History

A 51 year-old HIV-positive homosexual man on HAART presents with two liver lesions on PET scan and MRI measuring 1.4 cm and 1.6 cm. Liver tests are elevated with an alkaline phosphatase of 745, AST 140, and ALT 398. He carries a history of Burkitt lymphoma of the submandibular region for which he is on chemotherapy. Because of concern for recurrent Burkitt lymphoma, a liver biopsy of one of the lesions is performed.
Inflammatory Pseudotumor of the liver

- Spindle cell proliferations with inflammation that can be predominantly fibrohistiocytic or lymphoplasmacytic.
- Have gone by a variety of names, including plasma cell granuloma, xanthogranuloma, fibroxanthoma, and histiocytoma.
- Can be a non-specific term for a reparative lesion in the liver or elsewhere. For instance, a similar process can represent biopsy next to an abscess.
- When faced with IPT, several entities that should be considered which resemble IPT.

Inflammatory Pseudotumor of the liver

- Tumors that can be inflamed
  - Angiomyolipoma
  - Sarcomas
  - Hodgkin’s lymphoma (etc.)
Angiomyolipoma

- Mesenchymal tumor that is part of a family of tumors known as PEComa (perivascular epithelioid cell).
- Generally benign tumor of kidney and less often liver, associated with tuberous sclerosis (kidney > liver) composed of thick-walled vessels, adipocytes, and myoid cells.
- Myoid cells can be spindled to epithelioid, and stain with muscle markers (desmin, SMA) as well as melanoma markers (HMB-45, melan-A).
- Uncommonly, they can have an inflammatory component, usually lymphocytes, as well as plasma cells, histiocytes, and foamy histiocytes. In one series, 6 of 55 AMLs had IPT appearance (focal in 3, predominant in 3). Another series of 5 inflammatory AML described inflammation in greater than half of the tumor, mimicking IPT.

Inflammatory Pseudotumor of the liver

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  - Angiomyolipoma
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  - Hodgkin’s lymphoma (etc).
- Tumors that were once part of the spectrum of lesions known as “inflammatory pseudotumor” but that are now considered specific entities
  - Inflammatory myofibroblastic tumor
  - Follicular dendritic cell tumor

Inflammatory myofibroblastic tumor

- Predilection for children and young adults.
- Usually affects abdominopelvic region, lung, or retroperitoneum.
- Up to 30% have constitutional symptoms (fever, weight loss, malaise).
- Rearrangements in kinases found in about 60%, usually ALK1, but also ROS1 and others.
- ALK1 immunohistochemistry positive in 50%, particularly in tumors occurring in children or young adults
- Absent ALK1 staining does not exclude the diagnosis, esp in adults.

Follicular dendritic cell tumor, EBV related

- EBV-driven intra-abdominal FDC tumors occur almost exclusively in liver and spleen.
- Spindle cells, lymphocytes, and plasma cells. Spindle cells have oval nuclei, vesicular chromatin, and distinct central nucleolus. When binucleate and atypical, they can mimic Reed-Sternberg cells.
- Immunohistochemistry shows positive expression of dendritic cell markers (CD21 and CD35) and ISH for EBER is positive in spindle cells.
Inflammatory Pseudotumor of the liver

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- Tumors that were once part of the spectrum of lesions known as “inflammatory pseudotumor” but that are now considered specific entities
  - Inflammatory myofibroblastic tumor
  - Follicular dendritic cell tumor
- IgG4-related inflammatory pseudotumor
  - Often perihilar location, predominantly men, associated with ulcerative colitis.
  - Lymphoplasmacytic type, numerous IgG4 plasma cells, eosinophils, obliteratorive phlebitis

Syphilis

- Infection by Treponema pallidum
- The stages of syphilis
  - Primary: local infection, chancre.
  - Secondary: dissemination of the organism throughout the body → rash, malaise, sore throat, headache, low-grade fever, muscle aches.
  - Tertiary: After a variable latent period, the development of gummas, neurosyphilis, or cardiovascular syphilis.
- Syphilis can affect the liver in various ways
  - Secondary syphilis can cause hepatitis
  - In tertiary syphilis, gummas can occur in the liver that, when they heal/contract, result in hepatic lobatum
  - Inflammatory pseudotumors

Syphilis and the liver

- Syphilis is on the rise, particularly among gay men. But a recent news report noted the rise in congenital syphilis as well.
- 20-40% of HIV positive patients with syphilis develop hepatitis.
- Many case reports of syphilitic hepatitis involve gay men. HIV positivity and “unsafe” sexual practices are associated with syphilis.
- Presents with elevated liver tests, with disproportionately high alkaline phosphatase.
- Various histologic features have been described, including:
  - Granulomas
  - Pericholangitis with duct injury – mimic of drug reaction
  - Inflammatory cells in sinusoids

Syphilitic inflammatory pseudotumor

- Inflammatory pseudotumors related to syphilis have been reported in the skin, lung, meninges, lymph nodes, and liver.
- We reported 3 syphilis-related IPTs in the liver, in HIV-positive homosexual men (Hagen et al. AJSP 2014)
  - Variable cellular, with areas of bland spindle cells in a storiform pattern
  - Mixed inflammation: neutrophils were frequent and abscesses were present in two cases. Only one case had many plasma cells.
  - Background features similar to syphilitic hepatitis
    - Portal edema with duct injury and cholangitis/pericholangitis
    - Inflammatory infiltrate within sinusoids
    - One had granulomas
  - Possibly due to transport and massive localization of spirochetes to the liver.
42 year old man on PrEP (Pre-exposure prophylaxis), who presented with fever, jaundice, and a rash.
Antibody for Spirochetes

- Far more sensitive than Steiner or other histochemical stain for Treponema
- Depending on vendor, specificity may vary. For example, Biocare antibody cross reacts with Borrelia burgdorferi (the agent of Lyme disease) and....

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