Renal Cell Carcinoma Mimicking IgG4-Related Pseudotumor in Autoimmune Pancreatitis

Muhammad Ali Khan1, Sehrish Kamal1, Usman Ahmad2, Mohammed Andaleeb Chowdhury1, Ali Nawras2

1Internal Medicine and 2Gastroenterology, University of Toledo, Toledo, Ohio, USA

ABSTRACT

Context Autoimmune pancreatitis is classified into two distinct clinical profiles. Case report Type 1 autoimmune pancreatitis (AIP) is considered to be a manifestation of a novel clinicopathological entity called IgG4 related sclerosing disease, diagnosed using the Mayo Clinic HISORt criteria. Extra-pancreatic manifestations can include involvement of bile ducts, salivary gland, lung nodules, thyroiditis, tubulointerstitial nephritis, renal masses, and retroperitoneal fibrosis. Type 2 autoimmune pancreatitis on the other hand is confirmed by histologically seen duct centric pancreatitis without elevation of IgG4 or involvement of other organs. In type 1 autoimmune pancreatitis, extrapancreatic manifestations like bile duct strictures, tubulointerstitial nephritis, renal nodules, retroperitoneal fibrosis respond to steroid therapy. Conclusion We present a case of type 1 autoimmune pancreatitis in which the renal mass did not respond to steroid therapy and was later on found to be renal cell carcinoma. To the best of our knowledge this is only the third reported case of autoimmune pancreatitis in which the patient had renal cell carcinoma. Our case highlights the importance of close follow up of lesions that do not respond to steroid treatment which in this case proved to be renal cell cancer.

INTRODUCTION

Autoimmune pancreatitis is classified into two distinct clinical profiles. Type 1 autoimmune pancreatitis (AIP) is one of the presentations of IgG4 related sclerosing disease diagnosed using the Mayo Clinic HISORt criteria [1]. Extrapancreatic manifestations can include involvement of bile ducts, salivary gland, lung nodules, thyroiditis, interstitial nephritis, renal masses, and retroperitoneal fibrosis [2, 3]. Type 2 autoimmune pancreatitis on the other hand is confirmed by histologically seen duct centric pancreatitis without elevation of IgG4 or involvement of other organs.

CASE REPORT

We report a case of a seventy three year old Caucasian male who presented with obstructive jaundice, pruritus, a four month history of oily diarrhea, weight loss, and uncontrolled diabetes mellitus. Physical examination was unremarkable except for scleral icterus.

Liver function tests demonstrated a total bilirubin 16.9 mg/dL, AST 118 IU/L, ALT 213 IU/L, and alkaline phosphatase 438 IU/L.
had increased in size to 34mm x 30mm (Figure 1ab). The patient underwent partial nephrectomy and was found to have primary renal cell cancer.

**DISCUSSION**

Autoimmune pancreatitis is the prototypical manifestation of IgG4 related disease and it forms two percent of chronic pancreatitis cases [4]. Most of the early literature regarding this entity comes from Japan where the prevalence of the disease is 0.82 per 100,000 persons, however recently it has been described in several other countries.

Renal mass is one of the extra-pancreatic manifestations of IgG4 related disease which could range from tubulointerstitial nephritis to nodular lesions and pseudotumors mimicking renal cell carcinoma [5]. On radiological evaluation using contrast enhanced CT scan; the most common renal abnormality in IgG4 related disease was multiple low density lesions [6]. However, mass like lesions simulating renal cell carcinoma have been recognized in 3-27% patients [6, 7]. In the past, a number of such patients with renal masses underwent nephrectomies [8], which later revealed a dense lymphoplasmacytic infiltrate with an increased number of IgG4 positive plasma cells on pathological examination. Histologically, such renal lesions are characterized by an irregular pattern of fibrosis called storiform fibrosis which is considered to be pathognomonic for renal manifestations in IgG4 related disease [9]. Lately, CT guided renal biopsies have been performed in such cases, thereby preventing nephrectomies [10]. Being a rare clinical disorder, no formal guidelines evaluating the role of renal biopsy in such cases have been formulated (Figure 1cd).

Two case reports from Japanese literature demonstrated the occurrence of renal cell carcinoma in patients with AIP (Table 1). In the first case, renal cell carcinoma with metastases was found on autopsy, six years after the diagnosis of AIP [11]. In the second case, patient had a history of renal cell carcinoma ten years prior to the diagnosis of AIP [12]. We report a third case, in which renal cell carcinoma was diagnosed 5 months after the diagnosis of AIP. Although this may be an incidental finding, we do not know for sure whether there is a potential association between AIP and renal cell carcinoma which may be elucidated in future with more reported cases.

Our case emphasizes that not all renal masses in autoimmune pancreatitis are extra-pancreatic manifestation of the disease. It also illustrates the necessity for mandating further work up in the form of biopsy and/or surgery to rule out more ominous etiologies, especially if the renal mass is not responding to steroid treatment.

**Conflict of Interest**

Authors declare to have no conflict of interest.

**REFERENCES**


