E39

A case of IgG4-related sclerosing disease masquerading as pancreatic cancer

Sungho Jo[1], Sanghyun Song[1], and Wonae Lee[2]

[1]Department of Surgery, Dankook University College of Medicine, Korea
[2]Department of Pathology, Dankook University College of Medicine, Korea

Purpose:

IgG4-related sclerosing disease (IgG4-RSD) is a new and emerging disease entity, which is characterized by fibrosis, extensive infiltration of IgG4-positive plasma cells, and elevation of serum IgG4 level. This disorder mainly targets organs, including the pancreas, bile ducts, salivary glands, lacrimal glands, lymph nodes, retroperitoneum, aorta, lung, breast, and kidneys. We herein report a very rare case of IgG4-RSD involving pancreas and peripancreatic soft tissue which was misdiagnosed as pancreatic cancer.

Methods:

A 69-year-old male was admitted for interval increase in extent of ill-defined peripancreatic fatty infiltration around pancreas tail under suspicion of malignancy. He had undergone transanal excision for villous adenoma 7 years before. EUS-guided FNA carried out 3 months before revealed no malignant cells. Laboratory values at admission including CA 19-9 were within normal limit. Serum IgG4 level was not checked. Subsequently performed PET-CT showed ill-defined hypermetabolic lesions (SULmax 3.8) at and around pancreatic tail portion. With the impression of pancreatic cancer, we performed posterior radical antegrade modular pancreatosplenectomy (RAMPS). An about 15mm-sized hard mass was palpable at border of pancreatic body and tail portion and severe peripancreatic fatty infiltration was encountered. The hard infiltrating mass was adherent to the left kidney without true invasion. A few enlarged regional lymph nodes were also found.

Results:

Histopathological examination of the surgical specimen demonstrated IgG4-RSD with marked involvement of peripancreatic, perisplenic, and periadrenal soft tissue and partial spread into pancreatic parenchyma. All lymph nodes were proved to be reactive hyperplasia. Immunohistochemical staining for IgG4 showed increased positivity. The patient was discharged on 14th postoperative date without any complications.

Conclusion:

IgG4-RSD masquerading as pancreatic cancer is an extremely rare and confusing disease. Though surgical resection could be performed for this disease, high index of suspicion and multi-disciplinary workup are absolute to avoid unnecessary major surgery.