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Original Research Paper

Surgery

A CASE OF LOCALIZED IGG4-RELATED PERIAORTITIS AND VOCAL CORD PALSY

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ABSTRACT IGG4-related disease is a newly recognized idiopathic sclerosing lesions with wide spectrum of manifestations starting with autoimmune pancreatitis (AIP) ,a specific type of chronic pancreatitis and an original member of the IgG4-RD family ,sclerosing cholangitis,sialoadenitis,retroperitoneal fibrosis,tubulointerstitial nephritis etc. In this report, we describe a case of a 31-year old man with marked periaortic fibrous thickening localized to the aortic arch initially presented only as vocal cord palsy. Histologic examination revealed infiltration of lymphoplasmacytes and marked fibrosis with numerous IgG4-positive plasma cells which are hallmark of this disease. The serum concentration of IgG4 was 263 mg/dL,Twice the normal value.Treatment with steroid is well proven for this spectrm of disorder.

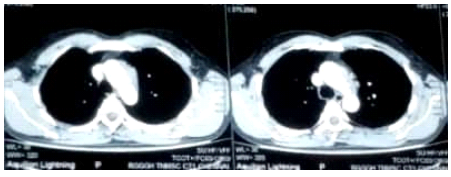
KEYWORDS : Periaortitis,IgG4 disease

INTRODUCTION

Chronic periarteritis and periaortitis are a part of a spectrum of idiopathic diseases characterized by fibroinflammatory reaction around arteries and that of aorta.Chronic Periaortitis includes retroperitoneal fibrosis and inflammatory abdominal aortic aneurysm with elevated inflammatory markers was considered as a systemic disorder.However,recent emergence of IGG4-related disease which is also systemic sclerosing disorder with wide range of involvement involving pancreas(1) ,bile duct(2),salivary gland(3),retroperitoneum(4).Recent studies have suggested that these two disorders overlap,at least in part .We have encountered a case periaortitis that presented initially only with left vocal card palsy.

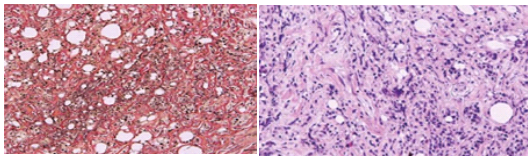
CASE REPORT

A 52-year-old male was admitted to our hospital with history of hoarseness of voice for 1 month with no previous history of any serious disease. He is a known smoker.He is also an diabetic on treatment for past 6 months. On physical examination, blood pressure was 100/60 mm Hg, and other vital signs were within normal range. Fibreoptic bronchoscopy showed left vocal cord was palsy.



Img (a,b).Ct chest periaortic mass in mediastinum

A computed tomography (CT) revealed infiltrative soft tissue density lesion in anterior mediastinum anterior to arch of aorta abutting pre-vascular space inferiorly extending upto descending aorta and superiorly upto origin of left subclavian artery with few discrete prevascular lymph nodes noted (img a,b) .MRI Chest was reported as soft tissue thickening in aortopulmonary window and upper para-aortic region. The following laboratory data were obtained: serum C-reactive protein, 0.62 mg/dL; erythrocyte sedimentation rate, 12 mm/hr; IgG4, 312 mg/dL; and absence of detectable antinuclear antibodies.Transbronchial node biopsy showed no evidence of malignancy. Right upper limb arterial Doppler showed monophasic flow.Chamberlain procedure was performed for diagnosis and few lymph nodes were biopsied from aortopulmonary window .



Img (c,d)-HPE staining shows lymphoplastmyitc infiltrstion with fibrosis

Histologic analysis of the biopsy specimen demonstrated diffuse infiltration of lymphoplasmacytes and marked fibrosis, with no significantly atypical cells(img a,b). Immunostaining for CD3 and CD20 showed that the lymphocytes were polyclonal and included mainly CD20-positive B cells and CD3-positive T cells. The number of CD20- positive B cells was larger than that of CD3-positive T cells. In addition, immunostaining for IgG4 revealed many IgG4-positive plasma cells within the lesion. On the basis of these findings, we made the pathologic diagnosis of IgG4-related periarteritis involving left recurrent laryngeal nerve. IgG4-positive disease frequently accompanies sclerosing pancreatitis.However, USG and CT showed no abnormality in the pancreas or retroperitoneum. No finding characteristic of sclerosing pancreatitis was observed. Patient was started with 40mg prednisolone for 2 months and showed good improvement,following which dose was tapered.

DISCUSSION

The spectrum of IgG4-Related disease(IgG4-RD) links many conditions once regarded as isolated, single-organ diseases without any known underlying systemic condition. IgG4 disease was established while evaluating extrapancreatic lesions in patients with autoimmune pancreatitis patients(4),an original member of IgG4-RD family and also the most common presentation followed by other manifestations like Mikulicz's disease [5], respiratory disorders [6], sclerosing cholangitis [9], retroperitoneal fibrosis [3], tubulointerstitial nephritis [7],dacryoadenits and prostatitis [8]. Pathologically, IgG4-related diseases are characterized by a diffuse lymphoplasmacytic infiltration, irregular "storiform" fibrosis, obliterative phlebitis, and severe infiltration of IgG4-positive plasma cells.IgG4-RD also includes systemic cardiovascular disease that involves arterial/aortic systems which has revealed concept of periarteritis/periaortitis

The idiopathic retroperitoneal fibrosis, also called as Ormond's disease(9) is now classified within grouping of chronic periaortitis .The three major components of chronic periaortitis are IgG4-

related retroperitoneal fibrosis, IgG4-related abdominal aortitis, and IgG4-related perianeurysmal fibrosis(10). The subtle presentation of IgG4-related chronic periaortitis non-can lead to diagnostic delay. A poorly localized pain in the back, flanks, lower abdomen, or thighs; leg oedema; and hydronephrosis from ureteral involvement are common presentation of this disease. The disease targets three sites: periaortic/arterial regions, involving connective tissue around the abdominal aorta or its first branches (appendix); periureteral areas, tending to cause ureteral obstruction and hydronephrosis; and a plaque-like mass that broadly involves the retroperitoneum.

IgG4-related disease is the cause of up to two-thirds of cases of idiopathic retroperitoneal fibrosis as evidenced by diffuse infiltration of IgG4-positive plasma cells in thickened fibrous tissue (10). In advanced disease, the ratio of IgG4-positive plasma cells to the total number of plasma cells in tissue can be more helpful diagnostically than the overall number of IgG4 positive plasma cells per high-power field. Even if the classic lymphoplasmacytic infiltrate is not evident in longstanding cases, both storiform fibrosis and obliterative phlebitis are commonly identified.

Although in most cases, the abdominal aorta and neighboring structures are affected, Most common site involved is infrarenal aorta. In few cases only the thoracic aorta is involved(11).

It was more prevalent in patients with an older IgG4-RD onset age and in those with a highly active disease state as evidenced by elevated serum IgG, IgG4, CIC, and sIL2R. High IgG4-RD activity state to most strongly influence the onset of periaortitis/periarteritis(12).

All patients who received corticosteroids showed improvements in wall thickening after therapy, although a fifth exhibited worsening of luminal dilatation .The exacerbation of luminal dilatation was defined as an expansion of luminal diameter > 5 mm at the same site as periaortitis/periarteritis lesions detected at the time of the initial diagnosis (14).Careful monitoring of patients with prior luminal dilatation is necessary.

Our case differs from common presentation of periaortitis in number of ways.Patient had just vocal cord palsy with no other constitutional symptoms.CT revealed a periaortic mass in aortopulmonary window and upper para aortic region compared to much wider and systemic involvement .Localized diseases such as malignant lymphoma should be ruled out in such cases. Presence of the diffuse infiltration of the IgG4-positive lymphoplasmacytic cells without the evidence of atypicality was useful in distinguishing IgG4-related periarteritis from malignant lymphoma in our case. Eventhough exact cause of such localized manifestation is not clear ,Some investigators have suggested that local inflammatory reaction to oxidized low-density lipoproteins and ceroids in the atherosclerotic plaques might cause periarteritis(15).Involvment of left recurrent laryngeal nerve in aortopulmonary window and upper para aortic region would be the propable cause for left vocal cord palsy.

In conclusion, we have presented a case with IgG4- related periarteritis localized to the thoracic aorta. After the induction of corticosteroid therapy, the size of the fibrous mass surrounding the aortic arch decreased recurrent nerve palsy disappeared. In this patient, accurate diagnosis led to suitable treatment

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