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Atypical Manifestation of IgG-4 Related Disease as Space Occupying Lesion Mimicking Hepatocellular Carcinoma in a Renal Transplant Recipient

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Authors' contributions

This work was carried out in collaboration among all authors. All authors read and approved the final manuscript.

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Case Report

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ABSTRACT

IgG_4-related disease (IgG_4-RD) is a multisystem fibro-inflammatory condition mostly prevalent in older males. The cornerstone for the treatment of IgG_4 related disease is Corticosteroids.It rarely affects immunosuppressive population such as solid organ transplantation. Here we present to you a case of a young lady with a history of renal transplantation evaluated for symptoms of abdominal pain and weight loss and was diagnosed as a case of IgG_4 related liver disease on the basis of liver lesion biopsy.

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1. INTRODUCTION

"IgG_4-related disease is an emerging multisystem fibroinflammatory condition that can affect essentially any organ and mostly prevalent in older males" [1,2]. "As its name suggests, there is abundance of IgG_4 positive plasma cells in the affected tissues along with the elevated serum IgG_4 levels in affected individuals" [3]. "It is usually characterized as growing soft tissue lesions that can closely resemble neoplastic lesions or lymphomas" [4,5]. The cornerstone for the treatment of IgG_4 related disease is Corticosteroids. In cases of steroid refractory disease, other immunosuppressive treatment can be utilized [6]. Here we present to you a case of a forty-six years old female with a history of renal transplantation evaluated for symptoms of abdominal pain and weight loss and was diagnosed as a case of IgG_4 related liver disease.

2. CASE PRESENTATION

A 46 years old lady with a history of live related renal transplant 12 years back, maintained on immunosuppression cyclosporine was evaluated for abdominal pain and weight loss. The patient denied history of altered bowel habits, fever or jaundice. She underwent percutaneous abdominal ultrasound showing a space occupying lesion in the right lobe of liver measuring 3.4x4cm. Later on, CT Scan abdomen was performed showing an arterially enhancing lesion in segment VII of liver measures 4.2 x 5.0 x 4.5cm with washout on Porto-venous and delayed images along with few mesenteric soft tissue density areas are also noted adjacent to this lesion likely representing lymph nodal deposits with one of them measures 1.7x 1.3cm (Fig. 1).

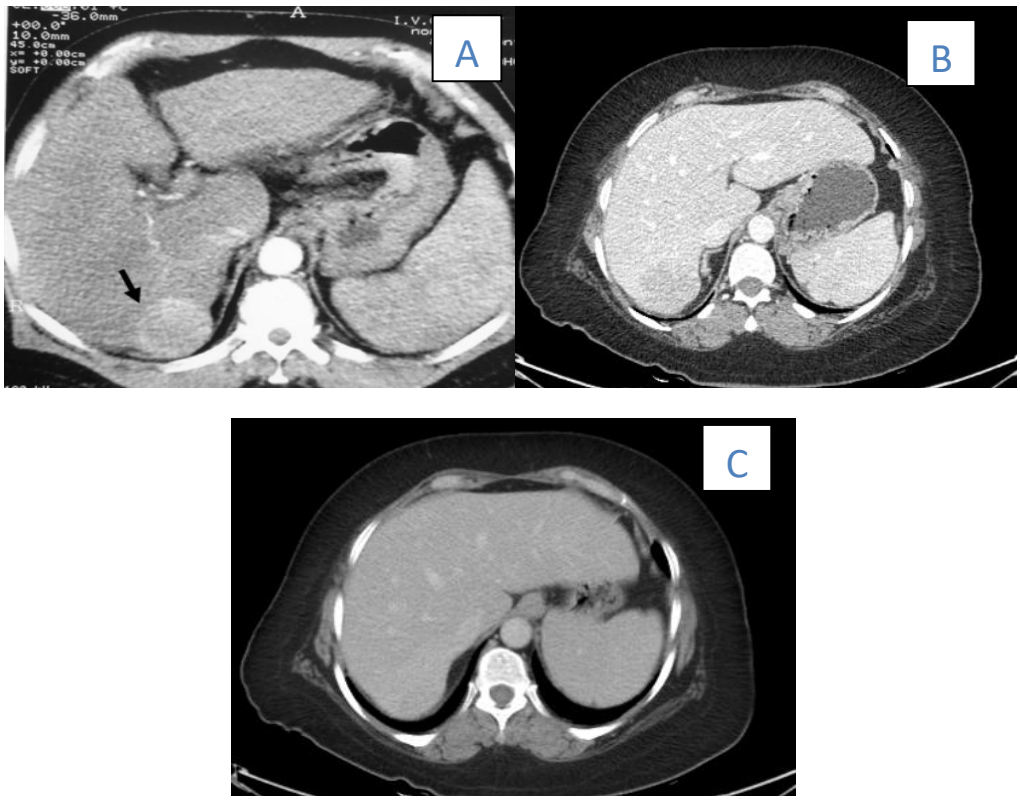

Fig. 1. CT-Scan abdomen shows lesion in segment VII of liver measures approximately 4.2 x 5.0 x 4.5 cm (black arrow) showing enhancement in the arterial phase(A) with subsequent washout in the Porto venous(B) and delayed phase (C)

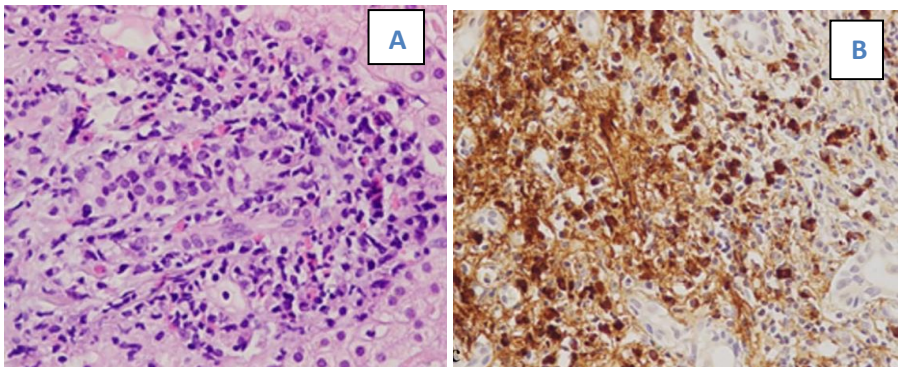

Fig. 2. Liver lesions biopsy. IA-H and E (H&E) staining showing severe expansion with sheets of plasma cells along with a few polymorphs and mature lymphoid cells. IB- IHC markers were applied and showed IgG 4 positivity in plasma cells. The morphological and IHC features are suggestive of IgG_4 related disease

Her tumor markers including CA19-9, Carcinoembryonic antigen CEA) levels and serum alpha fetoprotein were within normal limits. She subsequently underwent liver lesion biopsy to establish the diagnosis. The biopsy of the lesion showed expansion of portal tract with sheets of plasma cells along with a few polymorphs and mature lymphoid cells. Immunohistochemical (IHC) markers were applied showing Kappa and Lambda positive in almost equal proportion with IgG_4 positivity in many plasma cells. The morphological and IHC features were suggestive of IgG_4 related liver disease (Fig. 2). Her IgG_4 levels were also requested which were within normal limits. She was commenced on high dose steroids i.e.30mg/day in a tapering dose and was followed at eight weeks to document the resolution of symptoms and lesion on imaging.

3. DISCUSSION

This case of IgG_4 was a diagnostic dilemma, as both clinical presentation and imaging were suggestive of malignancy i.e. hepatocellular carcinoma. Although, there was a dynamic shift in the diagnosis when definitive tissue pathology was obtained by percutaneous liver lesion biopsy. Moreover, divergent treatment strategies between malignant and immune-mediated inflammatory processes were influential in initiating immunosuppressive agents until definite diagnosis was confirmed.

“In IgG4 –RD, there is usually a multi-organ involvement, ranging from the pancreas, kidneys, biliary tree, liver, salivary gland, orbit, aorta, to the lungs” [7]. “The liver-related manifestations of IgG4-related disorders (IgG_4 related disorders) are heterogeneous and less well described at present. They are classified into hepatic inflammatory pseudo tumor (IPT) or chronic active hepatitis that is much less prevalent than biliary involvement in IgG4-related sclerosing cholangitis. The fundamental differential diagnoses of IgG4- RD include lymphomas, hepatic abscesses and other malignancies such as hepatocellular carcinoma and metastases” [8].

Our case has rare presentation i.e. lesion in liver mimicking HCC. In such cases, early diagnosis and prompt treatment are the cornerstone of the management. “Optimal treatment regimens, dosing and duration remain ill-defined according to retrospective studies. However, early aggressive treatment is suggested to prevent serious organ dysfunction and failure in late stage disease and in the case of extensive fibrosis” [8]. The mainstay of treatment is steroid therapy; a 2- to 4-week course of prednisolone 30–40 mg per day followed by dose tapering [9]. A response is usually seen within 4–6 weeks and is determined by improvement in clinical symptoms and organ function, reduction in size of the lesions in the affected organ by radiological imaging and decrease in serum IgG4 levels. Such cases may be deceiving as IgG_4 levels can be within normal limits. Our patient also received steroids which resulted in resolution of his symptoms.

“Imaging findings of hepatic IgG_4-IPT are non-specific and tend to present with tumefactive lesions and therefore are often suspected to have malignancy. They can manifest as a single or multifocal mass. On ultrasound images, they show hypoechoic or hyperechoic masses with increase echogenicity through transmission and septation. Several patterns of enhancement are demonstrated on contrast-enhanced imaging. On MRI, these lesions are usually T1 hypointense

and T2 hyper-intense with heterogeneous enhancement" [10]. Our patient had an arterially enhancing lesion with porto-venous washout on cross-sectional imaging (CT Abdomen) which on biopsy was diagnosed as IgG_4-RD.

In solid organ transplanted patient, IgG_4-RD are rare. However, there is case report of IgG4 related lung disease in a renal transplanted recipient [11]. This is the first case of a renal transplanted recipient suffering from an IgG_4 related liver disease.

4. CONCLUSION

In conclusion, IgG4-RD with hepatic involvement is rare and has a wide variety of manifestations. Early recognition and prompt treatment with immunosuppressive therapy are important to prevent serious and irreversible tissue damage.

CONSENT

As per international standard or university standard, patient (s) written consent has been collected and preserved by the author(s).

ETHICAL APPROVAL

As per international standard or university standard written ethical approval has been collected and preserved by the author(s).

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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