

Abstract

Adult onset Still disease is a rare inflammatory disorder with an unknown etiology. Fever of unknown origin, generalized lymphadenopathy, splenomegaly and evanescent rash are the most common manifestations of Adult onset Still Disease. Pneumonitis and pleurisy are known to occur in the setting of full-blown disease. Adult onset Still disease presenting as pneumonia is a very rare manifestation and even more so to come as first and sole manifestation in the early disease course.

We present a 40-year-old man who had a past history of Adult onset stills disease, presenting with relapsing adult onset Still disease in the form of a severe pneumonia. He came with dry cough, shortness of breathing and pleuritic type chest pain for three days. Examination revealed evidence of a lower lobar consolidation in both sides.

Patient was initially managed as severe pneumonia with poor response to different antibiotics and later he developed evanescent rash and arthralgia. He fulfilled Yamaguchi criteria and responded well to steroid therapy. Later in the disease course he has developed generalized lymphadenopathy with lymph node biopsy evidence suggesting Kikuchi lymphadenitis. This is the first case of Relapsing Still Disease presenting as pneumonia with associated Kikuchi lymphadenitis. Even though pleurisy and pneumonitis are associated with florid disease, parenchymal lung involvement is extremely rare in AOSD.