Neurobrucellosis with bilateral sensorineural hearing loss and ataxia

A case report

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Summary

Brucellosis is a zoonotic infection and has endemic characteristics. Neurobrucellosis is an uncommon complication of this infection. In neurobrucellosis, various clinical and neuroradiological signs and symptoms can be confused with other neurological diseases. In inhabitants or visitors of endemic areas, neurobrucellosis should be kept in mind in cases which have unusual neurological manifestations. We present a case of a 50-year-old female who presented with headache, bilateral hearing loss for 1 year with ataxia and vertigo for 2 months. The case is reported because of the diagnostic dilemma and its rarity.

Introduction

Brucellosis, caused by various strains of Brucella, is acquired by consuming unpasteurised dairy products or by close contact with cattle [1, 2]. However, a history of exposure to cattle may not be available and in that case the route of infection is via aerosol inhalation [3]. Most of the patients with systemic brucellosis present with pyrexia of unknown origin. About 5% have predominant central nervous system (CNS) involvement [3, 4]. Here we present a patient with systemic brucellosis complicated by chronic meningitis.

Case report

A 50-year-old female presented with a history of headache and progressive hearing loss for 1 year. She was managed with analgesics, and later she presented with progressive vertigo in all positions and instability of gait which had been occurring for 2 months and she was bed-ridden for a week before admission. She had been married for 25 years with unevaluated primary infertility and had been postmenopausal for 1 year. She also gave a history of neck pain, vomiting on and off, paraesthesias of both upper and lower limbs, and constipation. There was also history of generalised body aches, arthralgias, fatigue and anorexia with weight loss. After a few days she also developed difficulty in passing urine. She came from a farming family who reared cattle.

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On examination she had pallor with pulse 82/minute, and blood pressure of 120/80 mm Hg when lying down, 110/76 mm Hg when seated, and 110/70 mm Hg when standing. Her temperature was 99 °F. A chest and cardiovascular examination was normal. The abdominal examination revealed a palpable liver 3 cm below the costal margin on the right side, which was smooth and non-tender. Neurological examination revealed higher mental functions were normal and an ophthalmology examination was normal. There was neck stiffness and the Brudzinski test was positive. Cranial nerve (CN) VIII showed sensorineural bilateral hearing loss with left > right (on audiometry) and CN IX, X , XI were also impaired on the left side with impaired gag reflex and less movement of uvula and soft palate on the same side. Motor examination revealed decreased bulk of muscles in both upper and lower limbs, increased tone bilateral lower limbs, and a power of Grade IV all over. Reflexes were exaggerated with ill sustained clonus on the left lower limb with plantars bilateral extensor type. The sensory system [touch, temperature, vibration, pressure] was intact with mild impairment in position sense in lower limbs. Gait was grossly ataxic with truncal ataxia as well. Cerebellar signs were impaired bilaterally showing dysdiadokinesia, impaired finger-nose test, and heel-shin test.

Investigations revealed mild anaemia with haemoglobin 9 g% and ESR 40. The rest of the investigations involving liver and renal functions, electrolytes, muscle enzymes, and the Mantoux test were negative. A cerebrospinal fluid (CSF) examination revealed: clear fluid, proteins 40 mg/dl, glucose 20 mg/dl, total cell count 260/mm^3 with polymorphs 23% and lymphocytes 73%, Adenosine Deaminase (ADA) 14.3 IU/ml, CSF Gram stain (no organism seen), CSF fungal stain negative, CSF polymerase chain reaction (PCR) for mycobacterium tuberculosis was negative, CSF Brucella agglutination titer 1:320, and CSF culture negative. Serum brucella agglutination titer was 1:160 with culture negative (CSF lactate and isoelectric focusing was not done). Contrast-enhanced magnetic resonance imaging of the brain showed a hyper-intense periventricular signal with patchy subcortical hyperintense foci seen in parietal lobe on T2 and Flair images with hyperintense signal intensity in post limb of internal capsule (fig. 1).

Based on these features and with laboratory support of brucella agglutination titer, a strong suspicion of neurobrucellosis was made and patient was put on rifampicin, doxycycline and ceftriaxone. The patient reported improvement during the hospital stay within 2 weeks with repeat CSF showing clear fluid, protein at 27 mg/dl and cell count at 92/mm^3 with lymphocytes at 93%. The patient was

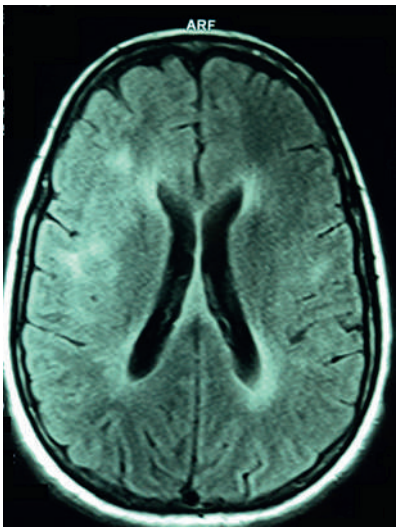

Figure 1
Contrast-enhanced Magnetic Resonance Imaging brain showed.

discharged in a stable condition on the same regimen. After 12 weeks of treatment the patient was able to walk normally with persistent hearing loss although this improved slightly. The patient is on follow-up and has improved markedly.

Discussion

Brucellosis is a disease caused by infection with Gram-negative microorganisms of the genus Brucella [5]. It is a zoonosis transmittable to humans. All Brucella infections are led by direct or indirect exposure (e.g., milk or milk products and raw meat) to animals [6]. The disease starts with nonspecific symptoms such as fever, sweats, malaise, anorexia, headache, and back pain. Among the most commonly involved systems are the hepatobiliary system and the skeletal system [5] because Brucella are in the gastrointestinal tract. However, nervous system involvement is not common. The incidence of this has been reported to be between 3 and 25% of the cases of generalised brucellosis [7]. Brucellosis can cause meningitis, encephalitis, neuritis, brain abscess, demyelination, transient ischaemic attacks, occlusive vascular disease, sensorineural hearing loss, vertigo and meningovascular syndromes [8–10]. Hearing loss in brucellosis may develop following involvement of the central auditory pathways [8]. Cranial nerve involvement in neurobrucellosis has been found as part of the varied neurological presentation, and it seems that there is a predilection for the vestibulocochlear cranial nerve, leading to sensorineural hearing loss [11, 12]. The criteria necessary for definite diagnosis of neurobrucellosis are (1.) neurological dysfunction not explained by other neurologic diseases, (2.) abnormal CSF indicating lymphocytic pleocytosis and increased protein, (3.) positive CSF culture for Brucella organisms or positive Brucella IgG agglutination titre in the blood and (4.) CSF response to specific chemotherapy with a significant drop in the CSF lymphocyte count and protein concentration. Our patient fulfilled all the above mentioned criteria for the diagnosis of neurobrucellosis [13].

The treatment of neurobrucellosis is still controversial [14–18]. There are few guidelines for the appropriate duration of neurobrucellosis treatment [19]. Recent reports recommend a regimen with a combination of three or four antibiotics for neurobrucellosis [20]. Doxycycline is the preferred tetracycline in neurobrucellosis because its tissue and CNS penetration is much better and it also has a longer half-life. Rifampicin and co-trimoxazole also offer a good penetration into the CSF [19]. Ciprofloxacin combined (not solely [21]) with other antibiotics is as effective as the standard regimen of doxycycline and rifampicin [22, 23]. Ceftriaxone also offers good *in vitro* activity and penetration into the CSF [24]. Ceftriaxone alone was given to three patients at onset of disease until definite diagnosis. With this antibiotic signs and symptoms abated. Ceftriaxone can be chosen as the third antibiotic in the hospitalisation period or for a patient for whom oral antibiotic could not be given. In neurobrucellosis, treatment should be maintained until improvement of clinical symptoms and CSF response. Serological findings have little value to decide when to stop therapy.

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