CLINICAL VIGNETTE

Uveitis and Hodgkin's Lymphoma

Nimit Sudan, MD

A 23-year-old male was diagnosed with uveitis and retinal vasculitis four years ago. He initially presented to Ophthalmology with headaches and visual changes without obvious or associated etiology for the uveitis and vasculitis. Four months prior he was evaluated by Rheumatology for cold sensitivity of hands and Raynaud's type symptoms. Initial evaluation was negative and observation was suggested. He then developed yellowish red lesions on his lower eyelids which was biopsied. Biopsy revealed mixed acute, chronic lymphohistiocytic and plasmacytic granulomatous inflammatory infiltrate in the dermis, with granulation tissue, hemorrhage, and foreign-body giant cell reaction. After much discussion, his clinical picture was thought to be suggestive of sarcoidosis and the eyelid lesions resolved after 2 weeks. He then developed cough and mild dyspnea and a CT of the chest revealed multi-compartmental bulky intrathoracic lymphadenopathy. This was initially thought to be secondary to sarcoidosis by Pulmonary who concurred that the CT findings were consistent with Sarcoidosis. He began treatment for presumed Sarcoidosis with prednisone and mycophenolate mofetil. Repeat CT Chest after six months revealed an increase in the lymphadenopathy, with the largest lymph node measuring 73 x 49mm, increased from 38 x 26 mm.

CT guided biopsy of mediastinal lymph node mass revealed classic Hodgkin's lymphoma, nodular sclerosis type with Reed-Sternberg cells present. Immunohistochemistry demonstrated CD30, PAX5 (weak), BOB1 (weak), and Fascin positivity. EBV-EBER was negative. A PET-CT revealed bulky confluent multi-station mediastinal lymphadenopathy with the largest para-aortic nodal mass on the left side measuring 74 x 50 mm. A 17 x 14 mm right supraclavicular lymph node was also noted. Mediastinal adenopathy demonstrated mass effect on the adjacent structures with narrowing of the brachiocephalic veins, peripheral superior vena cava and intrathoracic trachea. Intense uptake was also noted in the left vocal cord suggestive of vocal cord paralysis. No FDG lymphadenopathy was noted below the diaphragm. Symptomatically, patient complained of drenching night sweats once or twice a night. In the last 6 months, he had lost over 10 lbs, from his baseline weight of 163lbs. His weight loss had been curbed with edible marijuana use. He also complained of mild to moderate fatigue. In addition, he noticed a transient rash on his thighs, an occasional cough, a hoarse voice for 1-2 months and choking with drinking water. Physical exam was unremarkable. He was a well-developed, healthy appearing male. No lymphadenopathy was appreciated. Lab results revealed a normal sedimentation rate of 3. Hemoglobin was slightly decreased at 13.1 and absolute lymphocyte count low at 0.84. Otherwise, CBC and differential were normal. LDH was normal at 193. CMP was also normal other than a slightly elevated alkaline phosphate of 122.

Per NCCN guidelines, he had Stage IIB disease. He had involvement of 2 regional lymph nodes, mediastinal and supraclavicular, and B symptoms with night sweats. He did not appear to have any unfavorable risk factors. After discussion at tumor conference, given his history of potential Sarcoidosis and pulmonary symptoms, it was suggested to avoid bleomycin and to utilize A-AVD regimen (brentuximab vedotin, doxorubicin, vinblastine, dacarbazine) based on a recent study published with Stage III/IV patients.[1]

He began cycle #1 of A-AVD with treatment complicated by neutropenia. After 2 cycles, PET/CT revealed interval decrease in size of the multi-station mediastinal and right supraclavicular adenopathy, with residual FDG uptake below mediastinal blood pool level, Lugano 1. After completing Cycle #4 follow up PET/CT was consistent with complete remission (CR). He then received adjuvant radiation therapy and remains in CR two years after completing treatment.

Hodgkin's disease (HL) is a malignancy of the lymphoid tissues that accounts for approximately 0.5% of all new cancers and most common in individuals between the ages of 15-34 and over the age of 55.[2] With chemotherapy and/or radiation therapy, it is usually highly curable. Most patients usually present with constitutional symptoms, such as fevers, nights sweats, weight loss, or symptoms secondary to local involvement, such as mass effect. Ocular involvement is rare in HL and when present, these symptoms usually develop after diagnosis.[3,4] Manifestations of ocular involvement have been reported to include chorioretinitis, vitritis, papillary edema, necrotizing retinitis, retinal periphlebitis, infiltration of ocular structures, exudative retinal detachment, retinal hemorrhage and retinal exudates.[3,5] Mechanisms for the symptoms include direct or metastatic lymphomatous involvement, paraneoplastic vasculitis, or iatrogenic complications arising from treatment of HL or immunosuppression.[4-10] Complete remission of ocular inflammation is usually achieved with chemotherapy treatment of HL.[4] Symptoms usually resolve with treatment. Ocular symptoms do not seem to have any prognostic or predictive values for the disease course of HL.

This patient presented with ocular symptoms prior to the diagnosis of HL. His symptoms were initially thought to be secondary to sarcoidosis, however, did not respond to treatment

of sarcoidosis. After diagnosis of HL, symptoms resolved after chemotherapy. The Uveitis and his ocular findings resolved during treatment. Since completing treatment, he has had no further evidence of uveitis and is followed regularly by Ophthalmology.

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