

Hidden in Plain Sight: Castleman's Disease Presenting as Fever of Unknown Origin

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Case:

A 50-year-old female with a history of Crohn's disease presented to the emergency department with a two-day history of fevers, left ear and throat pain, nuchal rigidity and frontal headache. On presentation, she was febrile, tachycardic with a leukocytosis and lactic acidosis. CT neck ruled out peritonsillar abscess for reported dysphagia and revealed a prominent left cervical lymph node. She was started on empiric meningitis coverage and transitioned to broad spectrum antibiotics after a negative lumbar puncture, but she continued to have fevers. She developed acute renal failure thought to be iatrogenic from antimicrobials and IV contrast. Infectious, rheumatologic and endocrine workups for fever of unknown origin (FUO) were unremarkable. On day five of hospitalization, she developed right-sided neck pain. Ultrasound showed bilateral cervical lymphadenopathy and excisional biopsy was performed. Sections of the lymph node show scattered follicles with regressed and involuted germinal centers composed mostly of follicular dendritic cells, and containing few lymphocytes with sclerotic blood vessels. The interfollicular spaces exhibit prominent vascular proliferation with hyalinized walls, suggesting Castleman's disease (CD). CT chest did not show any evidence of lymphadenopathy. A final diagnosis of unicentric CD (UCD) was rendered. She was discharged with clinical improvement after lymph node excision.

Discussion:

Here we report a challenging case of UCD which presented with acute onset of suspected meningitis and met criteria for systemic inflammatory response syndrome (SIRS). CT neck revealed a single prominent lymph node, presumably due to reactive adenopathy from infectious etiology; however, fevers persisted beyond the expected timeframe on empirical antibiotics. An extensive FUO workup revealed a diagnosis of CD, which is a rare group of lymphoproliferative disorders associated with hyperactivation of the immune system and is classified as unicentric and multicentric. Multicentric CD affects multiple lymph node regions and is associated with systemic inflammatory symptoms, including weight loss, night sweats and generalized lymphadenopathy. In contrast, UCD affects only a single lymph node region, and its inflammatory presentation is usually mild. Patients with UCD are usually asymptomatic except for localized lymphadenopathy. Our patient's acute presentation of UCD with septic manifestations and without prior systemic "B" symptoms builds on previous case reports of this rare disease and expands it further. UCD can present with systemic inflammation and is typically managed with surgical removal of the affected lymph node. Furthermore, our case demonstrates the importance of considering a lymphoproliferative disorder in the differential diagnosis for FUO, even in the setting of a single enlarged lymph node.