Body Cavity Tumors

Key Points:
1. Body cavity lymphomas, also called primary effusion lymphomas are liquid phase tumors that invade body cavities and present as effusions.
2. HHV8, and to some extent EBV, are implicated as important contributors to pathogenesis.
3. There is no good treatment for primary effusion lymphomas, although chemotherapy is often tried. In HIV patients the prognosis is particularly poor.

What is it?
Body cavity tumors, also called primary effusion lymphomas, are a type of lymphoma that has a predilection for body cavities such as the pleural, peritoneal, or pericardial spaces. It is one of the three, and least common, subtypes of AIDS-related lymphomas (the others being NHL and primary CNS lymphoma). As we learn more about the disease, there appear to be two categories: HHV8 positive and HHV8 negative tumors.

Who is at risk for this disease?
- AIDS: especially in pts with CD4 < 100
- HHV8 positive: Pts with Kaposi’s sarcoma, especially in Africa, Mediterranean countries, and those of Jewish descent. Men more frequently than women
- HHV8 negative: Women more than men, higher EBV prevalence, Japanese.

What is the pathogenesis?
Not well understood. The cells are monoclonal B cells that express CD38 and appear to have some genomic material from HHV8 and/or EBV.
HHV8 positive: Believed to be a cytokine mediated disease. For example, release of IL-6 through HHV8 causes plasmacytosis and angiogenesis which essentially then acts as an autocrine growth factor. Vascular leakage mediated by the tumor cells and loss of immunosurveillance are also implicated. In HIV patients, up to 80% are also infected with EBV.
HHV8 negative-EBV positive: It is believed that chronic inflammation and irritation can lead to clonal stimulation of EBV-latently infected cells. There is some similarity to Burkitt’s lymphoma. Liver disease from HCV or alcohol (and even irritation of a cavity from a ventriculoperitoneal shunt!) have been cited as providing antigenic stimulation of cell proliferation.

How does the patient present?
With pleural or pericardial effusions, ascites, joint effusions, or occasionally meninges. B-type symptoms are less common.

Do imaging studies help?
Basically, they’ll help you find the effusion but won’t help you differentiate it. Sometimes there is thickening of the serosal surfaces, but there is not parenchymal involvement, lymphadenopathy, or solid masses.

What about cytology?
Almost always positive, as these tumors grow in the liquid phase. By microscope they show bridging diffuse B cell lymphoma and anaplastic large cell lymphoma. Flow cytometry shows expression of CD 45 and CD 30 but B and T cell-associated antigens are not usually present. Molecular evaluation for HHV8 and EBV should be performed.

Do I have stage these patients?
There’s not any staging to be done, as the tumor rarely spreads beyond the body cavity. Local destruction of surrounding tissues can happen.
How do you treat primary effusion lymphomas?

Definitely still a work in progress. Some cited treatments include:

- Chemotherapy: a modified CHOP regimen (cyclophosphamide, doxorubicin, vincristine, and prednisone for 5 days every 28 days for 4-6 cycles). You should see shrinking of the effusion after a few cycles if it’s going to work. When CHOP fails, you can try pegylated liposomal doxorubicin or daunorubicin or local radiation therapy.
- HAART is certainly important in HIV patients although has not definitively shown to help
- Anti CD20 antibodies

What is the prognosis?

Not good in HIV positive patients. Mean survival in a small study was 75 days even with aggressive treatment. Those with non-AIDS primary effusion lymphoma have shown better survival and in some cases remission from chemotherapy, simple drainage, treatment with anti-CD20 antibodies, or other approaches.

References:
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