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How A Pleural Effusion Workup Resulted in an AIDS-related Lymphoma Diagnosis in an HIV Negative Patient

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Abstract

Primary effusion lymphoma, a subtype of Non-Hodgkin's lymphoma, is linked with HHV-8. This disease is associated with immunocompromised patients such as those who have HIV, solid organ transplant carriers, or chronic hepatitis C. Primary effusion lymphoma frequently commences in the pleura, pericardium, and peritoneum.

A 77 year old middle eastern male with history of tobacco usage was admitted for fatigue and hypotension. Chest X-ray revealed bilateral pleural effusions. He had received a diagnostic and therapeutic thoracentesis. Cytology revealed that it was positive for HHV-8, a diagnostic criteria for primary effusion lymphoma. The patient was tested for HIV, HTLV, and hepatitis B/C. The testing was negative. The patient later revealed that he had Kaposi sarcoma a year prior to presentation on his right leg that was removed by his dermatologist. After having multiple thoracenteses, the patient later had a Pleurx catheter placed. Given the patient's other heart comorbidities, he was evaluated by oncology, and they had recommended a more palliative approach.

AIDS-related Lymphoma

- Primary effusion lymphoma (PEL) is one of the least common of the AIDS-related lymphomas, accounting for <1-4% of cases. However this can occur in the absence of HIV infection and rarely has been seen following solid organ transplantation.
- Malignant cells of PEL are monoclonal B cells that express surface CD-38 and contain genomic material from HHV-8
- PEL originates on serosal surfaces including the pleura (60-90%), pericardium (up to 30%), peritoneum, joint spaces and rarely the meninges.
- Most affected patients present with a symptomatic serous effusion containing high grade malignant lymphocytes. Patients usually present with symptoms such as dyspnea (from pleural or pericardial effusion), abdominal distention or joint swelling.

Evaluation and Diagnosis

- **DIAGNOSTIC WORKUP FOR PATIENT**
- *Chest X-Ray:* Pulmonary edema or bilateral multifocal pneumonia with pleural effusions
- *Thoracentesis:* Ultrasound-guided thoracentesis with removal of 1100 mL of blood tinged fluid from the left pleural cavity
- *Cytology:* malignant cells, primary effusion lymphoma.
- *Next step Treatment:* In our particular case the patient and family were offered evaluation by two different oncologists but decision was made not to proceed with chemotherapy due to multiple co-morbidities .

Diagnosis- KS diagnosed with skin biopsy from the center of the lesion.

- *Other diagnostic tools in suspecting KS: Bronchoscopy, Endoscopy, and Colonoscopy. (Bronchoscopy may be indicated in a person in whom you suspect KS who is having respiratory difficulties. EGD and colonoscopies are other tools to use to help diagnose KS located in the GI tract)*
- *Biopsy of skin lesion would be the first initial step to confirm the diagnosis of KS*
- *Fluid Examination: samples of the effusion are almost always positive for malignant cells due to the unique liquid phase of growth of these tumors. The effusions are exudative and often bloody.*

Management

Highly active antiretroviral therapy (HAART) should be initiated immediately in patients who have HIV once Kaposi sarcoma is diagnosed. Other treatment options depends on the individual case. It may include :

- **Radiation** therapy which helps ease the symptoms and cosmetic aspect of the disease.
- **Surgical** removal of small lesions on the skin surface or deeper lesions.
- **Freezing** (cryotherapy) of the skin lesions particularly on the face.
- **Laser** therapy (photocoagulation) to shrink skin lesions and ease pain and bleeding of deeper lesions.
- **Topical** applications such as retinoids to slow tumor growth.
- **Injectable** solutions (chemotherapy and biological therapy) such as anthracyclines, interferons, taxanes and vinca alkaloids.

Conclusion

This was a rather unusual diagnosis in an ailment that typically is associated with HIV. The first line treatment for primary effusion lymphoma is cyclophosphamide, doxorubicin, etoposide, vincristine, and prednisone along with HAART in those who are HIV positive. The therapy is less clear for those who are HIV negative due to being an extremely rare patient population. There has been less than 30 cases described in this subset population and they have typically been elderly, Mediterranean males. The key diagnostic criteria in diagnosing primary effusion lymphoma is the expression of HHV-8 in the malignant cells. Prognosis remains poor despite the use of chemotherapy; median survival is 6 months.

What are the types of Kaposi Sarcoma?

Epidemic Kaposi sarcoma (AIDS-related)

Seen in HIV patients and is an AIDS defining illness.
Most common cancers in HIV positive individuals
However this does not occur in every AIDS patient.

Classic Kaposi sarcoma (Mediterranean)

Seen in elderly Mediterranean, Middle Eastern and European men. HHV-8 infection is more common in these areas therefore, more likely to affect a person with weakened immune system.

Endemic Kaposi sarcoma (African)

It has been noted that not wearing shoes seems to increase the chances of endemic KS possibly due to some lymphatic disorder connected to direct contact with soil.

Immunocompromised Kaposi sarcoma

More likely to occur in an individual with organ transplant and on immune suppression medications.

Photos

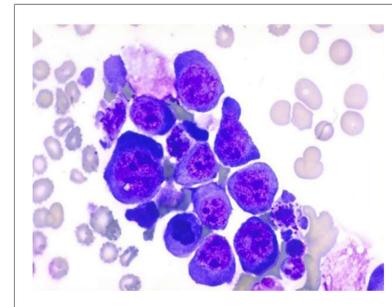


Figure 1. Brown-Red or purple nodules or blotches on the skin slightly raised.
Figure 2: Primary effusion lymphoma, Wright-Giemsa stained smear of pleural fluid.

References

- Dezube BJ. Clinical presentation and natural history of AIDS—Related Kaposi's sarcoma. *Hematol Oncol Clin North Am* 1996; 10:1023
- El Amari EB, Toutous-Trellu L, Gayet-Ageron A, et al. Predicting the evolution of Kaposi Sarcoma, in the highly active antiretroviral therapy era.
- Gao SJ, Kingsley L, Hoover Dr, et al. Seroconversion to antibodies against Kaposi sarcoma-associated herpesvirus-related latent nuclear antigens before development of kaposi sarcoma. *N Engl J Med* 1996; 335:233
- Figure 1- courtesy of Samuel Freire da Silva
- Banks PM, Warnke, RA. Primary effusion lymphoma. In: *World Health Organization Classification of Tumours. Pathology and Genetics of Tumours of Haematopoietic and Lymphoid Tissues*, Jaffe ES, Harris NL, Stein H, Vardiman JW (Eds), IARC Press, Lyon 2001. p.179. Figure 2