CLINICAL REPORT

IgG4-Related Disease (IgG4-RD) Presenting as a Mass in the Carotid Triangle Masquerading Paraganglioma

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Abstract IgG4-related disease (IgG4-RD) which is a protein disorder presented as a mass in the right carotid triangle in a 30 year male patient, who underwent battery of tests is described. The radiologist opined the mass as paraganglioma and the spindle shaped character of the mass also suggested neurogenic tumor in differential diagnosis. Reference to vascular surgeon also opined the same, and advised for incisional biopsy. Histopathology report suggested IgG4-RD and immunochemistry confirmed the final diagnosis. The neck mass which we opined as a single organ disease, proved to be multiorgan disease in PET scan.

Keywords IgG4-related disease · Autoimmune disease · Neck mass · PET scan

Introduction

IgG4-RD is a relapsing remitting immunomediated fibro inflammatory disease characterized by: tendency to mass formation, tissue destruction, in single or multiple sites, characteristic histopathological features consisting of

lymphoplasmacytic infiltration, storiform fibrosis, and usually responding to steroids [1]. In the head and neck region the occurrence of this disease is rare except for few case reports [2]. Demographic studies have shown that the disease is more common in middle age and older age group individuals [3]. Our patient presented with a solitary swelling in right carotid triangle, baffled the diagnosis. Both radiologist and vascular surgeon opined the mass to be atypical paraganglioma or neurogenic tumor due to its radiologic appearance (Fig. 1). FNAC of the lesion was found to be hemorrhagic and hence inconclusive. We have subjected the patient for incisional biopsy along with vascular surgeon and the tissue was sent for histopathological examination, the pathologist reported the tissue to be IgG4-RD and advised immunohistochemistry which confirmed the diagnosis (Fig. 3). A PET scan revealed multiorgan involvement (Fig. 4). As the tumor responds to glucocorticoids, as advised by the rheumatologist, the patient was put on long term steroids. The patient is under follow up for response to the drug regime.

Case History

A male Bengali patient aged 30 years presented to my outpatient complained of a small firm right sided painless slow growing neck swelling in the carotid triangle. The swelling was non pulsatile and not moving on deglutition without any neurological deficit (Horner's syndrome) and also without any local and distant lymphadenopathy. Fiberoptic flexible nasolaryngoendoscopy did not show any nasopharyngeal, laryngopharyngeal pathology. The patient also complained of mild dysphagia, and esophagoscopy did not reveal any obstructive esophageal lesion. Since the patient also complained of numbness of the limbs,

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Fig. 1 CT-scan: moderately defined homogenous minimal enhancing soft tissue thickening in right carotid space

neurologist opinion sought which was unequivocal. Reference was made to the vascular surgeon who opined the mass as atypical paraganglioma or a neurogenic mass, concurred with the radiologist CT report (Fig. 1). On 31.3.15, along with the vascular surgeon we explored the neck which was found to be firm, fibrotic, encircling the great vessels and very carefully the portion of mass excised and sent for histopathological examination. There was not much of bleeding encountered in the procedure. The patient had uneventful post operative course except, he developed hoarse voice on the next post operative day due to paralysis of right vocal cord seen on flexible nasolaryngeal endoscopy. The pathologist report after immunohistochemistry of the removed specimen contained abundant plasmacytes, histiocytes and storiform pattern of connective tissue suggestive of IgG4-RD (Figs. 2, 3). A PET CT revealed multiple organ involvement by surprise (Fig. 4). The patient is put on long term corticosteroid, suggested by the rheumatologist and patient is advised to report for follow up once in 2 months. Patient was serially reviewed upto 6 months. Patient was given 2 shots of injection Rituximab along with oral corticosteroids suggested by rheumatologist. He recovered from vocal cord palsy and his voice became normal. The residual neck mass disappeared with no further progression of other lesions in the body which was shown in the subsequent PET CT.

Fig. 2 Light microscopy showing storiform fibrosis with increased plasma cells ($\times 10$)

Fig. 3 Immunohistochemical staining of IgG4 plasma cells ($\times 40$)

Discussion

IgG4-RD is a protein disorder involving single or multiple organs. IgG4-RD is the new name given to a galaxy of diseases by multispecialty experts namely (1) autoimmune pancreatitis, (2) Mikulicz disease, (3) orbitopathy, (4) retroperitoneal fibrosis, (5) periaortitis, (6) lung interstitial fibrosis, (7) tubulo nephritis, (8) hypophysitis and pachymeningitis, (9) Riedel's thyroiditis, (10) Kuttner tumour, (11) omentitis and many others [1]. Earlier these group of diseases under various headings, described in literature which were presumed to be autoimmune disorder, are now grouped under IgG4-RD, a protein disorder. These disorders previously thought to be unrelated, commonly show features which include tumor like swelling of the involved organ with lymphoplasmacytic infiltrate, enriched with plasma cell with variable degree of fibrosis, of characteristic storiform pattern

Fig. 4 Increased FDG uptake in multiple areas

of tissue in histopathology (Fig. 2), is diagnostic feature of the condition [4]. The serum concentration of IgG4 is found to be raised in 60–70 % of patient of IgG4-RD but is not specific [5]. The disease most commonly occurs in middle age and older age group patients [3]. The patients affected with the disease usually have prevalence of allergic rhinitis or bronchial asthma and this patient had none [6]. They are also found in Castleman disease, sarcoidosis, allergic disorder, Churg Strauss syndrome and large number of other conditions [7]. Special tests like flowcytometry to detect plasmacytes in blood, is used as a “marker” [5]. Radiological investigations like MRI and PET scan are to be done as a routine. CT scan in this case showed the mass in the neck and incidental PET scan revealed multiple organ involvement. By and large, the tissue diagnosis showing the typical storiform connective tissue plasma cells and histiocytes is diagnostic [4]. Natural history of disease is uncertain. The disease responds to long term glucocorticoids oral therapy in early cases. In patients with adverse effects of steroids and

intractable cases, alternate drugs like Rituximab, Azathioprine and Mycophenolate Mofetil are administered [8]. Minority of patients improve with corticosteroids but they relapse after stopping the treatment [9]. Several types of lymphoma has been reported with IgG4-RD and now non-Hodgkin lymphoma is also reported in one case [2]. Hence the prognosis should be guarded.

Conclusion

We are presenting this case because of rarity and the disease masquerading paraganglioma. Hardly the patient had any symptoms related to the multiple organ involvement seen in PET scan (Fig. 4). The fibrous mass encircling the great vessels resembled paraganglioma, proved to be a protein disorder, IgG4-RD. The histopathological reports of presence of plasma cells, histiocytes and storiform pattern of connective tissue clinched the diagnosis [4]. We

have discussed in detail the clinical presentation of the disease which affects multiorgan. And the line of treatment in early cases with long term corticosteroid and in steroid toxicity cases, alternate drugs like Rituximab, Azathioprine, Mycophenolate mofetil are used. There are various tests like flowcytometry which detect the presence of plasmablasts in blood used as a “marker” [5]. It is expensive and not available in all centers. Tissue diagnosis is the best method universally accepted by multinational and multidisciplinary experts, and the role of ultrasound guided FNAC for unapproachable sites is doubtful [3, 4]. The disease is presented under various heads but all the heads are grouped under IgG4-RD which is universally accepted. There are only a few case reports in Head and Neck specialty and the unique presentation in the neck simulating paraganglioma is indented to bring awareness of this emerging disease to the medical fraternity of all disciplines. PET scan is a mandatory investigation as it’s a non invasive test which will detect multiorgan involvement and help in diagnosis and planning the line of treatment. The role of MRI is organ specific. PET scan in this case taken after surgery and not before, opened up new vistas of multiorgan involvement is mandatory.

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Compliance with Ethical Standards

Conflict of interest Prof. Dr. T. Ramadass, Dr. V Balaji, Dr. Sheba, Dr. Vali Ahmed, Dr. Raees declare that they have no conflict of interest.

Ethical Approval This article does not contain any studies with animals performed by any of the authors.

Informed Consent Informed consent was obtained from the patient included in the study.

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