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203.LYMPHOCYTES AND ACQUIRED OR CONGENITAL IMMUNODEFICIENCY DISORDERS

A Curious Case of IgG4-Related Disease Masquerading As an Occult Abdominal Malignancy

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Introduction

IgG4-related disease is an autoimmune, fibroinflammatory disorder involving multiple organ systems in the body. Diagnosis is often suggested by strong IgG4-positive stain on tissue biopsy. Patients respond well to steroids; and in cases of relapse, steroid-sparing agents like rituximab, azathioprine or cyclosporine can be used.

Case Presentation

Here we present the case of a 77 year-old African-American female with a past medical history significant for cerebrovascular accident, chronic hepatitis C infection status post treatment with antivirals, left chronic dacryoadenitis and chronic iron-deficiency anemia. She presented to the hematology office for a follow-up visit for chronic anemia and complained of new onset right upper quadrant pain and unexplained weight loss. Subsequent abdominal and pelvic imaging showed hypoechoic lesion in pancreatic head along with multiple epigastric, omental and gastro-hepatic nodules/lymph nodes suspicious for malignancy. This was followed by esophagogastroduodenoscopy (EGD) with endoscopic ultrasound (EUS) which revealed gastric body/antral erosions along with multiple enlarged abdominal lymph nodes and heterogeneous, hypoechoic irregular mass in the pancreatic head/neck, all of which were biopsied. While awaiting biopsy results, due to high suspicion for malignancy, PET-FDG was performed which showed hypermetabolic lesions corresponding to the EUS findings and a FDG-avid right paravertebral mid thoracic soft tissue mass favored to represent a neurogenic tumor, unrelated to abdominal disease. Laboratory work-up was significant for elevated IgG levels with particularly high titres of IgG subclass 4. Biopsy results from EUS showed no evidence of malignant cells, no clonal proliferation on flow cytometry in either the lymph nodes or pancreatic tissue. Cytology of pancreatic tissue biopsy revealed lymphoid infiltration, extensive edema and fibrosis with no malignant cells. Immunohistochemistry studies revealed strong positive stain for IgG4 suggestive of IgG4-related disease. Of note, the patient had been following up with ophthalmology for a few years for chronic inflammatory dacryoadenitis of the left eye, which can likely be attributed to IgG4-related disease. After shared-decision making with the patient regarding treatment options, treatment was initiated with a plan of rituximab for 4 cycles along with corticosteroid. As of this abstract submission, the patient received one dose of rituximab, pending assessment of response to treatment.

Discussion

Heterogeneous pancreatic masses on abdominal imaging should be considered malignant until otherwise proven. In cases where these masses are associated with diffuse abdominal and pelvic lymphadenopathy, definitive diagnosis is made through obtaining biopsies from the easily accessible sites. A benign, auto-immune, fibroinflammatory condition which mimics this presentation is IgG4-related disease, whose treatment is far less complicated than a metastatic carcinoma. It often involves lacrimal glands. A high index of suspicion backed by the biopsy results showing strong stain affinity to IgG4 and elevated blood IgG4 levels are required. Treatment involves initiation of steroids followed by steroid-sparing agents like azathioprine, cyclosporine or rituximab in cases of relapse after steroid induction. However, as in our case, combination therapy of simultaneous use of steroids with rituximab can be used after shared decision making with patients.

Disclosures No relevant conflicts of interest to declare.

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