Case Report

Necrotizing infundibulohypophysitis: A rare cause of central diabetes insipidus

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ABSTRACT

Diagnosis of central diabetes insipidus (CDI) requires a high index of clinical suspicion, especially when it manifests as a coexisting condition like traumatic brain injury or following neurosurgery. We would like to report a rare case of necrotizing infundibulohypophysitis (NIH) as a cause of CDI in a 21-year-young male who presented with a severe progressive headache not responding to routine analgesics followed by vomiting, altered sensorium. His baseline blood investigations were normal including his cerebrospinal fluid examination, plain and contrast magnetic resonance imaging (MRI) scan. The patient partially responded to the treatment of headache and after 2 days, complained of polyuria with severely reduced serum sodium level. A repeat plain and MRI of the brain was done which showed classical findings of NIH with CDI which we discuss along with the differential diagnosis and its prognosis.

Key words: Central diabetes insipidus, Magnetic resonance imaging, Necrotizing infundibulohypophysitis

Central diabetes insipidus (CDI) is commonly seen after traumatic brain injury, postpartum state, neurosurgery, stroke, or infections [1]. The disease requires a high index of clinical suspicion, especially when it manifests as a coexisting condition.

We describe a rare case of CDI in a young male with necrotizing infundibulohypophysitis (NIH) presenting with acute CDI. Less than 10 patients of NIH have been described in the literature so far as a rare cause [2,3].

CASE REPORT

A 21-year-old male presented with a progressive severe diffuse headache for 3 days which was not relieved by routine analgesics. This was followed by vomiting, retro-orbital pain, and tonic–clonic seizures.

Clinical examination revealed E4, M4, and B4 status on Glasgow coma score (GCS) score. Plantar reflexes were flexor and there was no sensory or motor deficit.

Routine blood investigations were done, that is, complete blood count, liver and renal function tests, random and fasting blood sugar, serum electrolytes, thyroid profile, and C-reactive protein which were all normal. A plain and contrast-enhanced magnetic resonance imaging (MRI) study of the brain was done and was also normal. The sella also showed normal size and signal of the pituitary (Fig. 1a and b). Following a normal MRI, cerebrospinal fluid examination was done by a lumbar puncture which showed 12 mg/dl proteins, 4.5 mg/dl globulins, sugar was 75 mg/dl, and it was acellular. Later, culture results also showed no bacterial growth. Fundus examination showed no papilledema.

A clinical diagnosis of migraine was made and the patient was given a combination of 250 mg aspirin and 65 mg caffeine thrice a day along with ibuprofen 200 mg twice a day orally, sumatriptan 20 mg once a day, and3 metoprolol 25 mg daily along with 400 mg of oral Eptoin for seizure control.

After 5 days, there was a partial relief in headache but his GCS status was unchanged. The patient then started complaining of polyuria. All baseline tests were repeated which were normal except serum sodium which was 115 meq/L. A repeat plain and contrast MRI study revealed enlarged pituitary gland and pituitary stalk with a contrast study showing non-enhancing central necrosis with a thickened enhancing stalk of the pituitary gland (Fig. 2a and c). His blood cortisol level, thyrotrophin/thyroid-stimulating hormone, growth hormone, and prolactin levels done were all severely reduced.

The patient was prescribed desmopressin nasal spray twice a day, hydrocortisone 20 mg/day, and thyroxine 25 microgram/day. The patient started improving and was asymptomatic after 1 week of treatment. No surgical intervention was done and the patient was discharged and advised regular follow-up.

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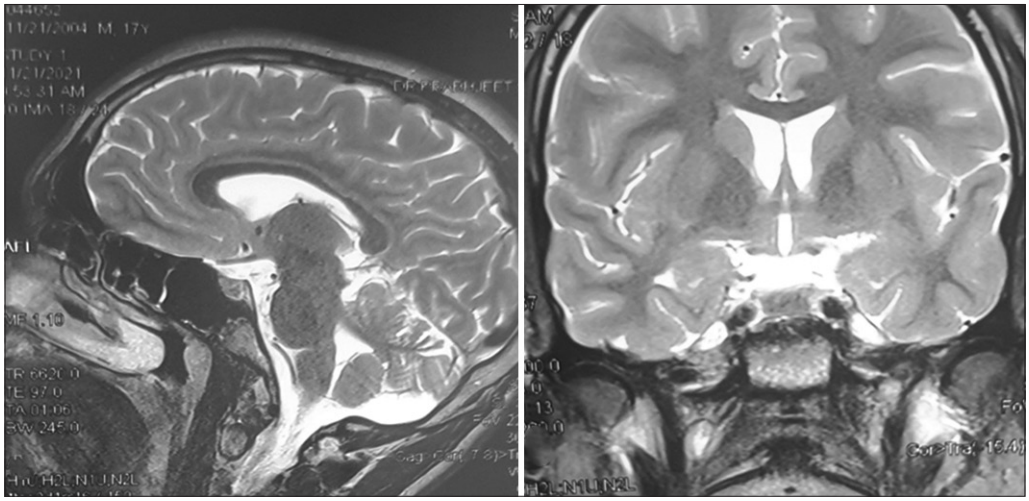

Figure 1: Plain T2W sagittal and coronal magnetic resonance imaging of the brain showing normal size and signal of the pituitary gland

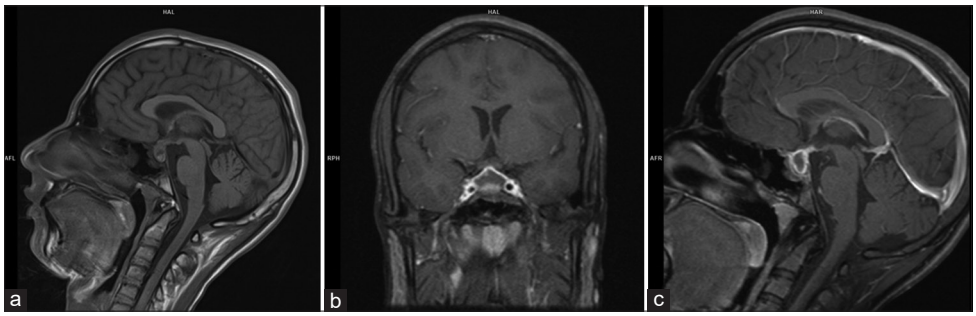

Figure 2: (a) Plain T1W sagittal magnetic resonance imaging (MRI) showing enlarged heterogeneous signal of pituitary gland with peripheral enhancing rim; (b) contrast coronal T1W MR showing enlarged necrotic pituitary gland; (c) contrast sagittal T1W MRI of brain with enhancing thickened pituitary stalk

DISCUSSION

NIH is a rare condition with less than 10 cases described in the literature so far [2,3]. It is one of the rare causes of CDI and presents as an acute condition like seen in Sheehan syndrome with acute headache, vomiting, and altered sensorium with or without seizures.

Early diagnosis is challenging as seen in this case since most of the initial investigations including clinical examination are unremarkable. MRI shows a triad of the enlarged pituitary gland, pituitary necrosis, and enlarged infundibulum. The condition mimics lymphocytic hypophysitis, pituitary macroadenoma, and granulomatous infections of the pituitary, however, in all these conditions, the presentation is chronic and never acute. Further in lymphocytic hypophysitis and macroadenomas, there is sparing of the stalk of the pituitary which can also be a differentiating feature on imaging. Other conditions in which pituitary necrosis can be seen are gestational diabetes, spinal anesthesia, and snake poisoning [4-6]. In one autopsy series, pituitary necrosis was seen in 10% of cases [7]. However, in all the above cases, the necrosis seen is usually patchy and never completes [8]. Pathologically, lymphocytic infiltration is seen in the pituitary in NIH with central necrosis suggestive of an acute necrotizing immune phenomenon. The reactive enhancing rim may be seen at the end of the 1 week on contrast MRI in such cases as seen in pituitary macroadenomas and apoplexy suggestive of a reparative cell reaction [9]. This rim is never seen in cases of lymphocytic hypophysitis due to its chronic autoimmune nature. The condition does not carry a good prognosis and most of the patients described were dependent on replacement hormonal therapy.

The exact cause for this is not clear but it is hypothesized that necrosis in the pituitary excites the production of autoantibodies resulting in ongoing necrosis with failure of the pituitary to return to its normal function [10]; the involvement of stalk of pituitary occurs as a secondary phenomenon due to spread of inflammation.

CONCLUSION

NIH is a rare condition and poses a diagnostic challenge. Serial MRI is helpful in making the diagnosis and shows a triad of necrotic enlarged pituitary mass with enlarged infundibulum with enhancing rim. The condition must be differentiated from pituitary macroadenoma so that surgical intervention can be avoided and clinical history and pituitary function blood tests must also be combined with the MRI findings to establish the final diagnosis.

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