An Unusual Cause of Central Diabetes Insipidus

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INTRODUCTION

Central diabetes insipidus (CDI) results from a deficient secretion of osmoregulated vasopressin. In most cases it is idiopathic or it can be induced by tumor, pituitary surgery, cranial trauma or infiltrative diseases. CDI can be isolated or associated to other pituitary hormone deficits. Clinical, biological and radiological follow-up is crucial given that idiopathic CDI can be the earliest sign of an evolving process (inflammatory or tumoral). We report the case of an unusual cause of isolated CDI.

CASE REPORT

A 23-year-old woman initially presented with polydipsia and polyuria. The diagnosis of CDI was established by a water deprivation test and a treatment with intranasal desmopressin was started. Circulating levels of anterior pituitary hormones were normal. Magnetic resonance imaging of the pituitary gland showed thickening of the pituitary stalk and loss of the normal hyperintense signal of the posterior pituitary on T1-weighted images (Fig.1).

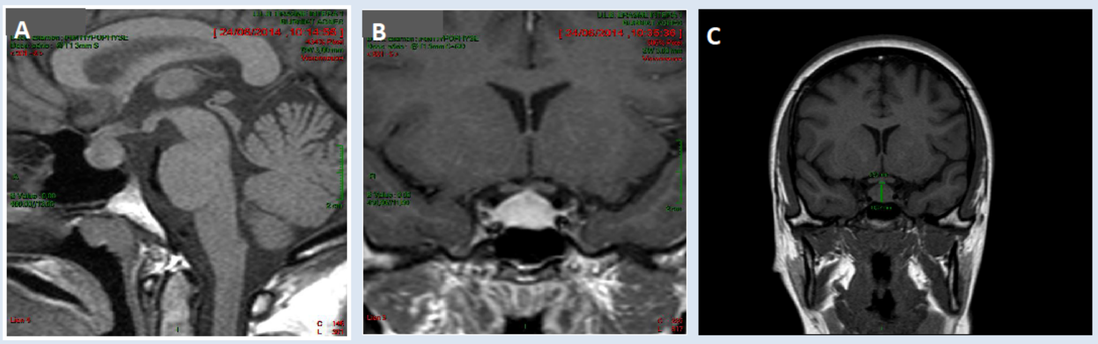


Fig.1. **A.** Unenhanced T1-weighted sagittal image: absence of the "bright spot" of the posterior pituitary.
B. Post-gadolinium T1-weighted coronal image: homogeneous enhancement of the pituitary gland.
C. T1-weighted coronal image: enlarged pituitary gland (12.8 mm) with thickening of pituitary stalk (3.63mm).

Alpha fetoprotein and beta human chorionic gonadotropin concentrations were normal in serum and spinal fluid, as were
C-reactive protein and angiotensin-converting enzyme plasmatic levels. Chest X-ray showed no pulmonary involvement. A ^{18}F-FDG PET/CT revealed a tumor-like lesion in the right kidney which was confirmed by a renal contrast-enhanced ultrasonography (CEUS). Granulomatous with polyangiitis (GPA) was suspected on the basis of associated maxillary sinus hypermetabolism on ^{18}F-FDG PET/CT and plasmatic anti-MPO ANCA positivity. Strikingly, urinalysis did not reveal proteinuria or hematuria and renal function was normal. Moreover, sinus biopsy did not show typical granulomatous inflammation. Ultrasound control performed 6 months later showed that the right renal mass had doubled in diameter (from 2.2 to 4.3 cm) (Fig. 2) and suspected new lesions in the left kidney. This was confirmed by ^{18}F-FDG PET/CT (Fig. 3). Renal function and urinary sediment still remained normal.

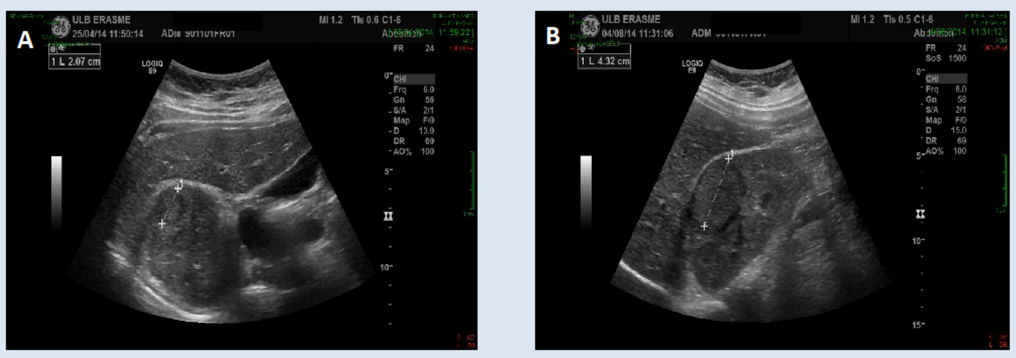


Fig.2. CEUS images of the right kidney : **A.** 2.2 cm mass (March 2014); **B.** 4.3 cm mass (September 2014).

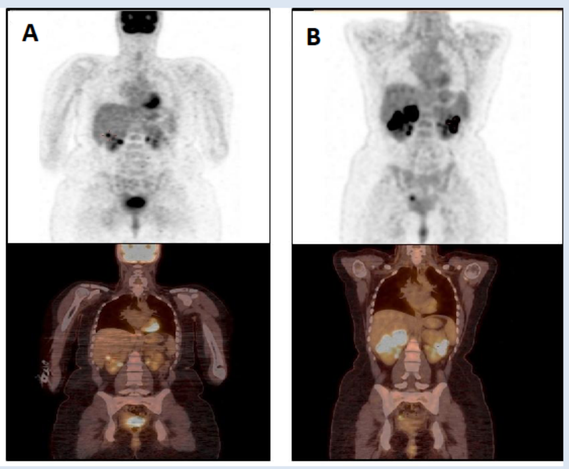
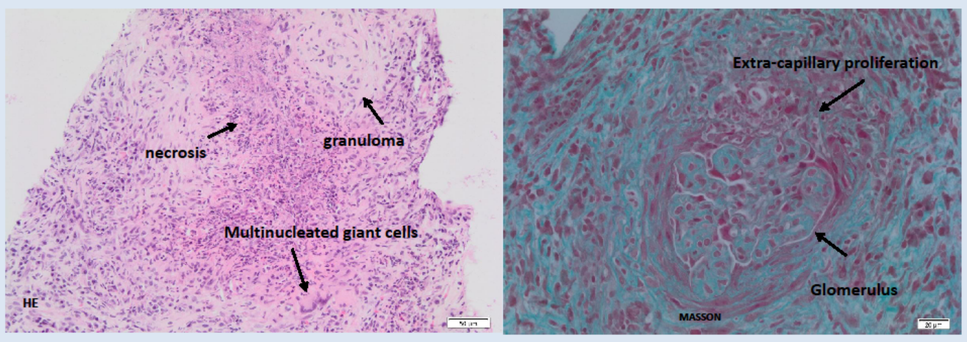


Fig.3. 18F-FDG PET/CT: **A.** Hypermetabolic lesion in the right kidney (March 2014); **B.** Expansion of the lesion in the right kidney and appearance of new lesions in the left kidney (September 2014).

CASE REPORT

Histopathological examination of the renal biopsy specimen revealed necrotizing granulomatous tubulointerstitial nephritis with multinucleated giant cells and extra-capillary glomerulonephritis, which was consistent with the diagnosis of GPA.



In order to avoid cyclophosphamide-induced ovarian failure, treatment with rituximab (375 mg/m^2 once weekly during 4 weeks) and prednisolone was initiated, resulting 6 weeks later in disappearance of renal tumors in ultrasound control.
Unfortunately, central DI persisted.

DISCUSSION

Granulomatosis with Polyangiitis (GPA) is an antineutrophil cytoplasmic antibody (ANCA)-associated systemic vasculitis of small and medium-sized vessels which classically affects the upper and lower respiratory tracts and kidneys. Pituitary involvement is a rare complication of the disease with only 30 previous case reports in the English literature. When it occurs, CDI usually follows rather than precedes lung and kidney involvement.
Renal impairment is usually characterized by segmental necrotizing glomerulonephritis with hematuria and/or proteinuria that often leads to rapidly progressive renal failure. The presence of a granulomatous renal pseudotumor is rare; only 16 cases have been reported.
The case described here is unique in many aspects.
It is the first report of GPA combining initially a CDI and subsequently bilateral renal pseudotumors.
Even if ^{18}F-FDG PET/CT is not usually recommended for the etiological exploration of CDI, it was decisive for final diagnosis in this particular case.
In addition, the pseudotumoral lesions have exceptionally been described by ^{18}F-FDG PET/CT and never before by CEUS.
Finally, whereas most of the cases of GPA-associated renal pseudotumors have been treated by surgery or less frequently by classical medical treatment (cyclophosphamide and steroids), our patient experienced rapid and complete renal response to rituximab. The persistence of CDI could be interpreted as irreversible damage, although the delay from the beginning of treatment is rather short.

CONCLUSION

In conclusion, GPA should be considered in the differential diagnosis of CDI on the one hand and of renal mass-lesions on the other hand. In particular, rapid recognition of GPA as the cause of CDI and rapid initiation of treatment could minimize the risk of irreversible pituitary function loss.

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