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Hypothalamic-pituitary disconnection: A diagnostic challenge in the middle skull base region

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Abstract

Infections gaining access to the skull base regions are particular challenging to diagnose, especially in the elderly people. We report the case of a 71-year old woman presenting with fever, impaired general condition and hypotension. MRI imaging techniques showed a tuberculoma in the sellar region, suggesting an extremely rare form of central nervous system tuberculosis.

Keywords

tuberculosis; middle cranial fossa; pituitary; skull base medicine

Introduction

The pituitary stalk, anatomic connection between the hypothalamus and the pituitary gland is involved in several endocrine processes such as the production of thyroid hormones, cortisol, oestrogens and prolactin [1]. If these two structures are disconnected because of a pathological process, the affected individual may present severe dysfunctions; however, such situations may be difficult to diagnose. We hereby report a severe and complicated case of an elderly woman.

Case Presentation

A 71-year old woman presented at the emergency department with an impaired general condition, hypotension and fever. She was referred from her nursing home, where the nurses reported a fever and a 77/53 mmHg hypotension, as well as an unspecified weight loss.

At the emergency department, although she was foreign and presented with a complete language barrier, her Glasgow coma score was 15/15; she also presented with head and upper limb tremor bilaterally, abnormal gait, limb ataxia and increased muscle tone. From her electronic health record, she supposedly suffered from tuberculosis 5 years ago. Blood pressure at the emergency department was 94/67 mmHg. Clinical assessment of vision was unremarkable.

The biology reported a hypokalaemia (3.3 mmol/L), a hypocalcaemia (2.03 mmol/L) and hypophosphatemia (0.77 mmol/L), normocytic anaemia (9.1 g/dL). A sputum was realized, which was negative.

A brain CT-scan was realized at the emergency department (Figure 1) which showed a pituitary mass (13 mm x10 mm). A brain MRI was also realized (Figure 2) which confirmed the mass, and a brain diffusion MRI (Figure 3) showed dense regions in the hemispheres, suggesting a meningoencephalitis. The patient was hospitalized and osmolality as well as endocrine functions were investigated in a supplementary biology two days later; the biology reported a hypoosmolality (155 mOsm/kg), low TSH, T3 and T4 levels (0.21 mU/mL, 9.6 and 2.11 pmol/L respectively), and very high prolactin level (74.7 µg/L). A lumbar punction reported lymphocytes 89%, monocytes 11%, and hypoglycorrhachia (glucose 33.4 mg/dL); a PCR was done on the cerebrospinal fluid collected that reported mycobacterium tuberculosis; a diagnosis of panhypopituitarism and hypophysitis secondary to tuberculous meningoencephalitis was made. The patient was treated with thyroid replacement therapy for secondary hypothyroidism, and vasopressin replacement therapy for secondary diabetes insipidus, as well as an intensive antituberculous treatment (isoniazid 5mg/kg; rifampin 10 mg/kg; ethambutol 20mg/kg; pyrazinamide) for tuberculous meningoencephalitis for a duration of two months, followed of an additional treatment (isoniazid 5 mg/kg and rifampin 10 mg/kg) for a duration of 18 months. The neurological deficit improved, and the patient returned to her home country and was lost to follow up.

Discussion

Central nervous system manifestation of tuberculosis account for less than 1% of cases worldwide; it is a devastating complication with high mortality [2].

Involvement of the skull base region is a rare finding associated with the central nervous system manifestation of tuberculosis. In our case, the tuberculoma detected in the pituitary gland and invading the pituitary stalk, which is a far rarer finding [3] and caused in our case a severe panhypopituitarism, characterized by diabetes insipidus because of lack of vasopressin; hypothyroidism because of lack of TSH; hyperprolactinaemia because of lack of inhibitory signal from the hypothalamus.

Diagnosis can be complicated, especially in elderly patients. MRI is the gold standard imaging modality for diagnosis of cerebral manifestations of tuberculosis and shows hypodense regions in T1-weighted images [3].

However, gold standard measures for diagnosing tuberculosis, such as sputum and PCR must also be considered for a microbiological confirmation of the pathology [4].

A skull base surgical approach may be needed in case of raised intracranial pressure in the patient, which was not present in our case [3].

Future studies may endeavour to evaluate the symptomatology of tuberculous hypophysitis as well as management in elderly patients.

Figures

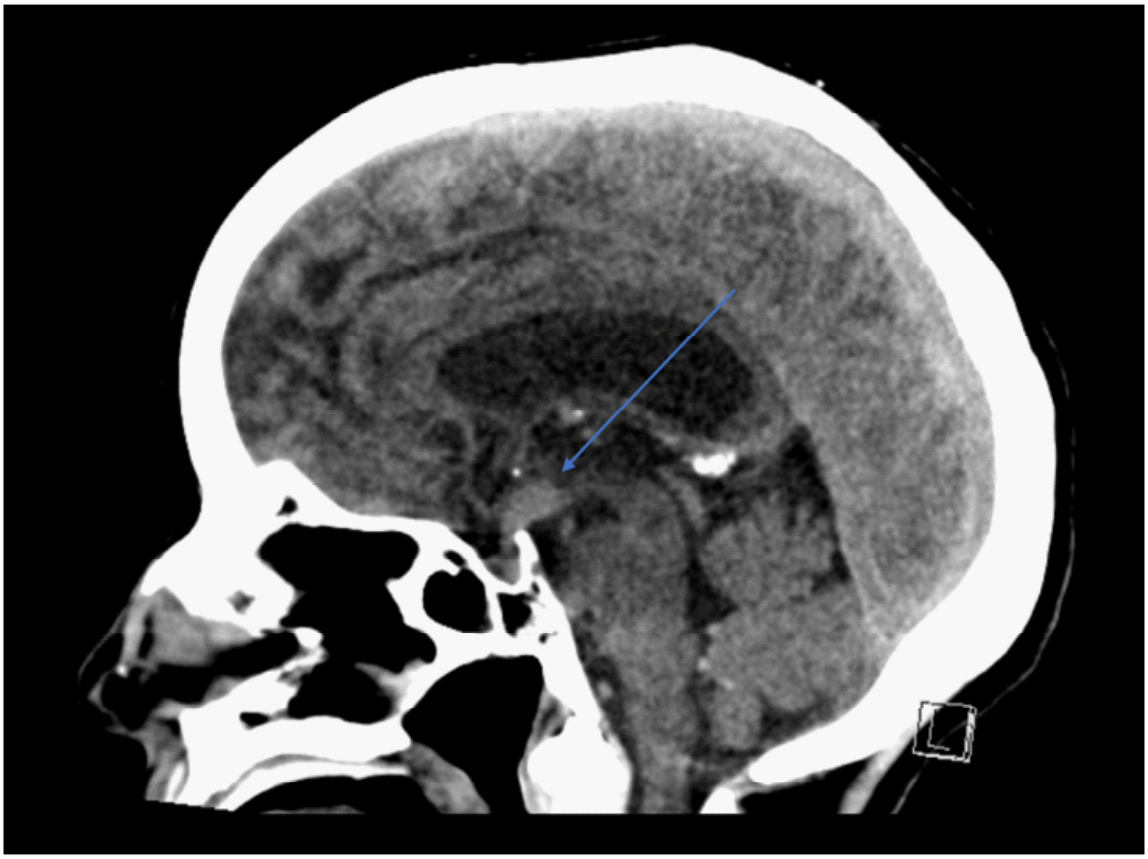

Figure 1: Brain CT-scan showing a pituitary mass.

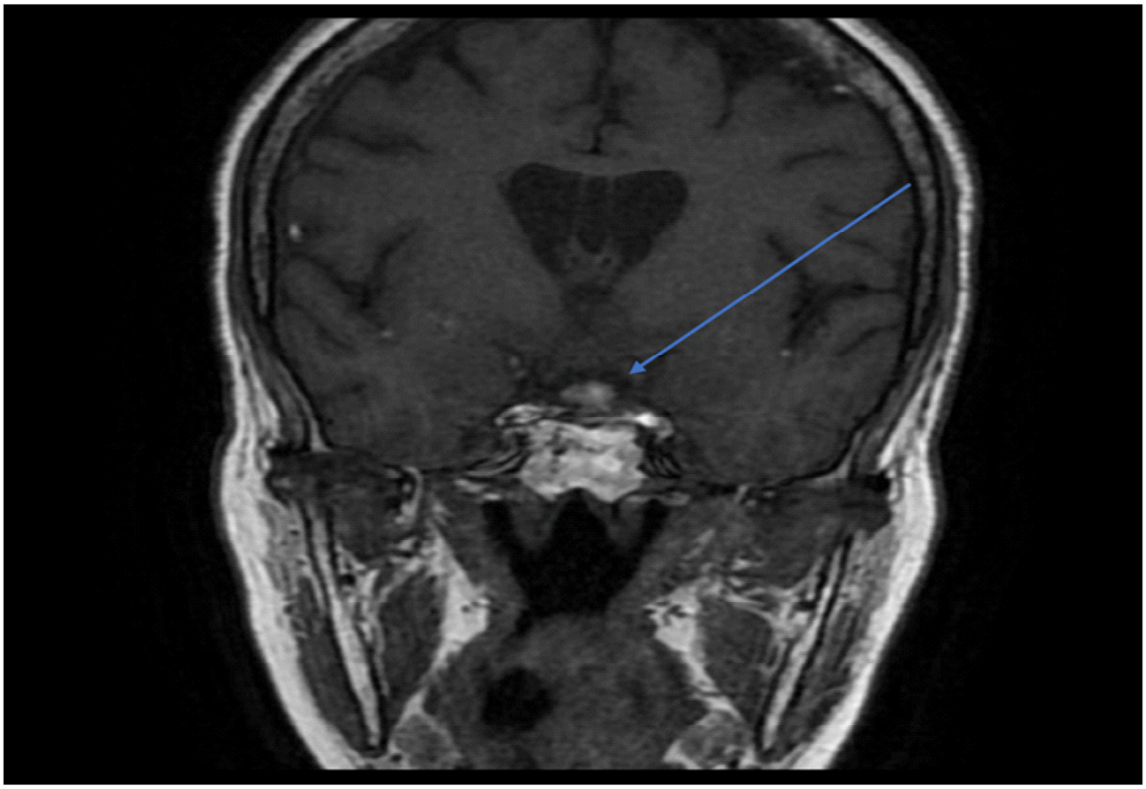

Figure 2: T1-weighted brain MRI reporting a pituitary mass as a hypodense region.

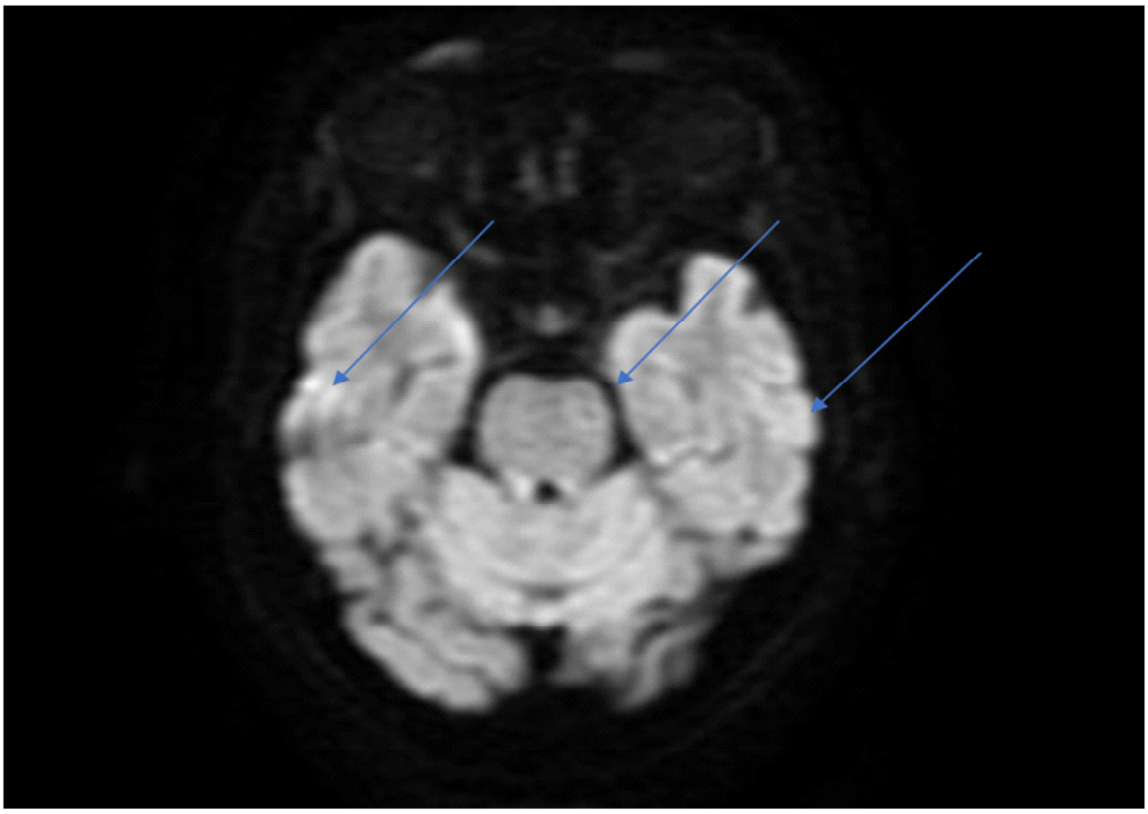

Figure 3: Diffusion brain MRI showing dense regions, suggesting a diffuse meningoencephalitis.

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