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Perspective

Necrotizing Lymphadenitis and Hyperinflammatory Complications

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DESCRIPTION

Kikuchi-Fujimoto disease, commonly referred to as Kikuchi's disease, is an uncommon, self-limiting disorder characterized by regional lymphadenopathy, fever and systemic symptoms. The disease is primarily observed in young adults, with a higher prevalence in women. Histologically, it is marked by necrotizing lymphadenitis with a predominance of histiocytes and plasmacytoid dendritic cells. Clinical presentation is often nonspecific, which makes early recognition challenging. Most patients experience cervical lymph node enlargement, low-grade fever, fatigue and occasionally, night sweats.

Kikuchi's disease generally follows a benign course, resolving within one to four months without aggressive treatment. However, rare cases are complicated by the development of secondary Histiocytic Lymphohistiocytosis (HLH), a severe hyperinflammatory syndrome characterized by excessive immune activation and multi-organ involvement. Secondary HLH represents an extreme systemic response, often triggered by infections, autoimmune disorders, or malignancies. In the context of Kikuchi's disease, it reflects an exaggerated immune reaction, where macrophages and cytotoxic T cells produce high levels of pro-inflammatory cytokines, leading to tissue damage and organ dysfunction.

Diagnosis of Kikuchi's disease with secondary HLH requires careful integration of clinical, laboratory and histopathologic findings. Lymph node biopsy remains the gold standard for confirming Kikuchi's disease, revealing characteristic necrotizing lesions without neutrophilic infiltration. Immunohistochemistry can support the diagnosis by highlighting histiocytes and plasmacytoid dendritic cells. For HLH, diagnostic criteria established by the Histiocyte Society are commonly used, which include a combination of clinical and laboratory parameters. Clinicians must maintain high suspicion for HLH in patients with Kikuchi's disease who present with persistent fever, cytopenias, hepatosplenomegaly and abnormal laboratory indices suggestive of systemic inflammation.

The management of Kikuchi's disease is largely supportive. Symptomatic treatment includes nonsteroidal anti-inflammatory drugs for fever and pain and short courses of corticosteroids may be considered in severe or persistent cases. In contrast, secondary HLH requires prompt immunosuppressive therapy due to its aggressive course. Standard treatment regimens often include corticosteroids, etoposide, cyclosporine, or intravenous immunoglobulin. Early initiation of therapy is associated with improved outcomes, whereas delayed treatment may lead to multi-organ failure and increased mortality.

Prognosis in isolated Kikuchi's disease is generally excellent, with spontaneous resolution within a few months. Recurrence is uncommon but has been documented in a minority of cases. When complicated by secondary HLH, prognosis depends on early recognition and treatment intensity. Despite the severe nature of HLH, patients who receive timely therapy may achieve complete recovery. Follow-up involves monitoring for recurrence of lymphadenopathy, systemic symptoms and laboratory abnormalities. Long-term complications are uncommon but may include persistent cytopenias or autoimmune manifestations.

Differential diagnosis is an important aspect of clinical management. Kikuchi's disease must be distinguished from infectious lymphadenitis, lymphoma, systemic lupus erythematosus and other autoimmune or neoplastic conditions. Misdiagnosis can lead to unnecessary interventions, including prolonged antibiotic therapy or chemotherapy. Awareness of the typical clinical course, combined with histopathological confirmation, helps avoid mismanagement. For secondary HLH, distinguishing it from sepsis or other hyperinflammatory states is critical, as delayed recognition can result in rapid clinical deterioration.

Epidemiologically, Kikuchi's disease is more common in Asia, although cases have been reported globally. Female predominance is notable and the disease typically affects individuals in their second to fourth decades of life. Associations with autoimmune conditions, such as systemic lupus erythematosus, have been observed, suggesting overlapping immunological pathways. HLH secondary to Kikuchi's disease

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does not appear to exhibit a clear demographic pattern but may be underreported due to its rarity.

In conclusion, Kikuchi's disease is an uncommon, self-limiting lymphadenitis that may rarely be complicated by secondary histiocytic lymphohistiocytosis, a severe hyperinflammatory syndrome. Clinical awareness, careful laboratory evaluation, and histopathological confirmation are essential for timely diagnosis. Management of Kikuchi's disease is generally supportive, while secondary HLH requires aggressive immunosuppressive therapy. Prognosis is favorable in isolated cases, and early intervention in HLH can significantly improve outcomes. Continued reporting and systematic study of cases will help refine diagnostic criteria, identify high-risk patients and guide therapeutic approaches.