) and peripheral wedge biopsy. The BAL revealed haemosiderin-laden macrophages. The open lung biopsy also showed haemosiderin-laden macrophages with minimal lymphocytic infiltrate and scattered alveolar wall fibrosis. Serologic testing was negative.

Outcome:
IPH is commonly treated with glucocorticoids plus or minus addition of immunosuppressants [4]. She was given oral prednisone and iron supplementation and nutritional advice for the anaemia and low weight, albuterol and fluticasone for asthma, omeprazole for her GERD. The rarity of IPH makes the prognosis difficult to determine although it is believed that children suffer a more aggressive course compared to adults [5]. In conclusion, this patient presented at the peak incidence of childhood with all 3 of the key symptoms of IPH, serology was negative, and pathology confirmed the diagnosis. The patient is doing well with treatment 10 years after diagnosis.

Differential Diagnosis List: Idiopathic pulmonary haemosiderosis., Mitral stenosis, Polyarteritis nodosa, Systemic lupus erythemotosus, Systemic vasculitis

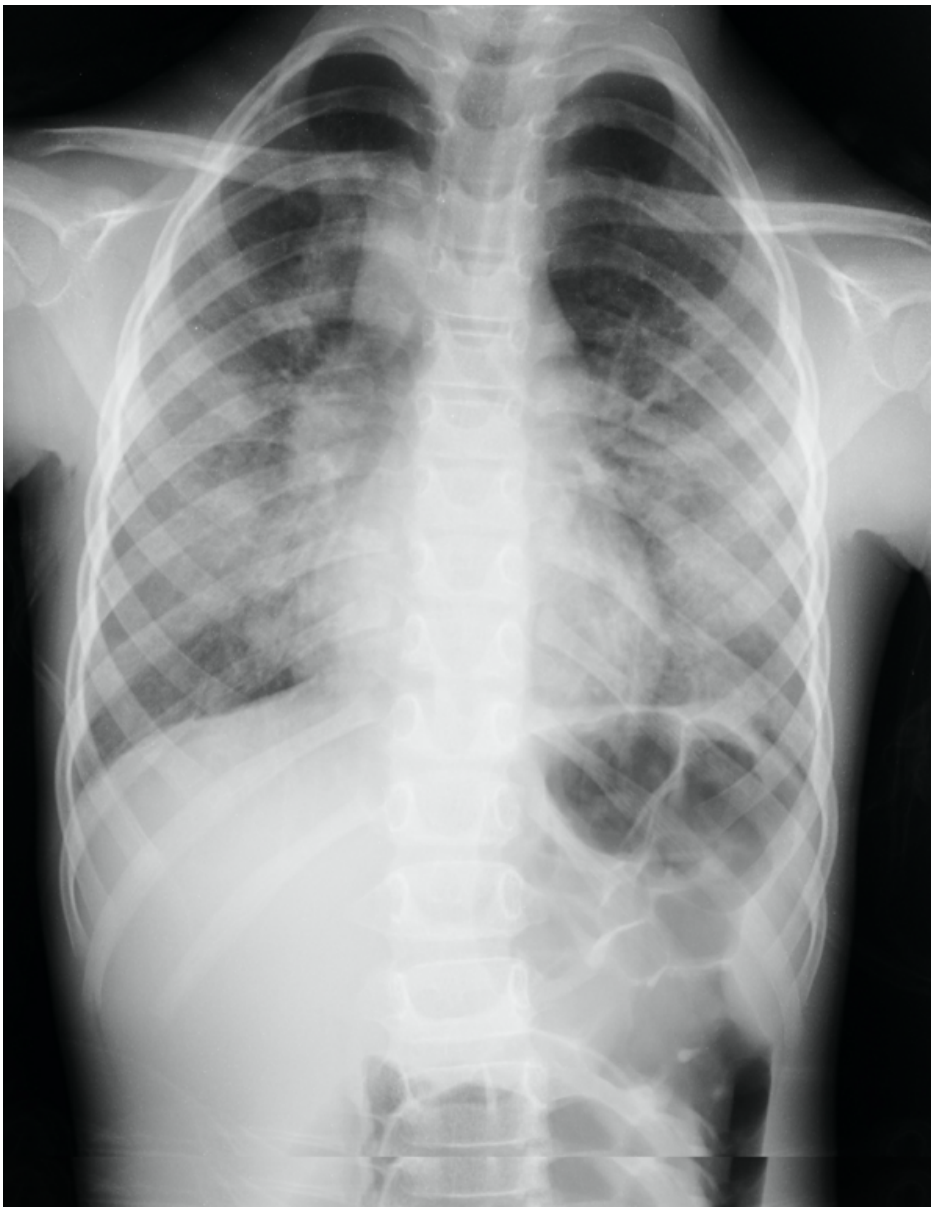
Final Diagnosis: Idiopathic pulmonary haemosiderosis.

References:

O.C Iochimescu, S. Sieber, A. Kotch (2004) Idiopathic Pulmonary Haemosiderosis Revisited. European Respiratory Journal (PMID: 15293620)
G. Custer, et al. (1982) Human Macrophage Hemoglobin-Iron Metabolism in Vitro. American Journal of Hematology (PMID: 7137164)
E. Kocakoc et. al (2003) Pediatric Idiopathic Pulmonary Hemosiderosis Diagnosed by Sputum Analysis: Plain Radiography and Computed Tomography Findings. Karger (PMID: 12634470)
N. Kiper, et al. (1999) Long-term clinical course of patients with idiopathic pulmonary hemosiderosis (1979-1994): prolonged survival with low-dose corticosteroid therapy. Pediatric Pulmonology (PMID: 10213256)
N. Milman (1998) Idiopathic pulmonary haemosiderosis. Epidemiology, pathogenic aspects and diagnosis. Respiratory Medicine (PMID: 10070562)

Figure 1

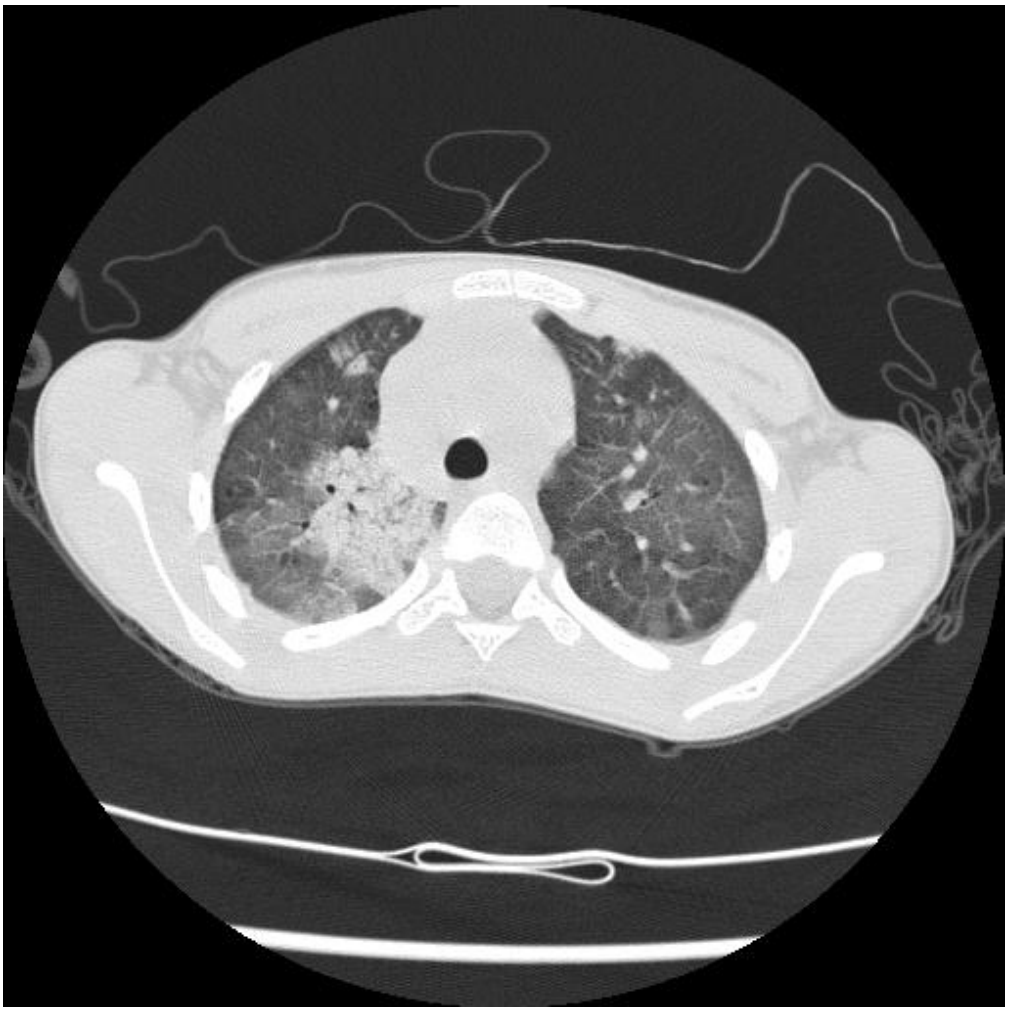
a



Description: Frontal chest radiograph demonstrates bilateral airspace opacities predominantly within the mid and lower lungs. **Origin:** Department of Radiology, Augusta University, Augusta, Georgia

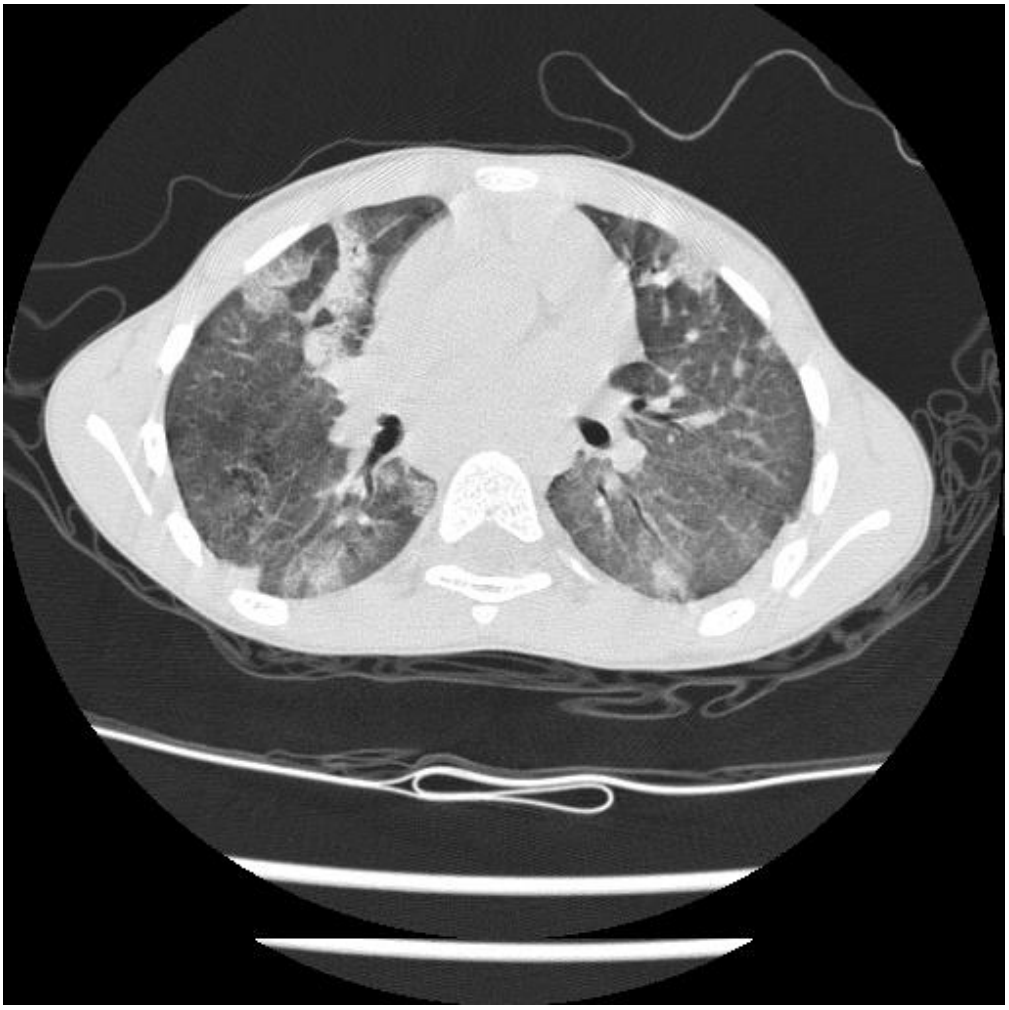
Figure 2

a



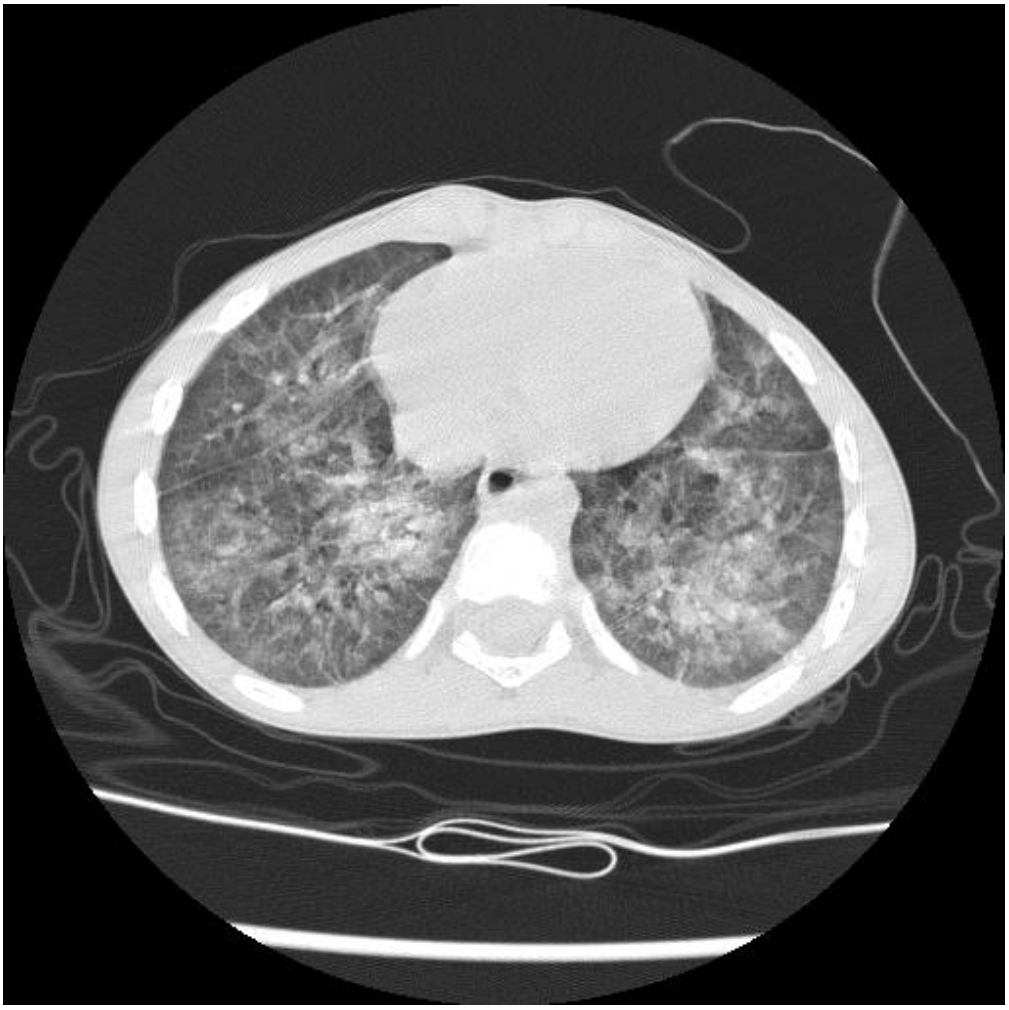
Description: Non-contrasted axial CT demonstrates bilateral ground glass opacities within the right greater than left upper lobes. Denser airspace opacification is present within the posterior right upper lobe. **Origin:** Department of Radiology, Augusta University, Augusta, Georgia

b



Description: Non-contrasted axial CT demonstrates bilateral ground glass opacities within the right greater than left upper lobes. Denser airspace opacification is present within the posterior right upper lobe. **Origin:** Department of Radiology, Augusta University, Augusta, Georgia

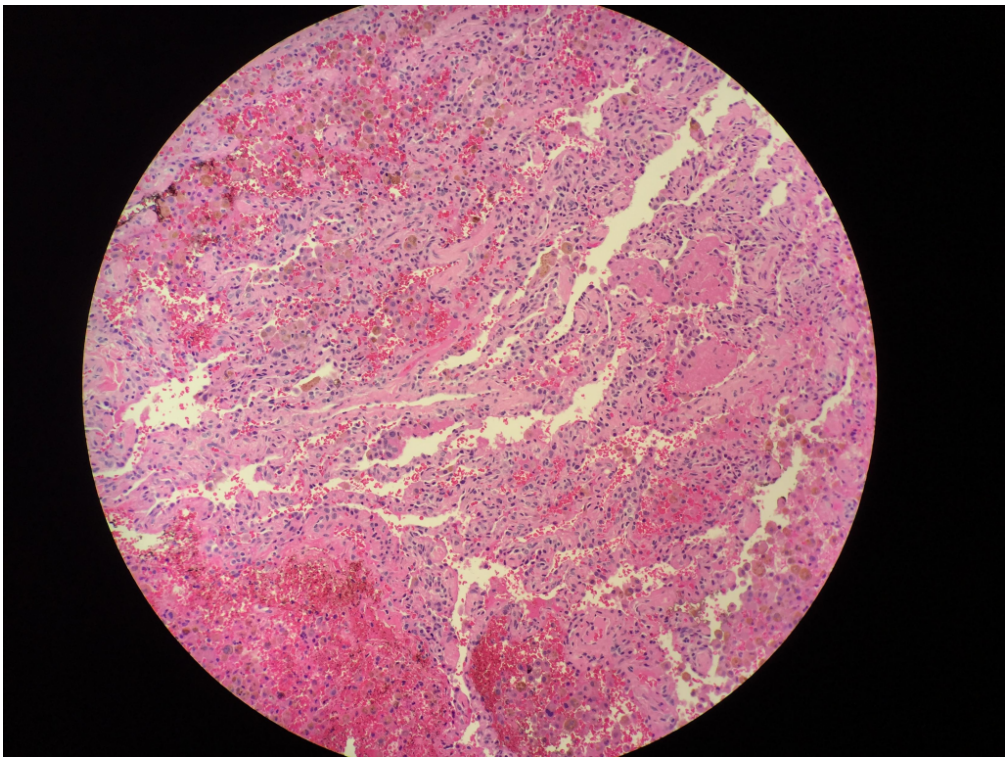
c



Description: Non-contrasted axial CT demonstrates bilateral ground glass opacities within the right greater than left upper lobes. Denser airspace opacification is present within the posterior right upper lobe. **Origin:** Department of Radiology, Augusta University, Augusta, Georgia

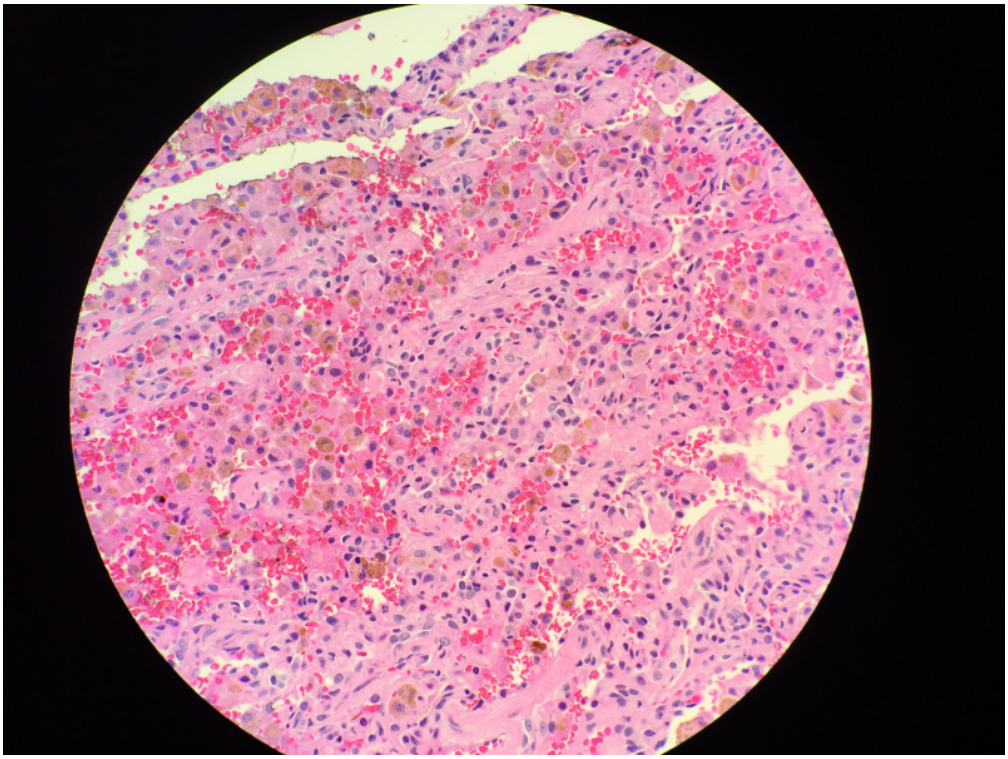
Figure 3

a



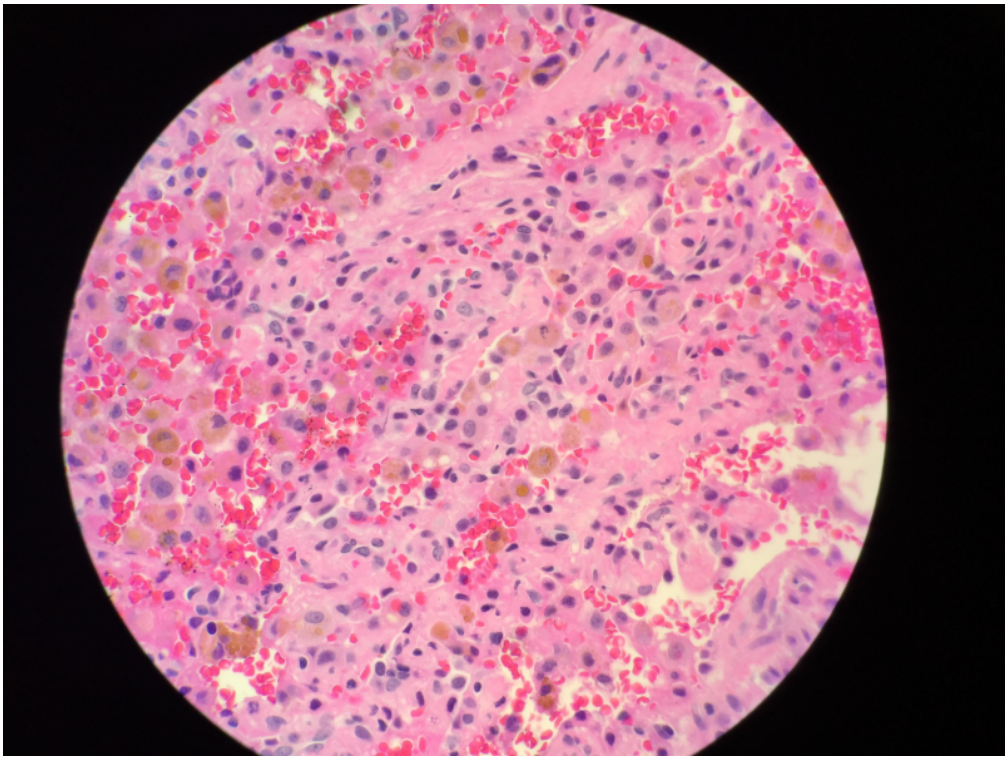
Description: H&E staining at low power demonstrates the presence of hemosiderin laden macrophages. **Origin:** Patel N, Department of Pathology, Augusta University Health, Augusta, Georgia

b



Description: H&E staining at medium power demonstrates the presence of hemosiderin laden macrophages. **Origin:** Patel N, Department of Pathology, Augusta University Health, Augusta, Georgia

c



Description: H&E staining at high power demonstrates the presence of hemosiderin laden macrophages. **Origin:** Patel N, Department of Pathology, Augusta University Health, Augusta, Georgia