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Ormond disease: an old disease with a new name

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Abstract

John Ormond is credited with the first description of idiopathic retroperitoneal fibrosis (RF) in the English literature. Seventeen years later, he speculated correctly that the disease was systemic and was in the same group of diseases as lupus, scleroderma and periarteritis. Presently, idiopathic RF is considered part of a recently defined group of diseases known as IgG4 related diseases. The disease is generally treated medically but surgery can be required if there is ureteric obstruction.

Keywords

Retroperitoneal fibrosis; Ormond disease; IgG4-related disease.

Introduction

Although the French urologist Joaquin Albarran (1860–1912) first described retroperitoneal fibrosis (RF) in 1905[1], it is the American urologist John Kelso Ormond (1886–1978) who is credited with the first detailed description of the disease in the English literature in 1948[2]. He was in charge of the department of urology at a Detroit hospital for many years. From 1952 to 1955, he worked in a urology department in a medical centre in India. He later engaged in private practice.

Ormond described 2 patients with diffuse fibrosis of the retroperitoneal tissues[2]. His description of the clinical and pathologic features of idiopathic RF established the condition as a clinical entity[2–5]. In his 1965 review of the condition, Ormond wrote that at the time of reporting his 2 cases, he was unable to find similar cases in the literature. However, he wrote: "Since then, Pugh has drawn attention to a report read before the French Urological Society by Perard and Orsini in 1937, and Hache and associates have cited 3 cases described by Albarran in the early 1900s. I have read these reports and agree that they probably represent cases of idiopathic retroperitoneal fibrosis."

This disorder was initially called Ormond disease or syndrome[6], but other names used include periureteritis fibrosa, periureteritis plastica, chronic periureteritis, sclerosing retroperitoneal granuloma, and fibrous retroperitonitis. RF may be idiopathic or secondary to other causes[6].

Idiopathic RF is part of the disease spectrum of chronic periaortitis, a condition characterized by inflammation and fibrosis surrounding the aorta and iliac arteries. Chronic periaortitis also includes inflammatory abdominal aortic aneurysms and perianeurysmal RF. Although these conditions are grouped together because of similarities in their clinical and histologic characteristics, their pathogeneses and epidemiology may differ.

Pathogenesis

RF may be idiopathic or can be caused by:

- drugs (ergot derivatives, methysergide, bromocriptine, β-blockers, methyldopa and hydralazine)
- malignancy (carcinoid, Hodgkin and non-Hodgkin lymphoma, sarcomas, colorectal, breast, prostate and bladder carcinoma)
- infections (tuberculosis)
- radiation therapy for testicular seminoma, colon, pancreatic cancer, retroperitoneal haemorrhage
- surgery (lymphadenectomy, colectomy, aortic aneurysmectomy)

Ormond wrote that the course of a typical case could be divided into three stages:

1. Onset of the process
2. Fibrosis enveloping the retroperitoneal structures
3. Healing and contracture of the fibrous mass causing compression of the enveloped structures

Ormond speculated that RF was a systemic disease rather than local and hence it was unlike keloid, Dupuytren contracture or Peyronie disease. It was more closely associated with the collagen diseases such as lupus, rheumatoid arthritis, scleroderma and periarteritis nodosa. He was convinced it was a hypersensitivity reaction to mostly drugs or chemicals[5].

Recent evidence shows that an increasing number of cases of RF are associated with the presence of IgG4-producing plasma cells and RF is a manifestation of IgG4-related disease. IgG4-related disease is a multisystem disease characterized by tumour-like swelling of involved organs, a lymphoplasmacytic infiltrate enriched in IgG4-positive plasma cells, and variable degrees of fibrosis[7]. Cases of isolated IgG4-related RF have been reported, but in most cases of RF associated with IgG4-related disease, other manifestations of the disease are present[7]. In one report, all 14 of the reported cases of IgG4-related RF exhibited involvement of other organs, including the pancreas (11 patients), salivary glands (3 patients), lymph nodes (2 patients), pituitary gland (1 patient), and one patient had mediastinal periaortitis[8]. Recently, a consensus has been reached on the term IgG4-related disease for the overall condition and on nomenclature for its individual organ system manifestations[9,10]. Guidelines for diagnosis include the critical histopathologic findings. The 3 major histopathologic features associated with IgG4-related disease are dense lymphoplasmacytic infiltrate, fibrosis arranged at least focally in a storiform (i.e. matted and irregularly whorled) pattern, and obliterative phlebitis. A mild-to-moderate eosinophil infiltrate is usually present. The inflammatory process frequently forms a tumefactive mass that may cause destruction of the involved organ. Immunohistochemical confirmation is required with IgG4 immunostaining. Although histopathology is the gold standard in diagnosing IgG4-related disease, a serum level twice the upper limit of normal or more is almost diagnostic of the condition.

Treatment

RF is essentially an obstructive uropathy. Untreated patients may eventually progress to end-stage renal disease. Percutaneous and endoureteral techniques seem to be safer than open and laparoscopic procedures. The goals of therapy are to relieve the obstruction, stop the progression of the fibrotic process, and to prevent recurrence. In patients who have hydronephrosis but no significant impairment in renal function, a trial of medical therapy can be initiated provided there is close supervision of renal function and ultrasonographic surveillance of the hydronephrosis. Treatment of secondary RF is aimed at the cause; drug-induced RF may respond to immunotherapy. Idiopathic RF is treated initially with prednisolone but if after 4–6 months of prednisolone therapy there is an inadequate response clinically, serologically, or by imaging,

azathioprine, methotrexate, mycophenolate mofetil or rituximab may be added[11,12]. Some use tamoxifen as initial therapy[11].

Conclusion

John Ormond is credited with the first description of idiopathic RF in the English literature. Seventeen years later, he speculated correctly that the disease was systemic and was in the same group of diseases as lupus, scleroderma and periarteritis. Presently, idiopathic RF is considered part of a recently defined group of diseases known as IgG4-related diseases. The disease is generally treated with medical therapy but surgery is needed if there is a ureteric obstruction.

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