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IgG4 DISEASE OF THE MEDIASTINUM

Medical Education

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KEYWORDS

INTRODUCTION

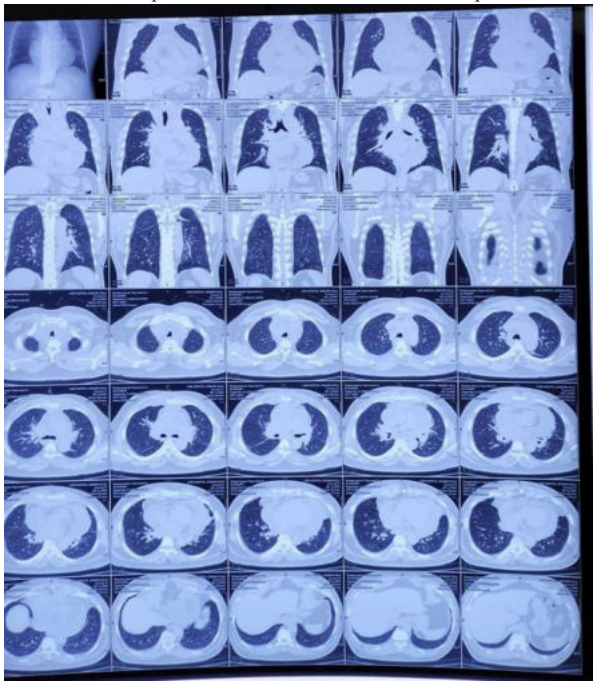
IgG4-related disease (IgG4-RD) is a systemic fibro-inflammatory condition characterized by a tendency to form tumefactive lesions[1]. Its thoracic presentation often manifests as interstitial lung disease or fibrosing mediastinitis. It is sporadic for IgG4-RD to form a well-defined mass in the anterior mediastinum, mimicking an encapsulated thymoma.

It is more frequently associated with the hepatobiliary system, salivary glands, lymph nodes, and kidneys, and it generally manifests as a tumorlike mass in 1 or more sites . It is most commonly diagnosed in the fifth or sixth decades of life, and there is a male predominance[2].

Isolated presentations in the mediastinum are rare.We present a case of IgG4-related disease forming a mass in the anterior mediastinum.

CASE DETAILS

- A 49-year-old male patient presents with a cough associated with shortness of breath and chest pain since the past 2 months, aggravated in the last 15 days.
- H/o present illness : Chest pain was gradual onset, intermittent in nature, aggravated on leaning forward.
- SOB (grade 2- mMrc) was gradual onset, progressive in nature, aggravated on exertion, relieved on rest (in Fowler's position) associated with orthopnea, postural variation was noted.
- Cough was gradual onset, progressive in nature, aggravated on exertion and supine position. Postural variation is noted.
- The complaints are not associated with fever, loss of weight, hemoptysis, expectoration
- General examination: Patient is conscious, coherent and cooperative, he appears dyspneic and visible skeletal muscle wasting is noted. His vitals were within the normal range.
- O/E : JVP was distended , CVS : S1+ , S2+ ; RS : B/L NVBS +, with no added sounds.
- ECG shows low voltage QRS complexes and inversion of the T-waves with ventricular hypertrophy
- Chest x-ray showed enlargement of cardiac silhouette, indicating cardiomegaly with widening of the mediastinum, no obliteration of costo-phrenic angles and no other abnormality was noted in the lung parenchyma .
- Upon his blood examination: ESR was elevated, Serum IgG4 were elevated > 135mg/dl.
- On subsequent FDG PET-CT scan findings show- Moderate pericardial effusion with intensely hypermetabolic ill-defined soft tissue thickening in the pericardial region, mediastinum, bilateral lower cervical region, and diaphragm extending into the anterior chest wall.
- The patient underwent a VATS procedure, and a lymph node biopsy was taken which reported: a dense lymphoplasmacytic infiltrate organized in a storiform pattern, obliterative phlebitis, and a mild to moderate eosinophilic infiltrate. Concentrated serum IgG4 was elevated > 135mg/dl.
- As per the Japanese CCD diagnostic criteria for IgG4-RD : The presentation and investigation of this patient was inclined towards IgG4-Related disease.
- Patient was treated with oral corticosteroid - Prednisolone 40mg/day after and was asked to follow up after 3 months.
- Subsequently in the follow up visit after 3 months, the patient complained of persistent symptoms , despite the treatment given. Following investigations were done -
- 2D ECHO reports showed - Pericardial effusion + Posterior-1.9cm; Anterior - 1.1cm ; Lateral - 3.4 cm ; Apical - 1.0 cm ; RA Side - 3.8 cm; RV - 1.5 cm .
- Upon further examination of Chest-CT showed - Ill defined heterogeneously enhancing soft tissue infiltration of mediastinal fat from the level of thoracic inlet up to the carinal bifurcation predominantly involving the prevascular compartment of the superior mediastinum, encasing the mediastinal vasculature including the superior vena cava, bilateral brachiocephalic vein, aortic arch, main pulmonary artery, brachiocephalic trunk, left subclavian artery and proximal bilateral common carotid arteries causing mild to moderate narrowing of SVC and bilateral brachiocephalic veins with extension of soft tissue up to the anterior chest wall as described above indicating - likelychanges/ sequelae of mediastinal fibrosis. Moderate Pericardial effusion was noted.
- The patient's dosage of Prednisolone was further increased, as per the treatment protocol and was advised for a follow up in 3 months.



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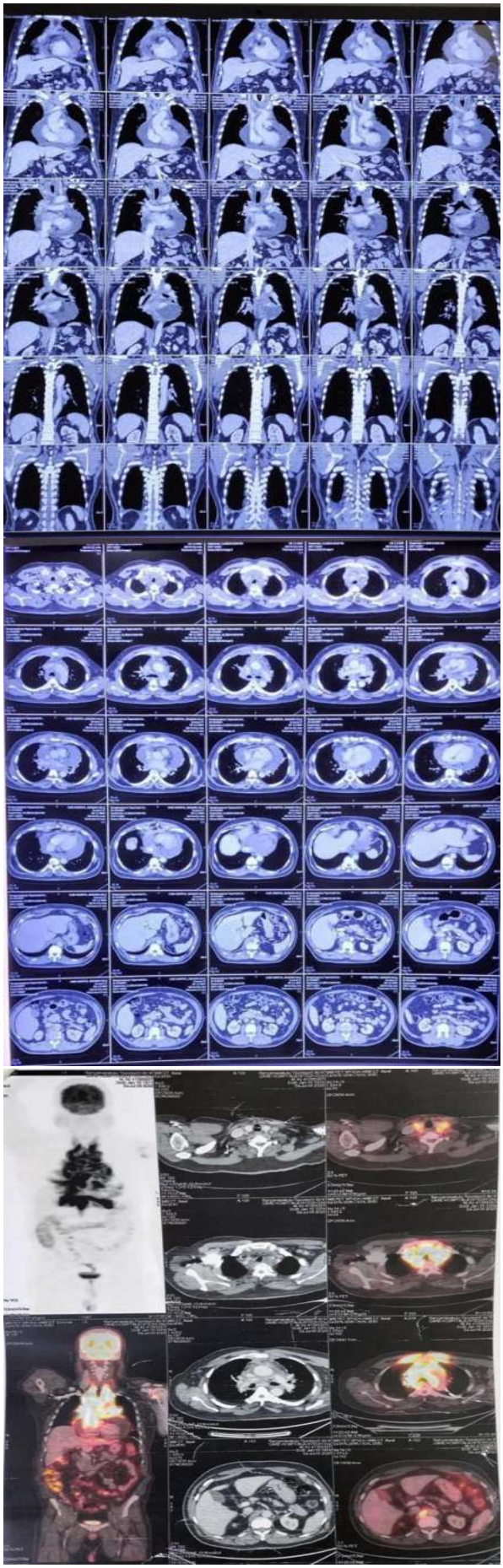

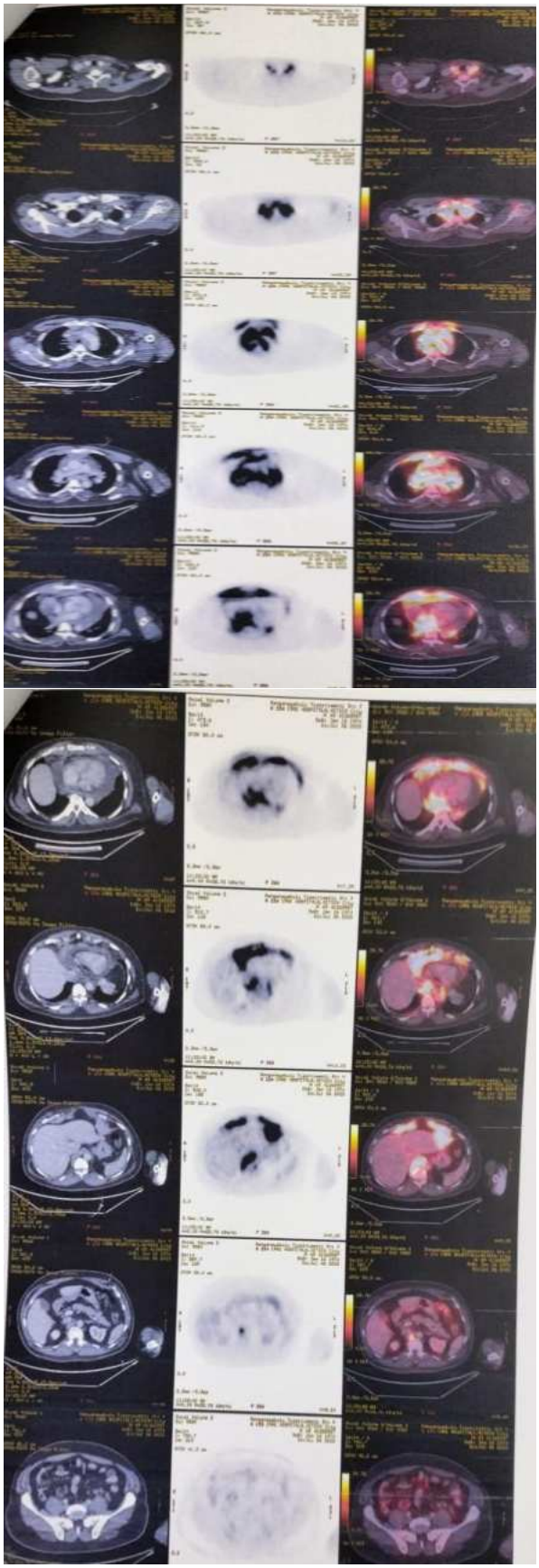

DISCUSSION

IgG4-related disease (IgG4-RD) is a complex disease which requires a multidisciplinary approach. In the respiratory system, it commonly presents as pulmonary nodules, lymphadenopathy, or sclerosing mediastinitis.[3]

Immunosuppressive drugs such as azathioprine, methotrexate, cyclophosphamide,cyclosporine and tacrolimus have also been used. Each of these treatments had varied success rates.

Rituximab can also be considered an effective treatment option for IgG4-ROD that is steroid dependent or steroid intolerant. Rituximab therapy resulted in swift clinical and radiological improvement, many months free of relapse, and few side effects.

CONCLUSION

IgG4-RD had potentially been categorized as Castleman's disease [4]. This disease predominantly involves the exocrine organs. Concerning its thoracic presentation, it often manifests as interstitial lung disease or fibrosing mediastinitis. The differential diagnosis of an anterior mediastinal mass includes thymoma, germ cell tumor, and mucosa-associated lymphoid tissue (MALT) lymphoma[5] . Several studies have reported that intrathoracic involvement may be seen in about 50% which commonly includes interstitial lung disease, inflammatory pseudotumors, fibrosing mediastinitis, and lymphadenopathy[6,7]. The formation of a mass in the anterior mediastinum, mimicking an anterior mediastinal tumor, is very rare [8].

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