Late-onset Posttransplant Lymphoproliferative Disorder after Simultaneous Pancreatic and Kidney Transplantation: Rare and Fatal

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**Background**

- Post-transplant Lymphoproliferative Disease (PTLD) is a rare and life-threatening complication of solid organ transplantation (SOT) or hematopoietic stem cell transplantation (HSCT).
- Often associated with intensive immunosuppression and multiple organ transplantation, it is usually seen within the first-year post-procedure, although rarely, it can occur as long as 10 years post-transplantation.

**Case Presentation**

- A 38-year-old African American woman presented with left lower extremity jerking movement and right frontoparietal headache.
- She has a history of Type 1 DM requiring simultaneous pancreatic and left kidney transplant in 2008 which was complicated by a failed kidney transplant in 2013 and is presently on hemodialysis.
- She has been on prednisone, tacrolimus, and mycophenolate mofetil (MMF).
- On presentation, her vital signs were stable with oxygen saturation of 97% on ambient air. She was alert and oriented. Neurological exam was significant for low amplitude jerking movement of the left lower extremity with normal muscle strength and sensation.

**Diagnostic Workup and Treatment**

- Electroencephalography was abnormal with intermittent sharp slowing left more than the right temporal lobe and occasional sharp without seizure activity.
- A right frontal, stereotactic assisted open brain biopsy of the right frontal mass was done. Pathology showed histopathology and immunohistochemistry showing an EBV-positive, monomorphic post-transplant lymphoproliferative disorder (Diffuse Large B-Cell Lymphoma) with positive CD20, CD3, CD10, BCL2, BCL6, Mum 1 and Ki-67 of 80% on special staining. (Figure 2)
- She was started on dexamethasone intravenously and was continued on tacrolimus while MMF was held. She was then referred to another tertiary institution service for initiation of chemotherapy, however, later succumbed to her illness.

**Discussion**

- The highest risk for PTLD among SOT are heart, lung, intestinal and multi-organ transplant subjects while liver and kidney transplant recipients have a lower risk at about 1-5%.
- Like our patient with diffuse large B cell lymphoma, CNS involvement usually presents as a space occupying lesion manifesting as headaches, seizures or focal neurological deficits. It occurs at a rate of about 10-15% with the highest incidence seen in pancreas and kidney transplant recipients.

**References**