Pictorial Review of Thoracoabdominal Manifestations of IgG4-Related Diseases

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Abstract

Immunoglobulin G4-related diseases (rD-IgG4) are a group of entities that share common clinical, serological and pathological features. They were thought to be individual diseases, but with advances in diagnosis they have been found to be part of the same spectrum (and those yet to be described).

Their main characteristic is the infiltration of tissues by IgG4+ plasma cells and lymphocytes, with or without the presence of fibrosis.

This entity was first described in pancreas (known as IgG4-related autoimmune pancreatitis, IgG4-rAIP), but it can affect practically the whole organism.

Definitive diagnosis can be difficult, especially early in the course of the disease. For this reason, multiple diagnostic criteria have been developed, differing according to the different working groups, which are intended to assist the clinician in its suspicion and diagnosis.

We will focus on developing the radiological features at thorax and abdomen, that may lead us to suspect this entity. From the point of view of Pathological Anatomy, it is important to consider the suspicion, because immunohistochemical tests are needed to reach the diagnosis.

On the other hand, it is also important to bear in mind that this pathology can simulate tumors, infections, or other autoimmune diseases, so radiologists should be careful when making differential diagnosis.

Its treatment is based on the use of oral corticosteroids, with the possibility of resorting to immunomodulators or monoclonal antibodies in refractory cases, relapses or in patients with multisystemic involvement.

Image

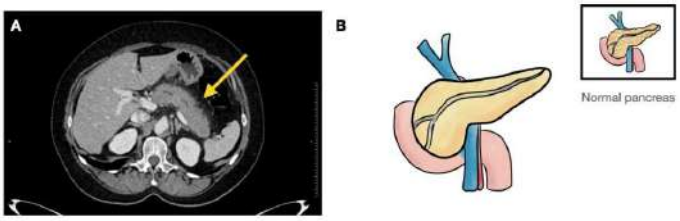


Figure 1: A) Axial CT of the abdomen with intravenous contrast in portal phase. IgG4-rAIP (diffuse pattern) showing loss of pancreatic lobulation (yellow arrow). B) Representative drawing of the described findings. Drawing of normal pancreas to emphasize the differences.

Recent Publications (minimum 5)

1. Álvarez-García M, Álvarez Torices JC, Díez Liébana MJ, de la Fuente Olmos MR. Manifestaciones toracoabdominales de las enfermedades relacionadas con la inmunoglobulina G4. Radiología. 2023;65(2):165-175. DOI: 10.1016/j.rx.2022.06.002
2. Martínez-de-Alegría A, Baleato-González S, García-Figueiras R, Bermúdez-Naveira A, Abdulkader-Nallib I, Díaz-Peromingo JA, et al. IgG4-related disease from head to toe. Radiographics. 2015;35:2007-25..
3. Stone JH, Khosroshahi A, Deshpande V, Chan JK, Heathcote JG, Aalberse R, et al. Recommendations for the nomenclature of IgG4-related disease and its individual organ system manifestations. Arthritis Rheum. 2012;64:3061-7.
4. Umehara H, Okazaki K, Masaki Y, Kawano M, Yamamoto M, Saeki T, et al. Comprehensive diagnostic criteria for IgG4-related disease (IgG4-RD), 2011. Mod Rheumatol. 2012;22:21-30.
5. Hubers L, Maillette de Buy L, Doorenspleet M, Klarenbeek P, Verheji J, Rauws EA. IgG4-associated cholangitis: A comprehensive review. Clin Rev Allergy Immunol. 2015;48:198-206.

Photograph



Biography

Dr was born in León, Spain. She studied medicine at the University of Salamanca (Spain). After taking the entrance exams to the medical residency, she specialized in Radiology at the Río Hortega University Hospital (Valladolid, Spain) for 4 years, within one year learning in external rotations at different hospitals in Madrid and León. She has also published her first article as first author and has completed the master's degree in Innovation in Biomedical and Health Sciences at the University of León. She is currently completing a University Expert Course in Head, Neck and Locomotor System Ultrasound and a master's degree in Updating Diagnostic and Therapeutic Techniques in Radiology (both from the CEU-Cardenal Carrera University).

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Notes/Comments:

I declare that I have no conflict of interest.