CASE REPORT

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Neurobrucellosis Presenting as Pseudotumor Cerebri: First Report from Oman

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ABSTRACT

A ten-year-old boy presented to the hospital with body ache and joint pains for two months and headache, vomiting, and skin rash for three days. He was drowsy and lethargic at admission. Physical examination revealed bilateral papilledema. There were no cranial nerve involvement, neuromotor deficit, or signs of meningeal irritation. Computed tomography and magnetic resonance imaging of the brain did not reveal any evidence of cerebral edema or space occupying lesion. In view of the high endemicity of brucellosis in the area, Brucella agglutination test (BAT) was done at the time of admission and was negative. However, on the eighth day of admission, blood culture showed growth of Brucella melitensis. A repeat BAT at this time was strongly positive with a titer of 1:1 280. The initial one was negative due to prozone phenomenon caused by very high antibody titers. A diagnosis of neurobrucellosis with pseudotumor cerebri was made. He was treated with gentamicin, rifampicin, and septrin along with acetazolamide for raised intracranial pressure. The boy responded well to therapy and made a complete clinical recovery with resolution of papilledema. In areas endemic for brucellosis, a high index of suspicion for neurobrucellosis should be entertained in any child presenting with diverse neurological signs.

Brucellosis is an important zoonotic disease in Oman.[1] It is mostly confined to the Governorate of Dhofar due to the presence of infected camels, cattle, goats, and sheep.[2] The transmission of infection to humans is through close contact with animals and consumption of raw milk.[3] Neurological manifestations are uncommon in brucellosis, and such symptoms in children are rarer still.[4] Neurobrucellosis usually presents as meningoencephalitis. Brucellosis presenting as pseudotumor cerebri is very rare.[5] Here, we report the first case of neurobrucellosis from Oman who presented with pseudotumor cerebri.

CASE REPORT

A ten-year-old boy was admitted to the pediatric ward with complaints of body ache and joint pains for two months and fever for three weeks. His condition worsened with the onset of headache and vomiting three days before admission. There was no history of cough, bowel/urinary complaints, or seizures. There was no history of recent travel. His development was normal with good school performance. There was a history of ingestion of raw camel milk from early childhood. His brother had been treated for brucellosis (manifested as polyarthralgia) two months prior.

At admission, he was febrile (39.4 °C). He had a generalized erythematous maculopapular rash. There was no pallor, icterus, lymphadenopathy, or throat congestion. His abdomen was soft without hepatosplenomegaly. His joints were normal.

The boy was drowsy, lethargic, and partially responsive to verbal commands. He had bilateral papilledema, which was confirmed by two ophthalmologists from separate institutes. There was no involvement of cranial nerves or sensorimotor deficit. There were no signs of meningeal irritation. A provisional diagnosis of brucellosis with papilledema was made.

Investigations showed hemoglobin levels of 12.5 g/dL, a total leucocyte count of 5.6×10^9/L (neutrophils 65%, lymphocytes 29%) and platelets of 250×10^9/L. His C-reactive protein level was 63 mg/L with mildly elevated alanine aminotransferase (89 IU/mL) and aspartate aminotransferase (64 IU/mL). His urea, electrolyte, and glucose levels were normal. His Brucella agglutination titers were negative. Chest X-ray, ultrasound abdomen, cranial computed tomography (CT) and brain magnetic resonance imaging (MRI) were normal. His parents

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refused lumbar puncture for cerebrospinal fluid (CSF) examination.

A differential diagnosis of autoimmune disease, prolonged viral illness or intracranial infection was entertained. Investigations were negative for antinuclear antibodies and rheumatoid factor. Tests for Monospot, Widal, cytomegalovirus, and Epstein-Barr virus were all negative. Treatment was initiated with ceftriaxone, acetazolamide, and supportive care.

On the eighth day of admission, blood culture showed pure growth of Brucella melitensis. A repeat Brucella agglutination was strongly positive with 2-mercaptoethanol (ME) titer > 1:1 280. A final diagnosis of neurobrucellosis with pseudotumor cerebri was established.

The patient was treated as per Saudi Paediatric Infectious Diseases Society (SPIDS) protocol with gentamicin (6 mg/kg/day intravenous infusion once daily) for two weeks and oral doxycycline (100 mg twice daily), rifampicin (15 mg/kg/day in two divided doses), and trimethoprim-sulfamethoxazole (10 mg/kg/day in two divided doses) for six months.[6] Acetazolamide was administered for the first six weeks. The patient steadily improved and was discharged after 33 days of hospital stay with full neurological recovery. The papilledema resolved completely in two months. On follow-up, the boy was doing well and attending school. Repeat Brucella agglutination titer at three months fell to 1:160 and turned negative after nine months.

DISCUSSION

In Oman, brucellosis is mostly confined to the Governorate of Dhofar, where the incidence has been reported as 6.8 per 10 000 persons.[1] Presentation with neurological manifestations in brucellosis is rare. A study of 375 cases of brucellosis in children from Dhofar, failed to show a single case.[3] Other studies have found the incidence of neurobrucellosis amongst children infected with Brucella to be 0.8%.[7,8] Yetkin et al,[9] reported that patients with neurobrucellosis had a higher incidence of headache (70%) and nausea/vomiting (30%) compared to those without neurobrucellosis (31% and 11%, respectively). Generalized maculopapular rash was another unusual clinical sign observed in our patient. Dermatological manifestations in brucellosis are rare and reported in less than 5% of cases.[10]

A review of 187 cases of neurobrucellosis contained in 35 publications revealed that most presented with meningeal irritation (37%), with the remaining having cranial nerve involvement, radiculoneuropathy, stroke, central nervous system abscess, and paraplegia/hemiplegia.[11] Presentation of neurobrucellosis with raised intracranial pressure in children and adolescents is uncommon.[12] Yilmaz et al,[8] and Sinopidis et al,[13] both reported cases of children with headaches, vomiting, diplopia, and papilledema. Similarly, Akhondian et al,[14] reported an 11-year-old girl from Iran presenting with pseudotumor cerebri and recurrent transient ischemic attacks due to neurobrucellosis. Single cases of children presenting with pseudotumor cerebri due to neurobrucellosis have also been reported by Emadoleslamia[15] and Panagariya.[16]

The presence of neurological features in blood culture positive brucellosis is enough to diagnose neurobrucellosis and evaluation of CSF is not mandatory.[17] In a large study conducted by Guven et al,[17] it was reported that CSF culture was positive in only 15% cases and approximately half (58%) had leucocytosis and raised protein in the CSF. The same study also found that only 27% of patients diagnosed with neurobrucellosis had findings consistent with the diagnosis in cranial MRI and/or CT. Pseudotumor cerebri was confirmed in our patient by the presence of papilledema with normal ventricular size in cranial MRI.

Brucella agglutination test was negative at the time of presentation. It turned strongly positive when performed at a higher dilution a week later. This is due to the prozone phenomenon, where the high level of antibodies prevents agglutination of the antigen by acting as blocking antibodies resulting in a false-negative test.

We used the protocol endorsed by the SPIDS for treating neurobrucellosis.[6] Our patient received gentamicin for the first two weeks along with doxycycline, rifampicin, and trimethoprim-sulfamethoxazole for six months. For children below the age of eight, the protocol recommends that doxycycline be replaced by ciprofloxacin.

The prognosis in neurobrucellosis is generally good if appropriate treatment is started early. Mortality rates have been reported between 0–27% in neurobrucellosis.[18,19] Sequelae occur mostly in cases where therapy had been delayed.

CONCLUSION

Neurobrucellosis should be considered in any patient in an endemic area presenting with prolonged illness and associated neurological symptoms. The prozone phenomenon should be kept in mind when the clinical suspicion of brucellosis is high and the agglutination test is negative. Appropriate treatment should be administered for six months for neurobrucellosis to prevent sequelae and ensure full recovery.

Disclosure
The authors declared no conflicts of interest.

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