PANCREATIC NEUROENDOCRINE TUMOR MIMICKING IMMUNOGLOBULIN G4- RELATED DISEASE (IgG4-RD)

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Case-Report

A 22-year-old patient, (PLAM), presented with episodes of fever and lymphadenopathy in the left supraclavicular region, confirmed by ultrasound (USG). The patient began treatment with antibiotics (amoxicillin + clavulanic acid and later levofloxacin) and nonsteroidal anti-inflammatory drugs (NSAIDs) while undergoing etiological investigation. Laboratory tests showed mild leukocytosis, elevated inflammatory markers, and negative serologies for hepatitis B, C, HIV, and syphilis, as well as negative tests for CMV, Epstein-Barr virus, and dengue. Biochemistry and renal and hepatic function were normal. Cervical USG and chest CT did not reveal significant abnormalities. While awaiting lymph node biopsy, the patient developed dyspepsia with epigastric pain.

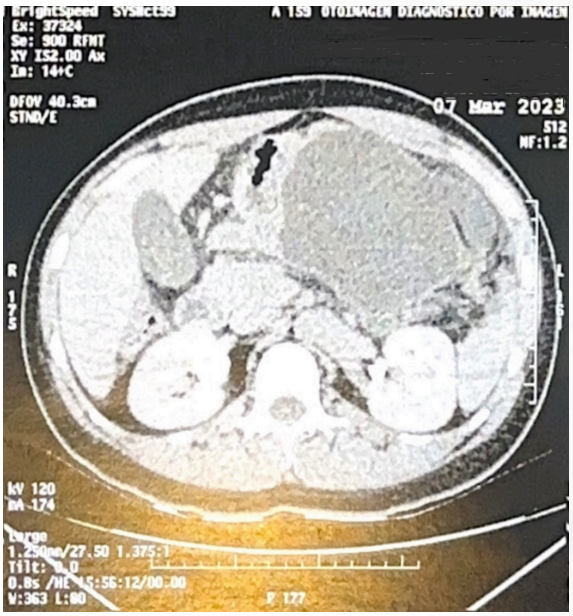


Figure 1. Neoplastic-appearing mass in the pancreatic body

New tests, including an abdominal USG, showed an image compatible with pancreatitis, with no apparent cause, and elevated amylase (428, normal <76) and lipase (2709, normal <39) levels. GGT was 21, ALT was 17, total bilirubin was 0.09, direct bilirubin was 0.3, and indirect bilirubin was 0.091. IgG4 was 6270 (normal up to 2010). A follow-up USG a week later revealed an anechoic fluid image in the left hypochondrium measuring 4.7 x 2.1 x 3.9 cm (estimated volume of 20.6 ml) adjacent to the stomach. Subsequently, a total abdominal CT showed a large peripancreatic cystic formation measuring 19 x 10 x 10 cm with an approximate volume of 988 ml, associated with focal dilation of the Wirsung duct in the body and tail of the pancreas. Complementary total abdominal MRI revealed a neoplastic-appearing mass in the pancreatic body associated with nonspecific globular lymphadenopathy in the splenic hilum (*Figure 1)*. Approximately three months after the onset of symptoms, the patient underwent partial pancreatectomy, with histopathological results showing a well-differentiated neuroendocrine tumor confined to the pancreas, with clear surgical margins. The patient showed clinical improvement and symptom remission.

Conclusion: Although the diagnosis of IgG4-RD can be made without a biopsy, in a subset of patients, even when the clinical picture is suggestive and IgG4 is elevated, a biopsy is generally necessary to elucidate and confirm the diagnosis in the presence of tumor-like lesions and/or lymphadenopathy.

Keywords: pancreatic neuroendocrine tumor; immunoglobulin; disease; igg4-rd.