“To Biopsy or Not” - A rare cause of renal failure

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Introduction

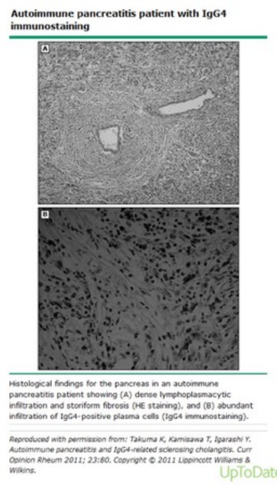
- Immunoglobulin G4-related disease (IgG4-RD) is an immune-mediated condition that can affect almost any organ and is now being recognised with increasing frequency.
- Awareness of the common features of IgG4 related disease is important as majority of patients respond to glucocorticoids.

Case Description

- A 75 years old gentleman with a background history of type 2 diabetes mellitus presented with 4 weeks history of abdominal pain and 10kg unintentional weight loss over 6 months. Physical examination was unremarkable.
- Blood investigations revealed acute kidney injury with urea 10.2, creatinine 198 and eGFR 29. CT abdomen showed a focal plaque of abnormal soft tissue in the left pelvic sidewall encasing the left common iliac vessels and bifurcation, measuring approximately 4.5x.1.5 cm, there is pelvicalceal and ureteric dilatation down to the level of this soft tissue mass which causes obstruction of the left kidney. Patient underwent left ureteric stenting.
- Further CT of the thorax and neck showed enlarged precarinal and right hilar lymph nodes measuring 14 and 16mm in diameter respectively. Pet scan showed increase uptake corresponding to the soft tissue mass in the left side of the pelvis.
- Differential diagnoses of lymphoma and retroperitoneal sarcoma were made after further discussions with other specialties.
- Because of the pelvic mass proximity to vascular structures and concerns about tumour seeding, subcutaneous image guided biopsy was deemed not feasible. Patient subsequently underwent an open biopsy at Royal Marsden Hospital which was complicated with bleeding post biopsy which he required a repeat operation. Fortunately, he recovered well.
- Histology appearances of the biopsy raised strong suspicion of IgG4-related sclerosing disease (IgG4-RD), with no evidence of lymphoproliferative disease.
- Following the biopsy, patient was started on high dose steroids. A follow up CT scan two weeks after commencement of steroids showed a significant reduction of the size of the lesion.
- IgG4 level was raised at 3.63(0-1.3), further supporting the diagnosis of IgG4-RD.
- Glycemic control was an issue whilst patient was on high dose steroids and insulin regime was intensified.
- CT scan 5 months after steroids showed a complete resolution of the pelvis mass and left hydronephrosis. Patient’s glycemic control improved with tapering of steroids. Creatinine is stable at ~170.

Discussion

- IgG4-RD was not recognised as a systemic condition until 2003[1]
- Common presentations include autoimmune pancreatitis, salivary gland diseases (previously named as Mikulicz syndrome), orbital pseudo tumours, retroperitoneal fibrosis, and IgG4-related hypophysitis.
- The commonly shared features include tumour-like swelling of involved organs, a lymphoplasmacytic infiltrate enriched in IgG4-positive plasma cells, and a variable degree of fibrosis that has a characteristic "storiform" pattern[2].
- Histopathological analysis of biopsy specimens remains the cornerstone in the diagnosis of IgG4-RD.
- Elevated IgG4 in tissue and serum are helpful in diagnosing IgG4-RD but neither one is a specific diagnostic marker. Elevated serum IgG4 are found in 60-70% of patient with IgG4-RD[3].
- IgG4-RD can often cause major tissue damage and can lead to organ failure.
- Aggressive treatment is needed when vital organs are involved. Glucocorticoids are typically the first line therapy[3].



Histological findings for the pancreas in an autoimmune pancreatitis patient showing (A) dense lymphoplasmacytic infiltration and storiform fibrosis (HE staining), and (B) abundant infiltration of IgG4-positive plasma cells (IgG4 immunostaining).

Learning Points

1)IgG4-RD can manifest as retroperitoneal mass causing obstructive uropathy. It mimics renal cell carcinoma or sarcoma.

2) It is important to biopsy the mass itself rather than peripheral lymph nodes as IgG4 in lymph nodes can be non specific.

3)Awareness of the various presentations of IgG4-RD need to be heightened as early disease responds very well to steroids.

4)Surveillance of other organs e.g. pancreas, pituitary, lungs for manifestations of IgG4-RD is important as IgG4-RD can present in multiple organs.

References

1)Kamisawa, T., Funata, N., Hayashi, Y. et al. A new clinicopathological entity of IgG4-related autoimmune disease. J Gastroenterol (2003) 38: 982.
2) UpToDate, Overview of IgG4-related disease.
3) John H.Stone et al. IgG4-Related Disease Review Article. N Engl J Med 2012;366:539-51.