A retinal change that can mimic many conditions in a young patient with PUO

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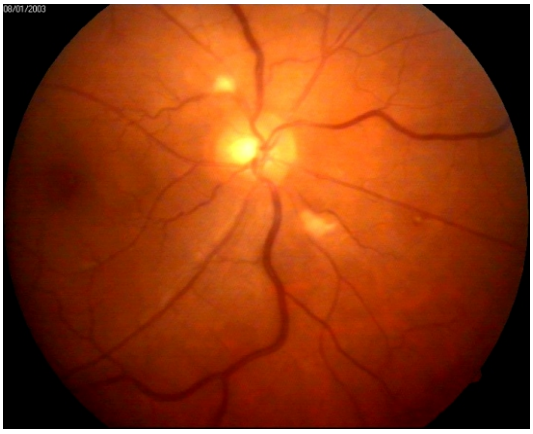
Case history

A 40-year old male developed a persistent febrile illness of unknown aetiology for three weeks of duration with severe sore throat, headache, blurring of vision, myalgia, multiple large joint pain and swelling including both knees, ankles and shoulders. His medical and family histories were unremarkable. On examination he was febrile (39.8^0C) and there were cervical lymphadenopathy and mild hepato-splenomegaly. He had no skin rash. Rest of the physical examination was unremarkable but the examination of optic fundi showed the following abnormality.



What is your diagnosis?

1. Infective endocarditis
2. Systemic lupus erythematosis
3. Adult-onset still's disease
4. Vasculitis
5. HIV
6. Haematological malignancy

Discussion

Laboratory test showed normochromic normocytic anaemia (Hb - 8.9mg/dl), neutrophil leucocytosis (24000/µl with 86% neutrophils), 404,000/µl platelet, ESR of 126mm, and CRP of 78mg/dL. Liver and renal function tests, serum calcium were within normal limits. Screening for TB, blood cultures and urinalysis were negative. 2D echocardiogram was normal while US scan abdomen and the CT imaging of chest and abdomen showed only mild hepato-splenomegaly. Retroviral studies, Rheumatoid factor, ANA, anti CCP, anti ds-DNA antibody, ANCA, hepatitis screening were all negative. Bone marrow biopsy and aspiration showed reactive marrow. Serum ferritin was 3089 ng/ml.

Unifying diagnosis: Adult-onset still's disease (ASOD) with Purtscher-like retinopathy

ASOD was diagnosed according to the Yhamagushi criteria consisted of fever, arthralgia, typical rash and leucocytosis as major criteria, and sore throat, lymphadenopathy and or splenomegaly, liver dysfunction in the absence of rheumatoid factor and ANA as minor criteria. ASOD is diagnosed when ≥ 5 criteria including ≥ 2 major criteria are present with a 96.2% sensitivity and 92.1% specificity. Laboratory data typically reveal marked hyperferritinaemia, anaemia, thrombocytosis and leucocytosis.

Purtscher's retinopathy is an occlusive micro-vasculopathy associated with cranial trauma or thoracic compression. When trauma is not the aetiology, it is called purtscher-like retinopathy. Purtscher-like retinopathy is a rare association of AOSD. He was treated with indomethacin and steroids and showed a dramatic clinical response. After one week of treatment his serum ferritin was 708 ng/mL and the retinal exudates showed an improvement.