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IgG4-related Disease of the Ovary Masquerading as Chronic Tubo-Ovarian Mass with Deep Infiltrating Endometriosis: A Diagnostic Challenge

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Abstract: ***IgG4-related disease (IgG4-RD) is a rare immune-mediated fibro-inflammatory disorder that can involve multiple organs and frequently mimics malignancy or chronic inflammatory conditions. Genital tract involvement is uncommon, and ovarian involvement is extremely rare.[1] So far only 4 cases of ovarian involvement have been reported, with all of them mimicking malignancy followed by HPE confirmed diagnosis. The youngest reported till date was 36 years. We report a case of a 20-year-old unmarried woman presenting with long-standing lower abdominal pain and cyclical bleeding per rectum, clinically and radiologically suggestive of a chronic tubo-ovarian mass with deep infiltrating endometriosis. Imaging had revealed a complex adnexal mass with pelvic adhesions and right-sided hydroureteronephrosis. The patient underwent exploratory laparotomy with right salpingo-oophorectomy following bilateral ureteric stenting. Histopathological examination revealed dense lymphoplasmacytic infiltration with fibrosis, consistent with IgG4-related disease of the ovary confirmed on IHC. This case highlights the diagnostic dilemma posed by ovarian IgG4-RD and emphasizes the importance of histopathology and immunohistochemistry in establishing the diagnosis and preventing unnecessary radical surgery.***

Keywords: IgG4-related disease, Ovary, Tubo-ovarian mass, Endometriosis, Pseudotumor

1. Introduction

IgG4-related disease is a systemic fibro-inflammatory condition characterized by lymphoplasmacytic infiltration rich in IgG4-positive plasma cells, storiform fibrosis, and variable elevation of serum IgG4 levels.[2] It most commonly affects the pancreas, salivary glands, biliary tract, kidneys, and retroperitoneum.[3] Involvement of the female genital tract is rare, with only isolated cases of ovarian IgG4-RD reported in the literature.[4- 6]

Ovarian IgG4-RD often presents as a mass lesion and closely mimics ovarian malignancy, tubo-ovarian abscess, or endometriosis, leading to extensive surgical intervention.[7] We report a rare case of IgG4-related disease of the ovary presenting as a chronic tubo-ovarian mass with deep infiltrating endometriosis.

2. Case Report

A 20-year-old unmarried woman presented with complaints of lower abdominal pain for eight years, insidious in onset and gradually progressive, with worsening over the last four months. Pain was predominantly localized to the lower abdomen and aggravated by walking and climbing stairs. There was no associated nausea or vomiting. She also reported cyclical bleeding per rectum since 2018, suggestive of deep infiltrating endometriosis and had received treatment for the same.[8]

She had no bowel or urinary complaints. Menstrual history revealed last menstrual period one month back, associated with bleeding per rectum. She was not sexually active. There were no medical comorbidities. The patient had undergone pelvic surgery at 3y of age for which documents were not available. She also had diagnostic laparoscopy in 2017in view of hematometra, hematosalpinx, endometriosis, was noted to have cervical atresia and was started on Anti tubercular therapy twice suspecting genital tuberculosis for 3 months. The patient was initially started with cat 1 drugs which she defaulted and later started on cat 2 drugs. The patient also underwent examination under anaesthesia followed by drainage of hematometra in 2018 with Cu-T insertion which was later removed in October 2018. The patient also gave history of cervical dilatation in July and november 2019

On examination, she was hemodynamically stable with no pallor or lymphadenopathy. Abdominal examination revealed a suprapubic mass corresponding to 16–18 weeks' size, firm, irregular, non-mobile, and non-tender. Per-rectal examination revealed a 1 × 1 cm non-tender mass, suspicious for deep infiltrating endometriosis.

Laboratory investigations showed hemoglobin of 8.9 g/dL. Tumor markers were within normal limits. Ultrasonography revealed a large thick-walled cystic lesion with tubular components in the right adnexa measuring 8.6 × 7.0 × 7.6 cm with pelvic adhesions and right-sided hydroureteronephrosis. MRI pelvis demonstrated a well-defined collection measuring 7.4 × 8.8 × 9 cm anterior to the right iliopsoas muscle and a

lesion in the posterior wall of the upper one-third of the rectum, suggestive of deep infiltrating endometriosis.

An initial diagnosis of Deep infilterating Endometriosis was made based on the clinical findings and sonographic investigations and A multidisciplinary approach was planned involving the General Surgery, Gynaecology and Urology teams. The patient underwent bilateral DJ stenting followed by exploratory laparotomy. Intraoperatively, a frozen pelvis with grossly distorted anatomy was noted. Necrotic tissues noted below the rectus- sample taken and sent for histopathology and CBNAAT. A right tubo-ovarian mass measuring approximately 8 × 10 cm was identified, densely adherent to the uterus, bowel, bladder, and lateral pelvic wall. On further dissection of the mass, purulent content was noted, collected and sent for testing and cultures. Very minimal ovarian tissue noted in right side and left ovary and tube were not visualised. In suspicion of malignancy, Surgical oncology team was called intraoperatively. Right salpingo-oophorectomy with extensive adhesiolysis was performed.

The patient was advised for colonoscopy, but couldn't be done due to financial constraints.

The pus culture reported to be MDRO positive for escherichia Coli and was started on Higher Antibiotics. However, the pus and tissue tested negative for tuberculosis and malignancy.

Histopathological examination revealed dense lymphoplasmacytic infiltration with fibrosis consistent with IgG4-related disease of the ovary.[9]

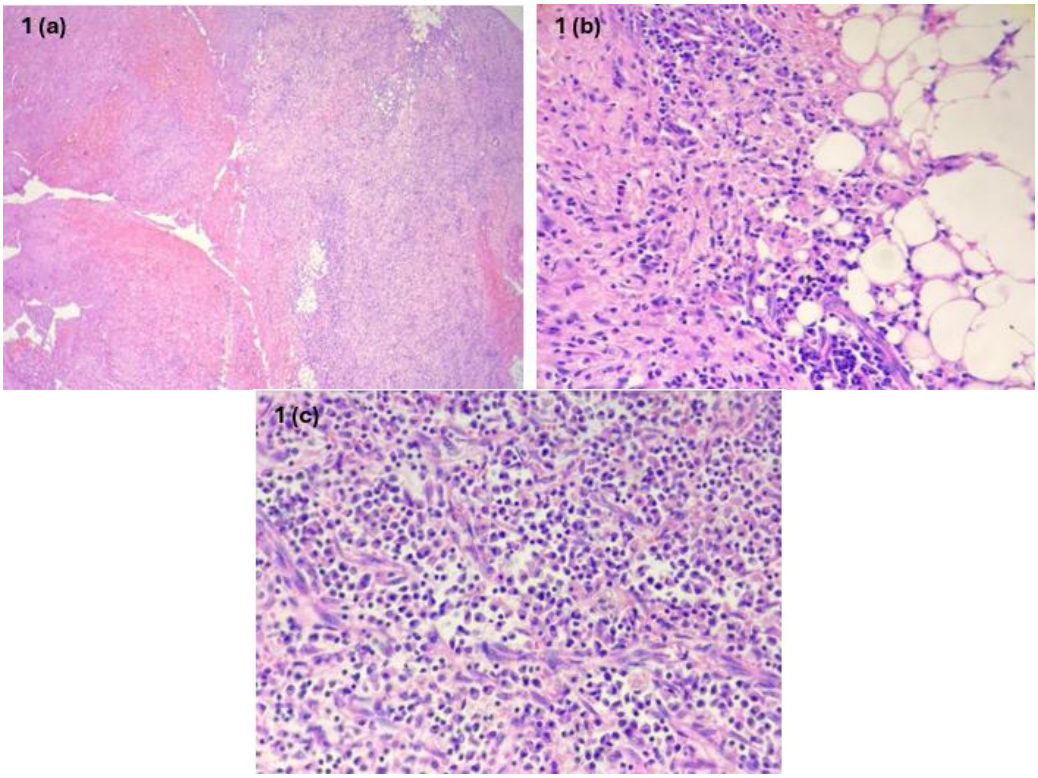

Figure 1: (a)H&E; 4x magnification. (b)H&E; 10x magnification. (c)H&E; 40x magnification. (a)(b)(c)-Omental tissue shows fibrocollagenous tissue with infiltration of chronic inflammatory cells-plasma cells, lymphocytes, foamy histiocytes and eosinophils

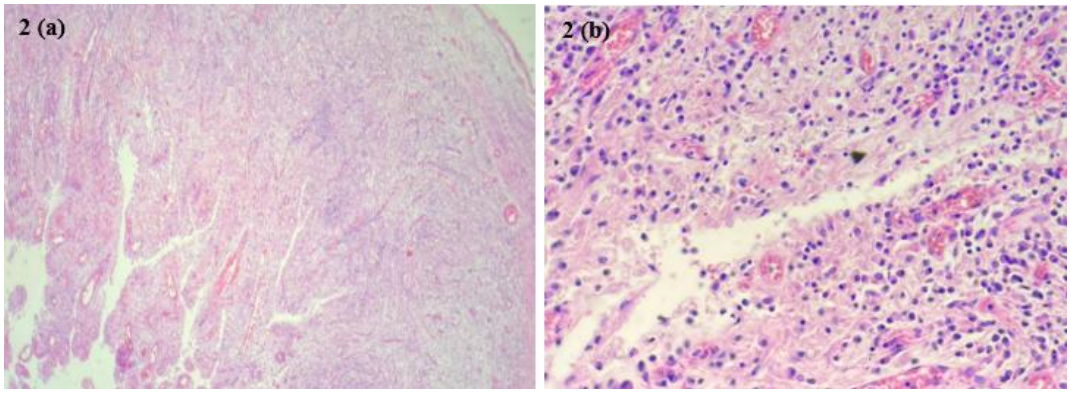

Figure 2: (a)H&E; 4x magnification. (b)H&E; 40x magnification. (a)(b)-Right tubo ovarian mass shows fibrocollagenous tissue with infiltration of chronic inflammatory cells-plasma cells, lymphocytes, foamy histiocytes and eosinophils with focal areas of hemorrhage.

For confirmation of the histopathological diagnosis, sample blocks were sent for immunohistochemistry and Serum IgG levels which reported 235mg/Dl, which confirmed the diagnosis.

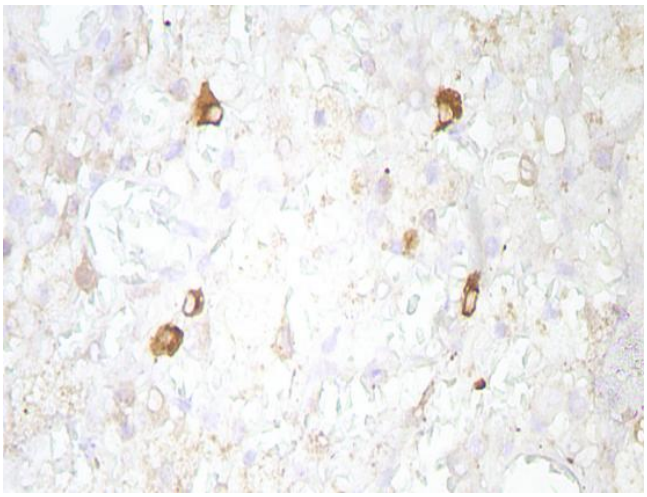

Figure 3: Immunohistochemical characteristics of the plasma cell infiltrate, A CD138 marker highlighting the prominent plasma cell component

The patient was then discharged after completing the course of antibiotics.

The patient was then started on oral steroids for 6 weeks. The patient was planned for further follow up with serial monitoring with sonography. Steroids were stopped after 6 weeks. Follow up scan after 3 months showed no nodules, endometriosis. The patient was then asked for annual follow up. The patient has been asymptomatic till date.

3. Discussion

IgG4-related disease is an under-recognized cause of ovarian mass lesions. The disease often mimics malignancy or chronic inflammatory conditions both clinically and radiologically.[4-7] Most reported cases of ovarian IgG4-RD have been diagnosed postoperatively following histopathological and immunohistochemical evaluation.[6]

The comprehensive diagnostic criteria for IgG4-RD include organ enlargement or mass, elevated serum IgG4 levels (>135 mg/dL), and histopathological features of dense lymphoplasmacytic infiltration, storiform fibrosis, and an IgG4/IgG ratio greater than 40%.[2, 10] The condition responds well to corticosteroid therapy when diagnosed early.[11]

In our patient, long-standing symptoms, distorted pelvic anatomy, and imaging findings led to extensive surgical management. This case underscores the importance of including IgG4-RD in the differential diagnosis of adnexal masses with normal tumor markers.

4. Conclusion

IgG4-related disease of the ovary is a rare entity that can closely mimic malignancy and chronic pelvic inflammatory conditions. A high index of suspicion, combined with histopathological and immunohistochemical evaluation, is essential for accurate diagnosis. Awareness of this condition may help avoid unnecessary extensive surgery and facilitate appropriate medical management.

Clinical Significance

- IgG4-RD should be considered in chronic atypical adnexal masses
- Normal tumor markers do not exclude serious pathology
- Histopathology and immunohistochemistry are essential for diagnosis
- Early recognition may prevent radical surgery

Declarations

Funding: None

Conflict of Interest: None declared

Ethical Approval: Not required for single case report as per institutional policy

Patient Consent: Written informed consent was obtained from the patient

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