10.7861/clinmed.23-6-s6

A rare presentation of Castleman disease

Authors: Aswin Surjit,[A] Sajira VK,[A] Shahanas PS[A] and Suhan Ahmad Najeeb[A]

Introduction

Castleman disease is a rare disease of the lymphoproliferative system with an estimated incidence of 5–16 per million patient-years. It commonly presents as fever with lymphadenopathy. Very rarely it can manifest as an autoimmune disease like rheumatoid arthritis, mixed connective disease etc. Here we present a rare case of Castleman disease that presented with many features of systemic sclerosis.

Case report

A 49-year-old man, who is a medical professional, presented with polyarthritis involving wrist joint and interphalangeal joints of hands. He gives history of occasional discolouration of fingers suggestive of Raynaud's phenomenon for the past year. On examination there was no pallor or lymphadenopathy, and vital signs were normal. His head to foot examination showed salt and pepper pigmentation in scalp and forehead and sclerodactyly with no swelling of hands. His systemic examination fine basal crackles were heard on chest auscultation. His blood investigations showed Hb-11.5 g/dL, ESR-65 mm/hr, random blood sugar 145 mg/dl, RA factor and ANA positive, extractable nuclear antibodies (ENA) panel was positive for U1RNP. Other laboratory investigations were normal. The HRCT thorax confirmed features of interstitial lung disease. In view of the clinical features and U1RNP being positive, a diagnosis of limited systemic sclerosis / MCTD was made and the patient was started with steroids and mycophenolate mofetil. His symptoms subsided for 6 months. He later presented with fever of 2 weeks' duration. A detailed evaluation including PET-CT showed lymphadenopathy in right axilla, left lower paratracheal, left external iliac and peripancreatic areas. An excision biopsy from an axillary lymph node reported Castleman disease – plasma cell variant. His HIV and HHV 8 tests were negative. His Interleukin 6 (IL6) levels were 11.34 H pg/mL (normal 0.00–7.00). Bone marrow aspiration and biopsy were normal. Kappa and lambda chains were within normal range. IgG levels were 1,800 mg/dl and serum electrophoresis did not show any M band. The skin biopsy done from the hypopigmented patch was positive for AA amyloidosis. The patient was started on rituximab pulse therapy, he responded well and symptoms subsided. Further imaging showed a reduction in the size of lymph nodes. He was lost to follow up for 4 years and again he presented with fever and joint pain. He was diagnosed to have relapse of the disease and was started on tocilizumab in view of the severity of the disease. Meanwhile, the patient developed invasive fungal pneumonia and succumbed to death.

Conclusion

Castleman disease can present as or mimic connective tissue disease or malignancy. Our patient has multicentric Castleman disease – plasma cell variant with secondary amyloidosis – and presented with most of the features of systemic sclerosis. At times, it can be a diagnostic dilemma when presenting in association with an autoimmune disease. Hence in such cases, when clinical features change or there is difficulty in treating an autoimmune connective tissue disorder, or a strong clinical suspicion of lymphoproliferative disorder, malignancy or other alternate diseases must be considered. ■

Authors: [A]Aster Medcity