

Abstract

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Necrotizing Sarcoid Granulomatosis (NSG) is a rare systemic disease characterized by confluent sarcoid-like granulomas with extensive necrosis and vasculitis. It primarily affects the lungs with varying presentations involving extrapulmonary manifestations in other organs such as the liver, kidneys, and eyes. Since its first description in 1973, the etiology and classification of NSG remains unclear. Since its initial characterization, there have been over 130 cases of NSG reported, with even more rarity in the pediatric population with those of the single digits. Clinical presentation and radiologic features are non-specific and can vary widely, hence it is important to obtain pathologic data to examine histologic features and determine a more accurate diagnosis. Similarly, the presentation as described can imitate various disease processes, such as infection, hypersensitivity reactions, and oncologic processes, which must be excluded before diagnosing NSG. This, alongside its rarity, further makes the proper diagnosis of NSG challenging. The aim of this review is to evaluate and differentiate NSG from other granulomatous diseases processes and determine the diagnostic criteria. The case review is to discuss uncommon manifestations of NSG in pediatric patients, which are rarely described. We present a case of a 17-year-old female presenting with abdominal pain, significant fatigue, fevers of unknown source with hepatosplenomegaly and hepatosplenic lesions found on CT scan and extensive negative infectious workup that ultimately led to a unifying diagnosis of NSG. To the best of our knowledge, there have been no prior cases in the literature describing a similar presentation, notably without respiratory involvement, in a pediatric patient.