

# IgG4-related Diseases – A Rare Polycystic Form of Ormond’s Disease

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**Abstract:** Currently, Ormond’s disease is classified among IgG4-associated diseases. Its clinical manifestation varies and is characterized by the presence of fibrous retroperitoneal tissue that often affects the ureters or abdominal aorta and iliac arteries. We present a unique case of the polycystic form of Ormond’s disease, imitating tumour in the retroperitoneal space. At the time of diagnosis, the disease was not metabolically active and did not require immunosuppressive therapy. The polycystic mass was removed surgically. There has been no exacerbation of the disease during the last 12 months.

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## Introduction

IgG4-related diseases comprise a relatively newly defined group of diseases characterized by IgG4 hypergammaglobulinaemia with the presence of IgG4-producing plasma cells in affected organs with fibrotic or sclerotizing changes (Kamisawa et al., 2015). A disease associated with IgG4 may affect virtually any organ – salivary glands, periorbital tissue, kidneys, lungs, meninges, aorta, prostate, pericardium or skin. Histopathological findings are uniform, characterized by a major lymphoplasmocytic infiltrate and the presence of IgG4-producing plasma cells, irrespective of the affected site. Ormond's disease (idiopathic retroperitoneal fibrosis) is an example of a disease associated with IgG4. It is characterized by chronic periaortitis and retroperitoneal fibrosis. The inflammatory process affects the infrarenal part of the abdominal aorta and the iliac arteries and is characterized by the presence of infiltrates encasing the ureters and inferior vena cava.

## Case report

A 46-year-old male patient presented to the Department of Urology, Na Homolce Hospital, for backache and fatigue. Clinical and ultrasound assessments and a CT (computed tomography) examination were performed, which revealed an obstructed ureter on the left-hand side. The CT scan showed a mass of 7×5×7 cm that was obstructing the left ureter and iliac arteries. The patient was referred for a surgical procedure to be performed in the Department of Vascular Surgery. Upon admission, the following data were collected. Family history: his mother died at the age of 68 following Grawitz tumour surgery; his father died at the age of 70 from a laryngeal tumour. The patient had been treated for hypertension. Health state: heart rate 97 beats/min, BP (blood pressure) 150/100 mm Hg, respiratory rate 16/min, temperature 36.5 °C, BMI (body mass index) 31.0, eupnoeic, oriented; no jaundice, cyanosis or anaemia; adequate hydration and skin turgor. Head was painless; ears and nose showed no pathological finding. Thyroid gland was not enlarged; cervical veins had an adequate filling; carotid arteries had a symmetric beat, with no murmur. The patient had a regular heartbeat, with two distinct sounds; the heart was not enlarged on percussion; breathing was alveolar, with no additional phenomena. The abdomen was painless on palpation; a resistant mass was palpable in the left mesogastrium; liver and spleen were not enlarged; tapotement was bilaterally negative. Extremities showed no signs of inflammation; pulse on peripheral arteries on both the upper and lower extremities was well palpable all the way into the periphery; joints showed no signs of inflammation and had a regular mobility. A preoperative assessment was performed that did not reveal any contraindications for the surgical treatment. Laboratory findings: serum minerals – Na, K, Cl, urea, creatinine, glycaemia, AST, ALT, cholesterol, triglycerides, all with no pathology, C-reactive protein 9 mg/l, Hb 113 g/l, erythrocytes  $3.74 \times 10^{12}/l$ , haematocrit 0.34 l/l, leukocytes  $9.6 \times 10^9/l$ , thrombocytes  $636 \times 10^9/l$ , lues, HbSAg, HIV negative, FW 28/52, cardiac and lung X-ray showed no

pathological finding. An 8 cm large tumour mass affixed to the retroperitoneum, well vascularized on the surface, was removed during the vascular surgical procedure. The ureter was encased into the tumour wall. The lower part of the tumour had close contact with the fascia of the psoas muscle and was affixed to the anterior vertebral fascia. The inferior mesenteric artery was encased by the anterior part of the tumour. Similarly, the tumour was growing behind the aorta, deviating it vertebrolaterally to the right. The tumour involved the inferior vena cava. A complete removal of the tumour was performed. The following histopathological examination showed. Macroscopic finding: a partially emptied cystic cavity (70×60×55 mm) with serous contents and a 1–10 mm thick wall made of homogenous grey tissue. Microscopic finding (Figure 1): in the examined excisions, the cavity wall is composed of hyalinised scarred tissue with a large number of blurred round-cell plasmocytic infiltrates which infiltrate the adjacent adipose tissue on the periphery; its remnants are also found in the scarred tissue of the wall. The cavity has a character of a pseudocyst with no clear epithelial lining. The scarred tissue comprises also some small neural rami and arteries. The mass is assessed as a tumour-like pseudocyst variation of interstitial retroperitoneal fibrosis (Ormond's disease). No tumour was detected. The finding is consistent with an idiopathic retroperitoneal fibrosis (Ormond's disease). IgG4-positive plasmocytes were detected, 30 IgG4+ cells/HPF. The subsequent

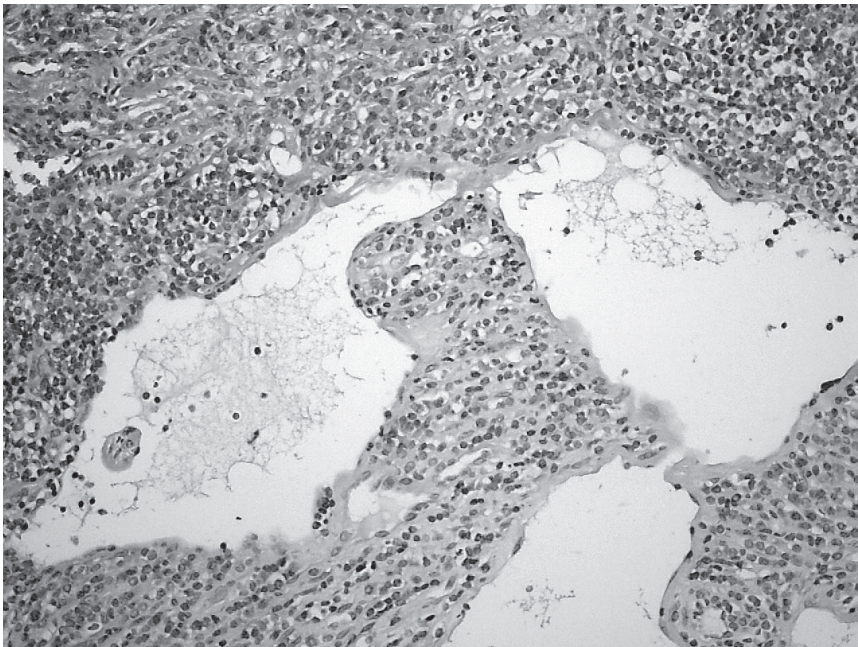


Figure 1 – Tissue with cysts.

PET/CT scan revealed only a slightly increased glucose metabolism and soft tissue lying paravertebrally and posteriorly to the aortic bifurcation, which may present a minor residual mass of active Ormond's disease. The immunology findings of the patient: IgA, IgG, IgM, C3 and C4 part of the complement with no pathological findings, ANA negative, anti-ds DNA negative, ANCA negative. IgG 12.1 G/l, IgG4 0.34 G/l. Since both laboratory findings and imaging methods showed no inflammatory activity, no immunosuppressive therapy was indicated. A cystic form of Ormond's disease was confirmed in the patient, which was not active at the time of diagnosis. The patient has been regularly followed up at the Department of Clinical Immunology. No activity of the disease has been seen in the last 12 months of follow-up.

### **Discussion**

Diseases associated with IgG4 are a newly defined group of diseases, some of which have been known previously (Stone et al., 2015). They are defined by an increased serum IgG4 concentration and the finding of plasma cells in inflammatory infiltrates producing these antibodies. Their clinical presentation varies greatly – the disease may affect completely different organs and systems. The pathogenetic role of IgG4 has not been established as yet. Our case report revealed an interesting clinical form of Ormond's disease. We did not find the described case of the cystic form of retroperitoneal fibrosis confirmed histopathologically in the literature, only one case of a patient with recurrent retroperitoneal fibrosis (RPF), in whom a pseudocyst in the periaortic fibrotic mantle was diagnosed without histopathological confirmation (Jansen et al., 2010). The absence of inflammatory activity at the time of surgery is another interesting feature. It confirms our experience that there are patients in whom the disease is not clinically active (Průcha et al., 2016). For practical reasons, it is important to mention that the disease responds well to systemic corticosteroid therapy. Immunosuppressive therapy is the treatment of choice and has no alternative (Scheel and Feeley, 2013). Patients with a hollow renal system or abdominal aorta are treated by both urologist and vascular surgeon. Systemic corticosteroid monotherapy, however, leads more often to exacerbation of the disease; therefore, combined immunosuppression is the preferred method of treatment, if it is possible.

### **Conclusion**

Diseases associated with IgG4 are a relatively rare group of diseases, often with unpronounced clinical symptomatology, affecting a great variety of organs and systems. Ormond's disease is one example of an IgG4-associated disease. We diagnosed a rare case of a polycystic form of Ormond's disease. Its diagnosis and treatment require a multidisciplinary approach that will be of greater value to the patient.

## References

- Jansen, I., Hendriksz, T. R., Van Bommel, E. F. (2010) Pseudocyst formation in retroperitoneal fibrosis relapse. *Br. J. Radiol.* **83**, e111–e113.
- Kamisawa, T., Zen, Y., Pillai, S., Stone, J. H. (2015) IgG4-related disease. *Lancet* **385**, 1460–1471.
- Průcha, M., Kolombo, I., Štádl, P. (2016) Combination of steroids and azathioprine in the treatment of Ormond's disease – a single centre retrospective analysis. *Prague Med. Rep.* **117**, 34–41.
- Scheel, P. J. Jr., Feeley, N. (2013) Retroperitoneal fibrosis. *Rheum. Dis. Clin. North Am.* **39**, 365–368.
- Stone, J. H., Brito-Zerón, P., Bosch, X., Ramos-Casals, M. (2015) Diagnostic approach to the complexity of IgG4-related disease. *Mayo Clin. Proc.* **90**, 927–939.