EDORIUM Journals
www.edoriumjournals.com

CASE REPORT PEER REVIEWED | OPEN ACCESS

Unmasking IgG4-related autoimmune pancreatitis from pancreatic cancer: A lesson learned

Ashwad Afzal, Seema Chittalae, Ivan Wong, Petros Efthimiou

ABSTRACT

Introduction: Autoimmune pancreatitis (AIP) is difficult to distinguish from pancreatic cancer. An AIP has a similar clinical presentation and is one component of a systemic disease, Immunoglobulin G4 (IgG4) related sclerosing disease. IgG4 related sclerosing disease is characterized by extensive IgG4 positive plasma cells and T-lymphocytes that may involve the pancreas and other organs.
Case Report: A 77-year-old male presented with nausea, vomiting, decreased appetite, and 39 lbs weight loss over three months. An endoscopic ultrasound showed one 3.3x2.7 cm mass on the tail of the pancreas extending towards the splenic hilum with no lymph node involvement. A fine needle aspiration was indeterminate. The patient underwent distal pancreatectomy and splenectomy for presumed pancreatic cancer, however, the pathology report was negative for malignancy. Instead, the microscopic examination revealed dense lymphoplasmacytic infiltrate staining positive for IgG4, onion-skin pattern fibrosis, and lobular atrophy in the medium sized pancreatic duct that was consistent with AIP.
Conclusion: IgG4 related sclerosing disease is a systemic disorder that may involve multiple organs. The clinical manifestations are similar to that of pancreatic malignancy. For this reason, diagnostic evaluation to differentiate the two disease entities is crucial, as treatments differ significantly. Treatment for AIP consists of early initiation with glucocorticoid therapy. However, poor response to treatment usually indicates advanced fibrotic changes or an alternative diagnosis. Failure to differentiate autoimmune pancreatitis from pancreatic cancer can lead to unnecessary surgery of the pancreas along with complications associated with surgery.

CASE REPORT PEER REVIEWED | OPEN ACCESS

Unmasking IgG4-related autoimmune pancreatitis from pancreatic cancer: A lesson learned

Ashwad Afzal, Seema Chittalae, Ivan Wong, Petros Efthimiou

ABSTRACT

Introduction: Autoimmune pancreatitis (AIP) is difficult to distinguish from pancreatic cancer. An AIP has a similar clinical presentation and is one component of a systemic disease, Immunoglobulin G4 (IgG4) related sclerosing disease. IgG4 related sclerosing disease is characterized by extensive IgG4 positive plasma cells and T-lymphocytes that may involve the pancreas and other organs. Case Report: A 77-year-old male presented with nausea, vomiting, decreased appetite, and 39 lbs weight loss over three months. An endoscopic ultrasound showed one 3.3x2.7 cm mass on the tail of the pancreas extending towards the splenic hilum with no lymph node involvement. A fine needle aspiration was indeterminate. The patient underwent distal pancreatectomy and splenectomy for presumed pancreatic cancer, however, the pathology report was negative for malignancy. Instead, the microscopic examination revealed dense lymphoplasmacytic infiltrate staining positive for IgG4, onion-skin pattern fibrosis, and lobular atrophy in the medium sized pancreatic duct that was consistent with AIP. Conclusion: IgG4 related sclerosing disease is a systemic disorder that may involve multiple organs. The clinical manifestations are similar to that of pancreatic malignancy. For this reason, diagnostic evaluation to differentiate the two disease entities is crucial, as treatments differ significantly. Treatment for AIP consists of early initiation with glucocorticoid therapy. However, poor response to treatment usually indicates advanced fibrotic changes or an alternative diagnosis. Failure to differentiate autoimmune pancreatitis from pancreatic cancer can lead to unnecessary surgery of the pancreas along with complications associated with surgery.

Keywords: Autoimmune pancreatitis, IgG4 related sclerosing disease, Pancreatic cancer

How to cite this article

Afzal A, Chittalae S, Wong I, Efthimiou P. Unmasking IgG4-related autoimmune pancreatitis from pancreatic cancer: A lesson learned. Int J Case Rep Images 2016;7(2):123–126.

doi:10.5348/ijcri-201620-CR-10607

Ashwad Afzal[1], Seema Chittalae[1], Ivan Wong[1], Petros Efthimiou[1]

Affiliations: [1]MD, Department of Internal Medicine, New York Methodist Hospital affiliate of Weill Medical College of Cornell University, Brooklyn, New York, USA.

Corresponding Author: Ashwad Afzal, New York Methodist Hospital, Affiliate of Weill Medical College of Cornell University506 6th street, Brooklyn, NY 11215, USA; Email: ashwad.Afzal@gmail.com

Received: 13 October 2015
Accepted: 05 November 2015
Published: 01 February 2016

INTRODUCTION

Autoimmune pancreatitis (AIP) is difficult to distinguish from pancreatic cancer. They both share similar clinical presentation including an insidious with anorexia, nausea, vomiting, decreased appetite, epigastric pain, and significant weight loss. The AIP also has a similar clinical presentation and is one component of a systemic disease, immunoglobulin G4 (IgG4) related sclerosing disease. IgG4 related sclerosing disease is characterized by extensive IgG4 positive plasma cells and T lymphocytes that may involve the pancreas, bile duct,

retroperitoneum, salivary glands and other organs. We illustrate a case of autoimmune pancreatitis presenting as pancreatic cancer.

CASE REPORT

A 77-year-old legally blind male with a past medical history of hypertension, hyperlipidemia, coronary artery disease with stents, and diabetes mellitus type 2 (established for 25 years) presented with nausea, vomiting, decreased appetite, and 39 lbs weight loss over a 3-month period. He was an ex-smoker with a 30-pack-year history. There was no significant surgical or family history. His vital signs were stable and his body mass index was 24.91 kg/m^2. The pertinent physical examination revealed mild tenderness to palpation over the epigastric area with no scleral icterus, and no lymphadenopathy. Routine blood work including complete blood count, renal and liver function tests were normal. An endoscopic ultrasound showed a 3.3x2.7 cm mass on the tail of the pancreas extending towards the splenic hilum with no lymph node involvement (Figure 1). A fine needle aspiration was indeterminate. The patient underwent distal pancreatectomy and splenectomy for high suspicion of pancreatic malignancy, however, the surgical pathology report was negative for malignancy. Microscopic examination revealed dense lymphoplasmacytic infiltrate with onion-skin pattern fibrosis and lobular atrophy in medium sized pancreatic ducts that was consistent with lymphoplasmacytic sclerosing pancreatitis (Figure 2). Further immunohistochemical staining showed 30–40 IgG4 positive plasma cells on high power field and labs revealed an elevated IgG4 level of 272.5 mg/dl, consistent with the diagnosis of AIP (Figure 3).

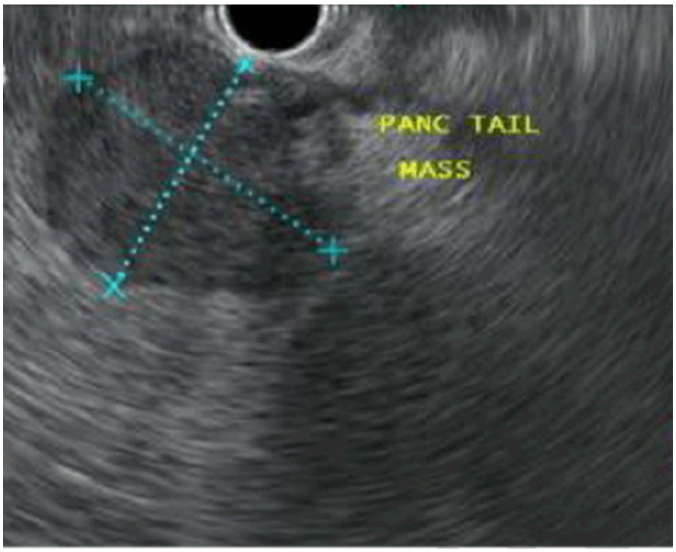

Figure 1: Endoscopic Ultrasound showing 3.3x2.7 cm hypoechoic mass on the tail of the pancreas extending towards the splenic hilum.

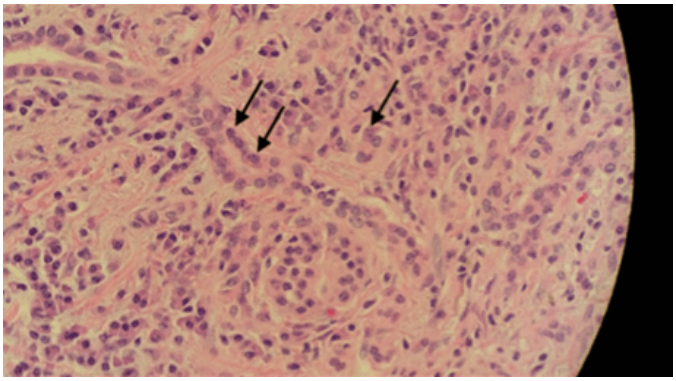

Figure 2: Microscopic Examination revealed dense lymphoplasmacytic infiltrate.

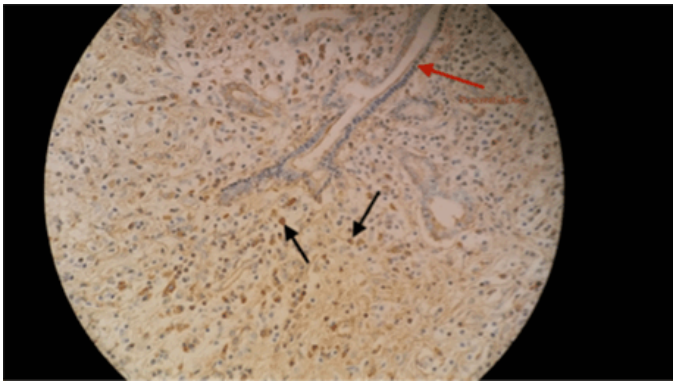

Figure 3: Immunohistochemical staining shows an abundance of IgG4 positive plasma cells (black arrow) and pancreatic duct (red arrow) surrounded by plasma cells.

DISCUSSION

IgG4 related sclerosing disease is a systemic disorder that may involve multiple organs. The clinical manifestations may include anorexia, weight loss, nausea, and painless jaundice similar to that of pancreatic malignancy. For this reason, diagnostic evaluation to differentiate the two disease entities is crucial, as treatment differ significantly. In AIP, serum IgG4 levels are elevated, and levels >2 times the upper limit of normal is highly suggestive of the disease [1]. However, this may also be seen in pancreatic malignancy and thus cannot be used to differentiate the two [2]. Type 1 AIP (lymphoplasmacytic sclerosing pancreatitis) has a greater prevalence of having elevated serum IgG4 levels and a higher relapse rate compared to Type 2 AIP (idiopathic duct centric pancreatitis) [3]. There are various imaging modalities which can be used to evaluate the pancreas including computed tomography, or magnetic resonance imaging identifying a diffusely enlarged pancreas resembling a “sausage-like” appearance or a focal mass, such as in this case. Endoscopic ultrasound may reveal diffuse hypoechoic pancreatic enlargement and bile duct wall thickening. A core needle biopsy is often required for histological evaluation as opposed to a fine needle

aspiration, which may be inadequate, as was portrayed in this case. The presence of lymphoplasmacytic tissue infiltration characterized by extensive IgG4 positive plasma cells and T lymphocytes deposition, fibrosis, and obliterative phlebitis on histology is diagnostic for AIP [4, 5]. Response to steroid therapy also supports the diagnosis [6]. Treatment consists of early initiation with glucocorticoid therapy [7]. A study by Matsubayashi et al. showed steroid response in 86% of patients within two weeks and in 97% of patients within four weeks by demonstrating a decrease in tumor size on abdominal ultrasound [6]. Steroid response was seen in almost 98% of patients with AIP in a study by Kamisawa et al. [8]. For resistant cases, B cell depleting agents such as Rituximab has been used with success [9]. A poor response to treatment usually indicates either advanced fibrotic changes or an alternative diagnosis.

CONCLUSION

Autoimmune pancreatitis (AIP) is a difficult diagnosis to differentiate from pancreatic cancer, as the presentation is very similar. Clinical suspicion with laboratory markers and imaging may help aid in the diagnosis. However, laboratory markers may be normal and a biopsy is required to make the diagnosis. Failure to differentiate AIP from pancreatic cancer can lead to unnecessary resection of the pancreas along with complications of surgery, as autoimmune pancreatitis responds well to steroid therapy. We recommend patients with pancreatic mass be evaluated for AIP to avoid invasive treatment.

****

Acknowledgements
We are thankful to Arpita Bose and the library staff at New York Methodist hospital for their dedication in helping residents with research.

Author Contributions
Ashwad Afzal – Substantial contribution to conception and design, Acquisition of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published
Seema Chittalae – Substantial contribution to conception and design, Acquisition of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published
Ivan Wong – Substantial contribution to conception and design, Acquisition of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published.
Petros Efthimiou – Substantial contribution to conception and design, Acquisition of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published.

Guarantor
The corresponding author is the guarantor of submission.

Conflict of Interest
Authors declare no conflict of interest.

REFERENCES

1. Ghazale A, Chari ST, Smyrk TC, et al. Value of serum IgG4 in the diagnosis of autoimmune pancreatitis and in distinguishing it from pancreatic cancer. Am J Gastroenterol 2007 Aug;102(8):1646–53.
2. Chari ST, Takahashi N, Levy MJ, et al. A diagnostic strategy to distinguish autoimmune pancreatitis from pancreatic cancer. Clin Gastroenterol Hepatol 2009 Oct;7(10):1097–103.
3. Sah RP, Chari ST, Pannala R, et al. Differences in clinical profile and relapse rate of type 1 versus type 2 autoimmune pancreatitis. Gastroenterology 2010 Jul;139(1):140–8.
4. Deheragoda MG, Church NI, Rodriguez-Justo M, et al. The use of immunoglobulin g4 immunostaining in diagnosing pancreatic and extrapancreatic involvement in autoimmune pancreatitis. Clin Gastroenterol Hepatol 2007 Oct;5(10):1229–34.
5. Deshpande V, Chicano S, Finkelberg D, et al. Autoimmune pancreatitis: a systemic immune complex mediated disease. Am J Surg Pathol 2006 Dec;30(12):1537–45.
6. Matsubayashi H, Yoneyama M, Nanri K, et al. Determination of steroid response by abdominal ultrasound in cases with autoimmune pancreatitis. Dig Liver Dis 2013 Dec;45(12):1034–40.
7. Hirano K, Tada M, Isayama H, et al. Long-term prognosis of autoimmune pancreatitis with and without corticosteroid treatment. Gut 2007 Dec;56(12):1719–24.
8. Kamisawa T, Shimosegawa T, Okazaki K, et al. Standard steroid treatment for autoimmune pancreatitis. Gut 2009 Nov;58(11):1504–7.
9. Khosroshahi A, Bloch DB, Deshpande V, Stone JH. Rituximab therapy leads to rapid decline of serum IgG4 levels and prompt clinical improvement in IgG4-related systemic disease. Arthritis Rheum 2010 Jun;62(6):1755–62.

Edorium Journals: An introduction

Edorium Journals Team

About Edorium Journals

Edorium Journals is a publisher of high-quality, open access, international scholarly journals covering subjects in basic sciences and clinical specialties and subspecialties.

Invitation for article submission

We sincerely invite you to submit your valuable research for publication to Edorium Journals.

But why should you publish with Edorium Journals?

In less than 10 words - we give you what no one does.

Vision of being the best

We have the vision of making our journals the best and the most authoritative journals in their respective specialties. We are working towards this goal every day of every week of every month of every year.

Exceptional services

We care for you, your work and your time. Our efficient, personalized and courteous services are a testimony to this.

Editorial Review

All manuscripts submitted to Edorium Journals undergo pre-processing review, first editorial review, peer review, second editorial review and finally third editorial review.

Peer Review

All manuscripts submitted to Edorium Journals undergo anonymous, double-blind, external peer review.

Early View version

Early View version of your manuscript will be published in the journal within 72 hours of final acceptance.

Manuscript status

From submission to publication of your article you will get regular updates (minimum six times) about status of your manuscripts directly in your email.

Our Commitment

Six weeks

You will get first decision on your manuscript within six weeks (42 days) of submission. If we fail to honor this by even one day, we will publish your manuscript free of charge.

Four weeks

After we receive page proofs, your manuscript will be published in the journal within four weeks (31 days). If we fail to honor this by even one day, we will publish your manuscript free of charge and refund you the full article publication charges you paid for your manuscript.

Most Favored Author program

Join this program and publish any number of articles free of charge for one to five years.

Favored Author program

One email is all it takes to become our favored author. You will not only get fee waivers but also get information and insights about scholarly publishing.

Institutional Membership program

Join our Institutional Memberships program and help scholars from your institute make their research accessible to all and save thousands of dollars in fees make their research accessible to all.

Our presence

We have some of the best designed publication formats. Our websites are very user friendly and enable you to do your work very easily with no hassle.

Something more...

We request you to have a look at our website to know more about us and our services.

We welcome you to interact with us, share with us, join us and of course publish with us.

Edorium Journals: On Web

Browse Journals

CONNECT WITH US