

# Delayed-Onset Idiopathic Pneumonia Syndrome Post Hematopoietic Stem Cell Transplantation

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## BACKGROUND

- Idiopathic pneumonia syndrome (IPS) includes a wide range of severe, non-infectious, diffuse lung injuries after hematopoietic stem cell transplantation (HSCT)
- The incidence of IPS post HSCT ranges from 3-15% (1). The median time of onset is 42-49 days post HSCT, and in most reports it occurs within four months after HSCT.
- Late onset IPS (after 4 months) is very rarely reported.
- Diagnostic criteria of IPS include clinical presentation of pneumonia in the setting of diffuse lung injury without evidence of active lower respiratory tract infection (1).
- The prognosis is poor, with a case fatality of 60-95% (2)

## CASE DESCRIPTION

- A 51-year-old female patient with Acute Myelogenous Leukemia underwent myeloablative HLA-haploidentical HSCT 10 months prior to admission.
- The patient presented with cough and progressive shortness of breath for 3 weeks.
- She was found to have hypoxemia requiring oxygen therapy.
- CT showed bilateral, diffuse air space opacities due to a combination of consolidative and ground glass opacifications (Fig 1).
- COVID-19 was negative, and the patient was started on empiric antibiotic therapy.
- Echocardiogram was normal.
- Flexible bronchoscopy with bronchoalveolar lavage did not show significant abnormalities.

## CASE OUTCOME

- Extensive Interstitial lung diseases and infectious work up including BAL was negative (Fig 2).
- Patient's respiratory status deteriorated, she was transferred to the intensive care unit (ICU) and placed on invasive mechanical ventilation and high dose Methylprednisolone.
- Given her unresponsiveness to steroids with further worsening respiratory status, she was given Tocilizumab. Her ICU stay was further complicated with small pneumothorax and pneumomediastinum (Fig 1, G&H).
- She failed to improve with further therapy with intravenous immunoglobulin, Rituximab, and cyclosporine. She continued to deteriorate, and the focus of her care was transitioned to comfort measures. She passed away 38 days after her presentation.

## WORK UP RESULTS

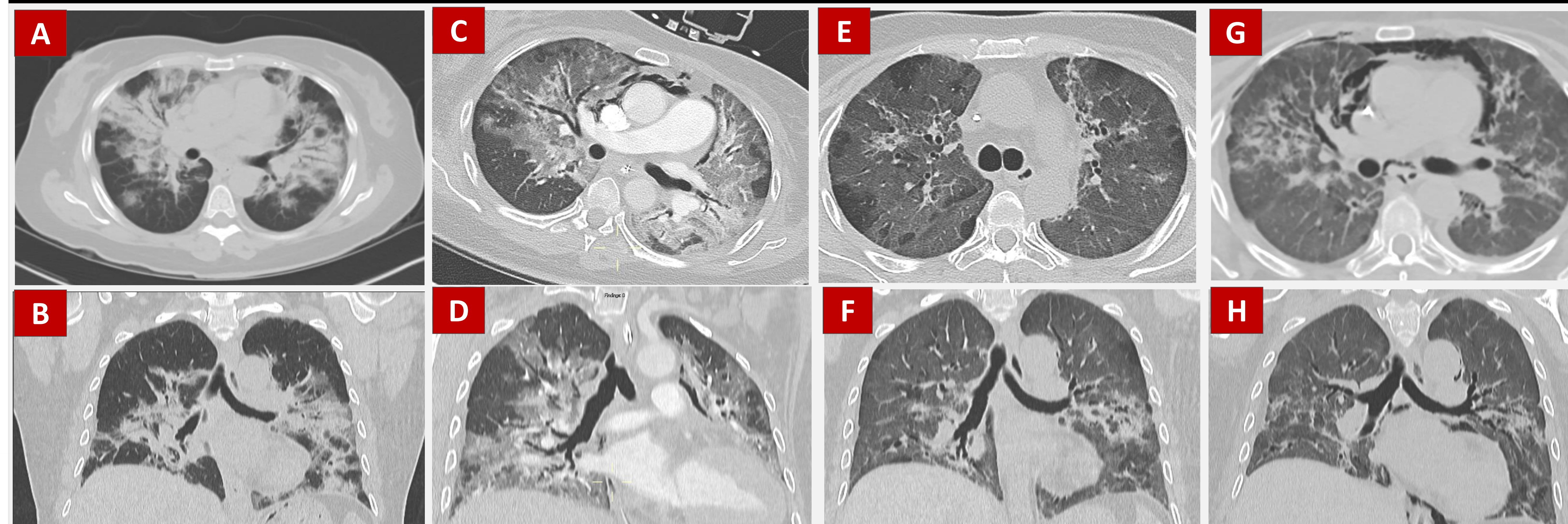


Figure 1: Cross sectional and coronal CT chest images on admission (A&B), week 1 (C&D), week 3 (E&F), week 4 (G&H)

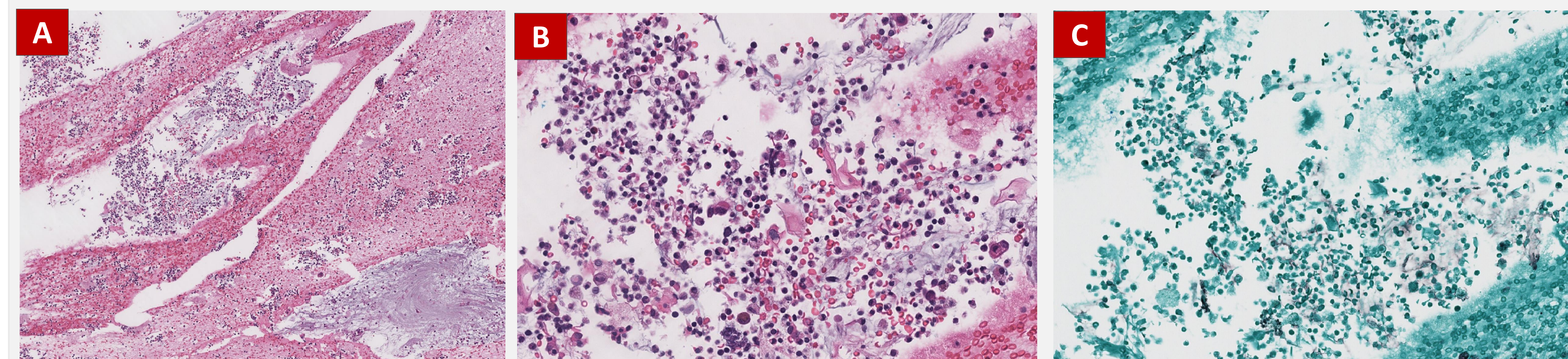


Figure 2: BAL cytology images. A- Low power: Cellular sample with background mucin and blood. B- Mixed acute and chronic inflammation. C- GMS stain is negative for fungi and *Pneumocystis jirovecii*

## DISCUSSION

- Here we present a rare case of delayed-onset IPS, occurring 10 months after HSCT, with rapid progressive deterioration and unresponsiveness to high dose steroids and immunosuppressive agents.
- Although infection should always be suspected in an immunocompromised individual with bilateral pulmonary opacities, IPS can manifest in a patient presenting with signs and symptoms of pneumonia after HSCT.
- Early therapy with systemic steroids and soluble TNF- $\alpha$  inhibitors should be considered when pulmonary dysfunction is determined to be noninfectious, ideally with enrollment in open clinical trials to assess outcomes and clinical response (3).

## CONCLUSIONS

- Clinicians who care for HSCT patients should remain vigilant to the diagnosis of non-infectious pulmonary complications.
- Late-onset IPS should be considered in the differential diagnoses for rapid progressive bilateral lung injury after ruling out infectious etiologies.

## REFERENCES

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