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IgG4 Related Disease of the Paratestis

Dr. K. S. Jansi Prema, Dr. Anila Abraham Kurien

Abstract: ***IgG4 related disease (IgG4-RD) is characterized by tumefaction or infiltration of the tissue by IgG4 positive plasma cells and increased serum IgG4 levels. It affects various organs like pancreas, submandibular glands, thyroid, lungs, pituitary, retroperitoneal tissue, kidney and the prostate. IgG4D in the paratesticular region is rare and only a few cases have been reported so far. Here we report a case of IgG4-RD which presented as a pseudotumor in the left paratesticular region.***

Keywords: IgG4 related disease, paratesticular pseudotumor

1. Introduction

IgG4-RD is a fibro inflammatory condition characterized by increased serum IgG4 levels and tissue infiltration by IgG4 positive plasma cells. The first case of IgG4RD was reported in 2001 by Hamano et al. He reported elevated serum IgG4 concentration in patients diagnosed with sclerosing pancreatitis. Subsequently several diseases are included under the umbrella term of IgG4RD: Mikulicz's disease, Riedel thyroiditis, hypophysitis, interstitial pneumonitis, prostatitis, interstitial nephritis, inflammatory aortic aneurysm, retroperitoneal fibrosis and inflammatory pseudotumor. IgG4D presenting as paratesticular fibrous pseudo tumor (PFPT) is rare.

Case Report: A 57 year old male diabetic patient presented with left scrotal swelling and pain for 45 days preceded by history of trauma in the scrotal region. Ultrasound evaluation showed left hydrocele with hypoechoic solid space occupying 2.6 cm * 1.9 cm lesion in the left spermatic cord. He was further treated with antibiotics and anti-inflammatory drugs for five days. No significant improvement noted. FNAC of the lesion was suggestive of mesenchymal / spindle cell lesion. On 17.9.2019, he underwent left inguinal high level orchidectomy.

Grossly, the lesion was a well circumscribed, firm mass measuring 2.9 centimeter in greatest diameter. Cut section showed a yellowish white solid lesion which was firm in consistency and had an ill defined whorled appearance. There were no areas of necrosis or haemorrhage. The lesion encased the vas deferens and diffusely infiltrated the paratesticular tissue (Figure 1).

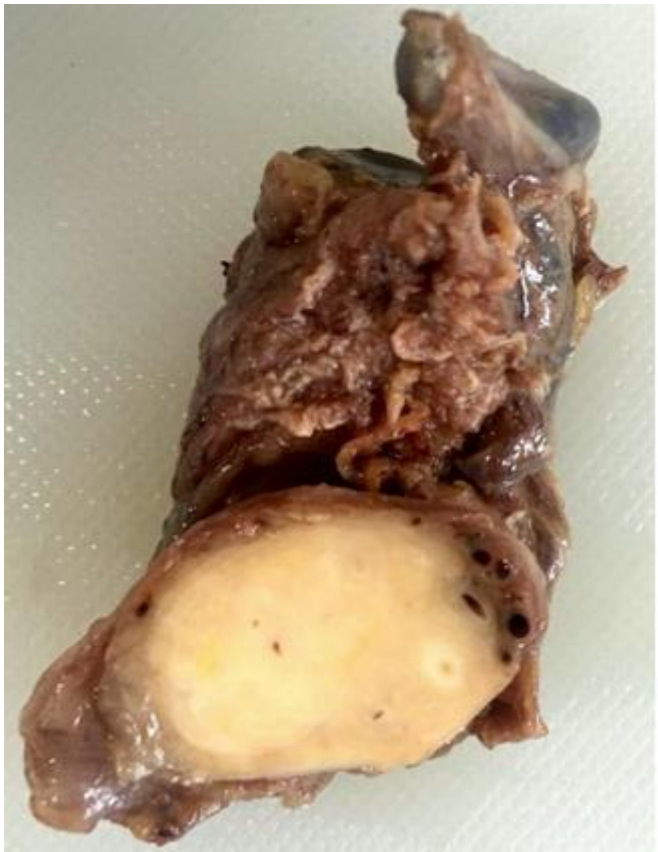

Figure 1: Cut surface shows a yellowish-white lesion with a whorled appearance encasing the vas deferens

Microscopic examination showed hyalinized, collagen rich tissue with storiform pattern of fibrosis densely infiltrated by lymphocytes and plasma cells. Obliterative phlebitis, with lymphoplasmacytic infiltration in the wall of many of the veins causing luminal obliteration, was noted (Figures 2 & 3).

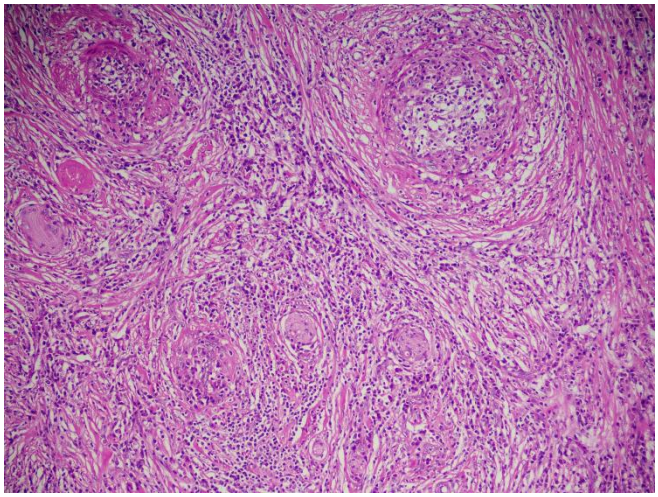

Figure 2: Hematoxylin and eosin stained section shows storiform fibrosis, dense lymphoplasmacytic infiltrate and obliterative phlebitis

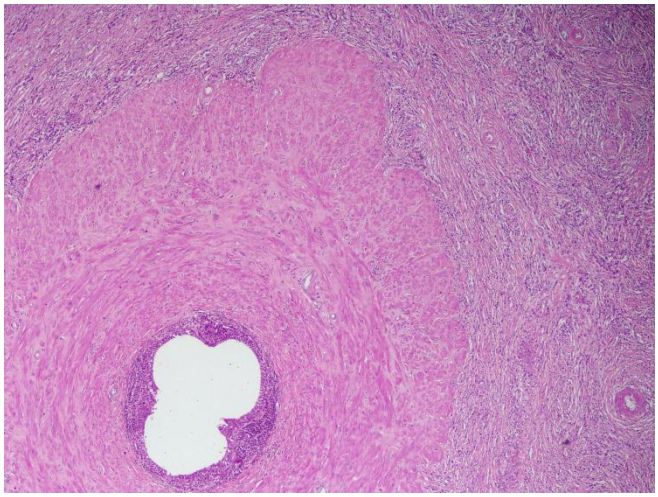

Figure 3: The vas deferens is encased by storiform fibrosis

There was no nuclear atypia or atypical mitosis. IgG4 immunostaining was performed. There were 20 to 30 IgG4 positive plasma cells per high power field (Figure 4).

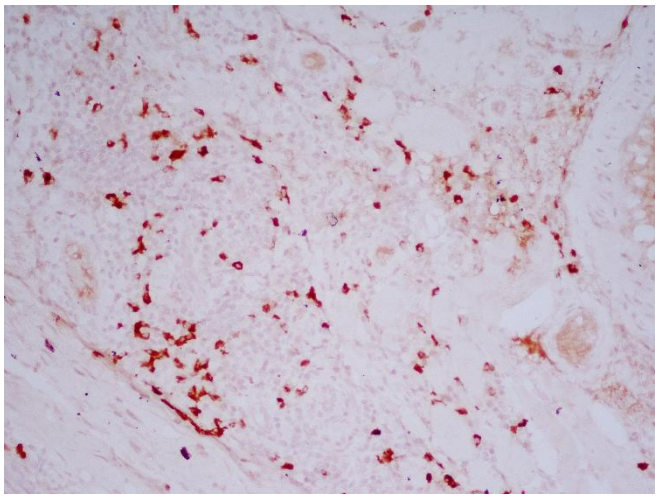

Figure 4: Immunostain with IgG4 shows a dense infiltrate of IgG4 positive cells, X200

2. Discussion

IgG4-RD is characterized by pseudotumor formation or diffuse fibrosis along with infiltration of IgG4 positive plasma cells. Serum IgG4 concentration is markedly elevated. [1] It is a fibroinflammatory condition with multiple organ involvement either synchronosly or metachronously. The first case of IgG4 disease in paratesticular region was reported by Hart et al in 2012. [2] Since then only a few cases are reported. [3,4]

PFPT are observed in the tunica vaginalis, tunica albuginea and epididymis. [4]

Most cases of PFPT were reported on the left side. Our patient also had left sided involvement. Paratesticular IgG4RD shows biphasic age involvement affecting adolescents and old age. [5]

The following are the comprehensive criteria to diagnose IgG4RD:

1) Clinical finding of diffuse or localized swelling in one or more organs
2) Elevated serum IgG4 levels (≥135mg/ dl) and
3) Histopathological finding of marked lymphoplamacytic infiltration and fibrosis (Infiltration by IgG4+ plasma cells. Ratio of IgG4+/IgG+ cells >40% and >10 IgG positive plasma cells per high power field).

However there are no specific diagnostic criteria for IgG4RD in the paratesticular region. Our patient presented with swelling in the paratesticular region. Microscopic examination of the tumor revealed marked lymphoplasmacytic infiltration with typical obstructive phlebitis. IgG4 immunostaining showed 20 to 30 IgG4 positive plasma cells per high power field.

He underwent high inguinal orchidectomy. For PFPT orchidectomy should be avoided and pharmacotherapy like corticosteroids should be tried. Not all patients of scrotal IgG4RD can be diagnosed preoperatively because of the lack of characteristic findings. [5]

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