A rare case of Castleman Disease in a young male

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Introduction:

- Castleman disease is a rare disorder of lymphoid tissue with presentation ranging from inflammatory to low grade neoplasm. It is important to recognize this condition due to its variability in presentation and treatment choices. We present a rare case of castleman disease in a young male patient.

Case Presentation:

- 34-year-old male with past medical history of saddle pulmonary embolus s/p thrombectomy under Eliquis twice daily, right leg DVT with factor V Leiden mutation presented with upper abdominal pain, nausea and vomiting. Lab workup was negative. Imaging positive for left upper quadrant retroperitoneal mass anteriorly displacing the left pancreas with no invasion. Underwent enbloc resection of the mass with distal pancreas and spleen. Histopathological examination of the specimen revealed hyaline vascular type of angiofollicular lymphoid hyperplasia, with negative margins and no disease was found in a lymph node and omentum. Confirmatory for unicentric Castleman disease. Workup later for HHV-8 and HIV were negative. The patient is being followed for 5 years, with imaging once a year.

Discussion

•Castleman disease(CD) also angiofollicular lymph node hyperplasia describes a group of lymphoproliferative disorders that share common histopathologic features. Unicentric CD involves 1 or more enlarged lymph node in a single region of the body, Multicentric CD(MCD) involves multiple regions of lymphnodes. MCD is further subclassified according to the presence of HHV 8.

•CAD is associated with non-Hodgkin's lymphoma, Hodgkin's lymphoma and POEMS syndrome(polyneuropathy, organomegaly, endocrinopathy, monoclonal protein, skin changes). Evaluation for associated conditions and ruling out underlying malignancies with appropriate follow up is the important post diagnosis and treatment. The exact cause of the condition is unknown; however, the most likely underlying pathogenesis is hypersensitivity or reactive changes to normal antigens in the body, with excess release of pro inflammatory cytokines including IL-6.

•Surgical resection is curative in most of the cases, asymptomatic and unresectable disease is followed up with Observation. Inflammation related symptoms are managed with novel anti-inflammatory agents (eg: anti- IL 6 therapy with siltuximab and tocilizumab). Compressive masses are managed by resection, rituximab and embolisation to decrease the size and symptom severity.

Reference

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