Neurolupus in a previously healthy young Caucasian male

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Introduction

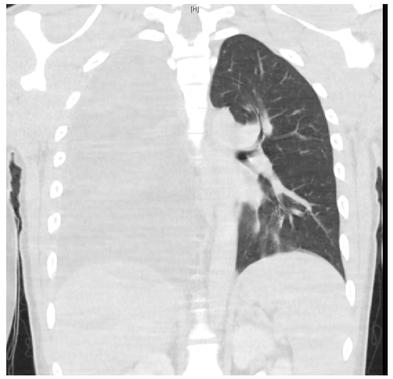
- A 25 year old male presented with a wide variety of physical complaints, most concerning being vision changes, headache and lethargy. He was found to have neurolupus as the presenting symptom of new onset systemic lupus erythematosus (SLE).
- SLE can present with a wide variety of systemic symptoms such as headache, arthralgias, myalgias, malaise, fevers and appetite changes as it can affect any part of the body.[1]
- Meningitis is an uncommon manifestation in SLE especially as the first presenting symptom, estimated at 1.5%.[2]
- Herein we present a rare case of a young Caucasian male with SLE, as the United States incidence rate is 0.8 per 100,000 person-years.[3]

Case Presentation

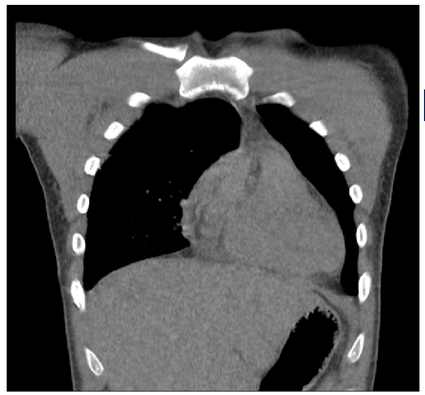
- A previously healthy 25 year old male presented with concerns of a headache and fevers.
- Further history revealed migratory arthralgias, primarily of knees and elbows, central chest pain worse with leaning forward, intermittent fevers and rashes.
- Examination revealed diffuse non-blanchable bright pink patches of bilateral arms, diffuse bilateral supraclavicular and axillary lymphadenopathy, nuchal rigidity, and positive Kernig and Brudzinski signs.
- Laboratory studies were significant for microcytic anemia (hgb 9.7g/dl, MCV 79.7), thrombocytosis (580/mcl) and hypocalcemia (7.8 mg/dl). LP analysis showed 338 total nucleated cells, 83% neutrophils, glucose 29 and total protein 148 and was culture negative.
- He was empirically treated with ceftriaxone and vancomycin without clinical improvement, prompting further testing (**figures 1-3**).
- Serum infectious panel was negative for syphilis, Borrelia burgdorferi, Ehrlichia, West Nile, Herpes simplex, human immunodeficiency virus, Rickettsia, Typhus, and Varicella Zoster.
- Anti-nuclear antibody screen revealed a 1:1280 titer of nuclear, speckled antibodies and anti-double strand DNA antibody was elevated 1946 IU/mL, confirming the diagnosis of SLE.
- His antimicrobials were transitioned to immunomodulating methylprednisolone then oral prednisone on discharge after evidence of significant clinical improvement.

Figures

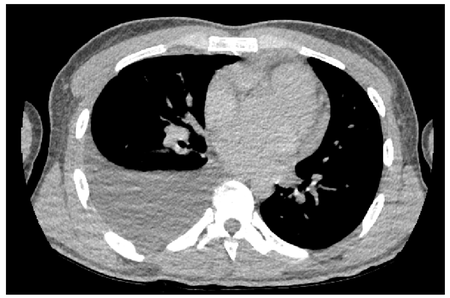
Pleural and pericardial effusions



- ***Figure 1. Large Right Pleural Effusion in coronal view.***



- ***Figure 2. Trace Pericardial Effusion in coronal view.***



- ***Figure 3. Trace Pericardial Effusion and Right Pleural Effusion in transverse view.***

Discussion

- This case describes one of the rarest initial clinical presentations of SLE, neurolupus as aseptic meningitis, even rarer in this demographic, a Caucasian male.
- The European League Against Rheumatism and American College of Rheumatology have created with EULAR/ACR criteria for SLE diagnosis for this multifaceted disease, met by this patient with his elevated ANA and clinical manifestations.[4]
- Neurolupus manifestations include chorea, delirium ,migraine headache, psychosis, seizures and aseptic meningitis with estimated total incidence of as low as 12%.[5]
- SLE aseptic meningitis pathogenicity remains unclear, it may be linked to the vasculitis inflammation within the CNS.[6]
- Most cases are attributed to anti-lupus therapies, this patient had not been exposed to IVIG, sulfa-antimicrobials, OKT3 monoclonal antibodies.
- Here is a rare case of an abnormal demographic for SLE, presenting in one of the rarest ways, aseptic meningitis.

Conclusions

- Neurolupus is rare, and aseptic meningitis an even rarer initial presentation of SLE, 1.5% and SLE should be maintained on a differential of unidentified aseptic meningitis.[2]
- SLE should be considered in young patients with nonspecific autoimmune symptoms as early identification and treatment leads to improved quality and quantity of life.

References

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