Oncology Case Reports Journal

Clinical Image
Published: 24 Apr, 2018

Massive Pancreatic Cysts in a Patient with Von Hippel Lindau Syndrome

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Clinical Image

40-year-old male with a history of Von Hippel-Lindau (VHL) syndrome presented to his primary care physician's office with intermittent nausea, vomiting and abdominal pain. Persistent symptoms prompted a Computed Tomography (CT scan) of the abdomen revealing a large conglomerate of pancreatic cysts. VHL is a rare inheritable cancer syndrome resulting from a germline mutation in the VHL gene that predisposes patients to develop cerebellar hemangioblastomas, retinal angiomas, endolymphatic sac tumors, renal cell carcinoma, pancreatic neuroendocrine tumors, renal cysts and pancreatic cysts. Pancreatic cysts are particularly common in VHL patients with one series describing up to 71% of patients having [1]. Although pancreatic neuroendocrine tumors are often found co-existent with pancreatic cysts in patients with VHL, there is no conclusive evidence that simple pancreatic cysts have the potential for malignant transformation in VHL patients. In addition, massive pancreatic cysts are rarely associated with symptomatic sequelae and surgery is rarely recommended [2]. This patient's symptoms resolved spontaneously and no surgical intervention was needed. Figure 1 shows massive pancreatic cysts in a patient with Von Hippel Lindau Syndrome.

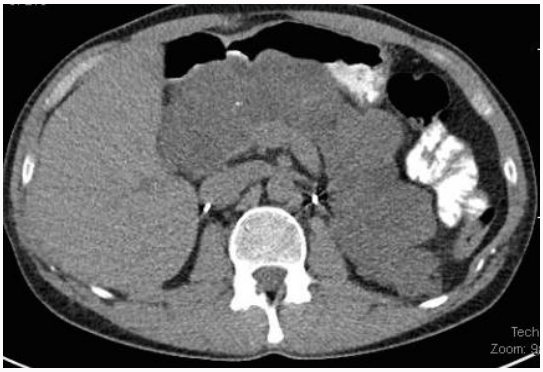

Figure 1: Massive pancreatic cysts in a patient with Von Hippel Lindau Syndrome.

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OPEN ACCESS

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Received Date: *13 Apr 2018*
Accepted Date: *20 Apr 2018*
Published Date: *24 Apr 2018*

Citation:
King GT. Massive Pancreatic Cysts in a Patient with Von Hippel Lindau Syndrome. Oncol Case Report J. 2018; 1(1): 1001.