Case Report ISSN 2768-6647

Medical and Clinical Case Reports

IgG4-Related Intense Cervicitis: A Rare Case Report

K Ikouch[1], O Wajih Fz[1], Hachami[1], S Tossi[1], H Boufettal[1], S Mahdaoui[1], N Samouh[1], Meryeme Azim[2], Nissrin Bennani-Guebessii[2] and Mehdi Karkouri[2]

[1]*Department of Gynecology Wing 08 CHU IBN ROCHD Casablanca.*

[2]*Department of Anatomopathology CHU IBN ROCHD Casablanca.*

***Correspondence:**
K Ikouch, Department of Gynecology Wing 08 CHU IBN ROCHD Casablanca.

Received: 14 Jul 2023; **Accepted:** 21 Aug 2023; **Published:** 28 Aug 2023

Citation: K Ikouch, O Wajih Fz, Hachami, et al. IgG4-Related Intense Cervicitis: A Rare Case Report. Med Clin Case Rep. 2023; 3(3): 1-3.

ABSTRACT

IgG4-related disease (IgG4-RD) is an inflammatory process. The literature on IgG4-RD is rather limited, and cervical involvement is rare. In such cases, the presentation may mimic other pathologies, particularly cancerous. This is a clinical case of a 42-year-old patient who presented with low back pain and dysuria. Radiological imaging revealed an advanced cervical process with repercussions on the upper urinary tract. IgG4-RD involving the cervix is a difficult condition, both in diagnosis and treatment.

Keywords

Intense cervicitis, IgG4-RD, Inflammatory pseudotumour.

Introduction

IgG4-related disease (IgG4-RD) is a fibro-inflammatory condition, characterised by lesions of tissue hypertrophy and lymphoplasmacytic infiltrate rich in IgG4 and plasma cells; the infiltrate is described as storiform fibrosis and phlebitis obliterans. Elevated serum IgG4 levels often occur, but the sensitivity and specificity of these antibodies are unknown [1,2]. Since the disease was first described and associated with autoimmune pancreatitis two decades ago [3], it has been described in virtually every part of the body [4-6].

The diagnosis of IgG4-RD in the cervical region is difficult. To date, the medical literature is based on sporadic case reports, and information on this specific IgG-RD entity is scarce. Given that treatment of the disease is considered simple and safe (mainly steroid-based), early diagnosis is of immense importance and could avoid unnecessary therapeutic and diagnostic steps, in particular surgery [7]. In this article we report the case of a young woman who presented with a cervical mass suspected of malignancy, the results of multiple biopsies of which were inconclusive, requiring laparotomy and total hysterectomy with preservation of the adnexa.

Clinical Case

Mrs LH, 42, 3G/3P, had a thyroidectomy 5 years ago, an operation for an extrauterine pregnancy 8 years ago and an occlusion 8 years ago.

The patient presented with low back pain associated with dysuria for 8 months prior to her consultation: Tissue lesion process centred on the cervix, well limited measuring 63x79mm; inferiorly it extends to the lower two thirds of the vagina; laterally it extends to the parametrium and encompasses the two ureters responsible for minimal upstream ureterohydronephrosis, completed by a pelvic MRI which concluded: infiltrating cervico-isthmic lesion process with discrete hydrometry Stage (IIIB N1 Mx). Initial cervical biopsy: pseudopolyploid exulcerated and suppurated endocervicitis with inconclusive specificity.

A second scan-guided biopsy with staged biopsy curettage was performed, with the following histological results: Cervical biopsy: Exo- and endo cervicitis, chronic pseudopolypoid, rich in plasma cells. Biopsy of the endometrium: Morphological appearance of a quiescent secretory endometrium. Examination under spinal anaesthesia revealed an impassable cervix with bilateral parametrial infiltration and infiltration of the anterior vaginal wall down to the lower third of the vagina. A biopsy of the cervix and vagina was therefore performed.

Histological Result

Cervical biopsy: Chronic junctional cervicitis with presence of a poorly preserved spindle cell population in the lower 2/3 of the cervix. Immunohistochemistry: Chronic follicular cervicitis with no evidence of malignancy. Vaginal biopsy: No malignancy; after a consultation meeting attended by gynaecologists, urologists and visceral surgeons, we opted for surgical exploration.

Surgical exploration revealed a tumour induration in relation to the uterine cervix, and a total interannexal hysterectomy was performed. The anatomopathological result showed intense cervicitis with IgG4.

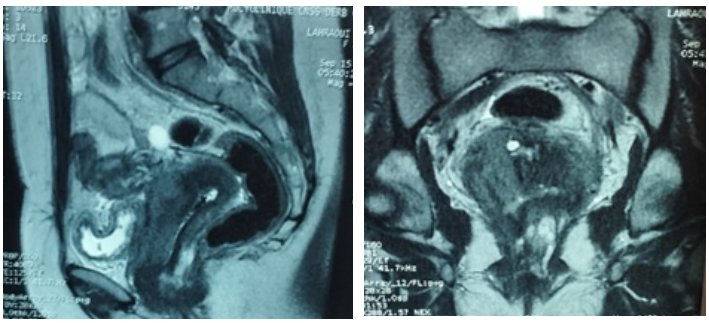

Figure 1: Thickening of the cervico-isthmic region measuring 102x65x86 mm with spiculated contours.

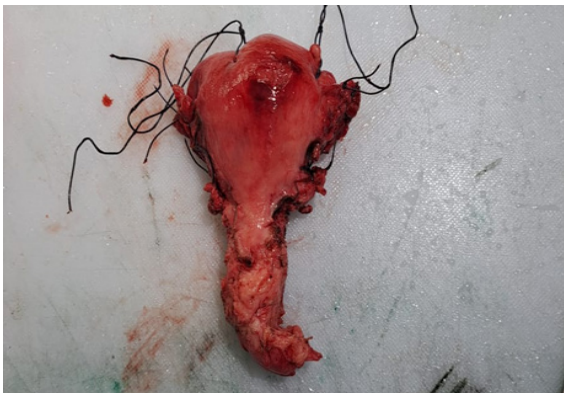

Figure 2: Macroscopic appearance of the total hysterectomy specimen with preservation of the adnexa.

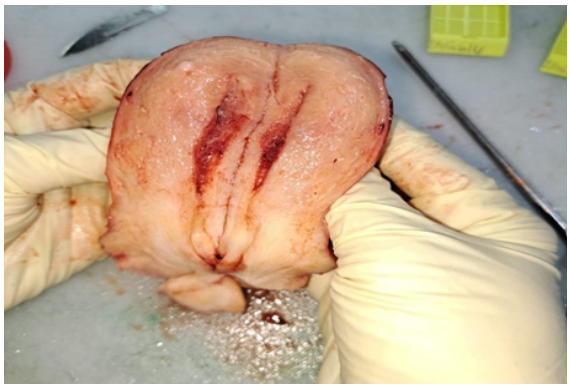

Figure 3: Macroscopic sectional view of the hysterectomy specimen.

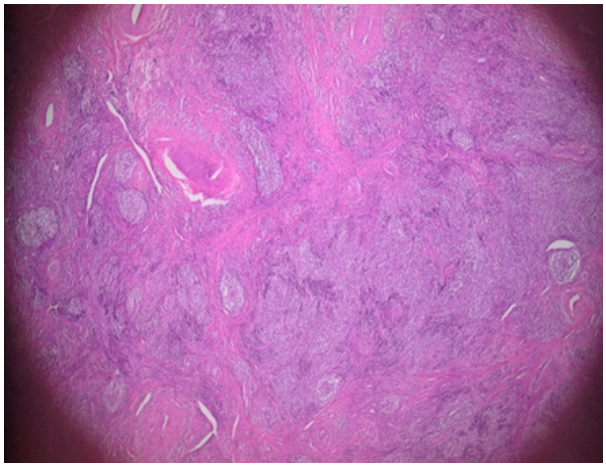

Figure 4: (H&E, x100): Abundant follicular cervicitis made up of plasma cells and lymphocytes with fibrosis.

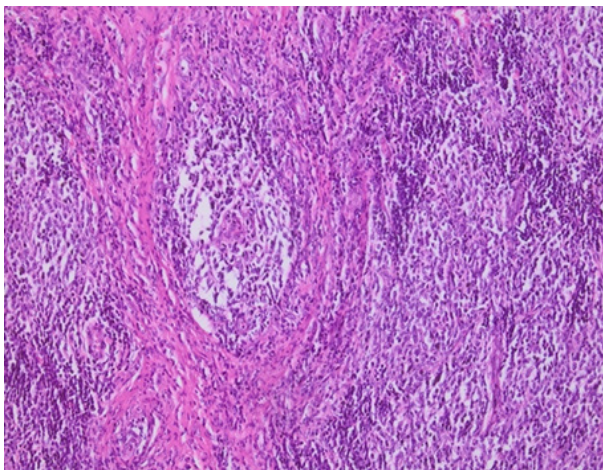

Figure 5: (H&E, x200): Abundant follicular cervicitis made up of plasma cells and lymphocytes with fibrosis.

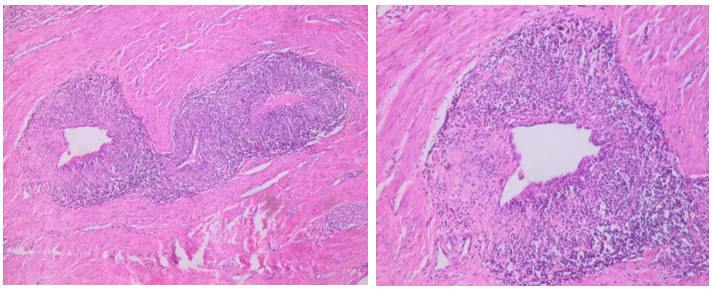

Figure 6: (H&E, x100): Obliterative phlebitis.

Discussion

Since IgG-RD was first diagnosed as autoimmune pancreatitis more than two decades ago [3], various patterns have been identified in different parts of the body. Sometimes the process is confined to a single organ, but the fibrotic and inflammatory process can also involve several organs. Due to the unspecified inflammatory and infiltrative pathophysiological nature, there is often a misdiagnosis

of infections, autoimmune diseases and space lesions [8]. IgG4-RD can mimic several conditions, including infections, malignancies and autoimmune diseases [2]. Thus, the differential diagnosis can be broad and include not only imaging studies, but also serological investigations, including human immunodeficiency virus (HIV) testing, antinuclear antibody (ANA) titres and neutrophil cytoplasmic antibody (NCA) titres, with the aim of excluding other conditions [9]. The gold standard for the diagnosis of IgG4-RD is histopathology. The main morphological features of IgG4-RD are threefold: a dense lymphoplasmacytic infiltrate, storiform fibrosis and phlebitis obliterans [2]. In the consensus statement on IgG4-RD pathology, an attempt was made to create criteria for pathological diagnosis in various organs such as the pancreas, lungs, lymph nodes and salivary glands.Of secondary importance, IgG4 immunostaining and HPF calculation and measurement of the IgG4/IgG ratio are recommended, with a full threshold value when the plasma cell ratio is greater than 40% [10]. Serum IgG4 levels are not mandatory for the diagnosis of IgG4-Rd. There are cases where levels are increased, but the sensitivity and specificity of immunoglobulin levels are still uncertain [1]. To date, there is a lack of randomised controlled trials examining the appropriate treatment of IgG-Rd, particularly in cases of purely organic involvement. Glucocorticoids are generally the first line of treatment. In 2010, a consensus statement from 17 medical centres in Japan suggested initial treatment with prednisolone for 14-30 days [11]. The diagnostic and imaging process is not a simple challenge for clinicians and can be time-consuming. However, whenever a clinical suspicion arises, whether due to treatment failure, disease recurrence or unusual surgical findings, the diagnostic-pathological process can be directed to provide a diagnosis in a short period of time.

Conclusion

IgG4-RD is a rare disease with variable presentations. Diagnosis requires a high degree of clinical suspicion, and clinico-pathological correlation with biopsies is mandatory.

References

1. Deshpande V, Zen Y, Chan JK, et al. Consensus statement on the pathology of IgG4-related disease. Mod Pathol. 2012; 25: 1181-1192.
2. Stone JH, Zen Y, Deshpande V. IgG4-related disease. N Engl J Med. 2012; 366: 539-551.
3. Hamano H, Kawa S, Horiuchi A, et al. High serum IgG4 concentrations in patients with sclerosing pancreatitis. N Engl J Med. 2001; 344: 732-738.
4. Ebbo M, Patient M, Grados A, et al. Ophthalmic manifestations in IgG4-related disease: Clinical presentation and response to treatment in a French case-series. Medicine. 2017; 96: e6205.
5. Ozawa M, Fujinaga Y, Asano J, et al. Clinical features of IgG4-related periaortitis/periarteritis based on the analysis of 179 patients with IgG4-related disease: A case-control study. Arthritis Res Ther. 2017; 19; 223.
6. Gradoni P, Frausini G, Pandolfini M, et al. Laryngeal involvement of immunoglobulin G4-related disease: Case report and. B-ENT. 2019: 15; 51-54.
7. Schiffenbauer AI, Wahl C, Pittaluga S, et al. IgG4-related disease presenting as recurrent mastoiditis. Laryngoscope. 2012; 122: 681.
8. Khosroshahi A, Wallace ZS, Crowe JL, et al. International consensus guidance statement on the management and treatment of IgG4-related disease. Arthritis Rheumatol. 2015; 67: 688-699.
9. Hofmeyr LM, Herbst G, Pretorius E, et al. Diagnosis of petrous apex IgG4-related disease via a middle cranial fossa craniotomy and temporal bone biopsy. Front Neurol. 2022; 9: 957.
10. Sato Y, Kojima M, Takata K, et al. Systemic IgG4-related lymphadenopathy: A clinical and pathologic comparison to multicentric Castleman's disease. Mod Pathol. 2009; 22: 589-599.
11. Kamisawa T, Okazaki K, Kawa S, et al. Japanese consensus guidelines for management of autoimmune pancreatitis: III. Treatment and prognosis of AIP. J Gastroenterol. 2010: 45; 471-477.