Case Report: Neurobrucellosis with Plastered Spinal Arachnoiditis: A Magnetic Resonance Imaging–Based Report

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Abstract. Diffuse spinal arachnoiditis in neurobrucellosis is a rare manifestation. We report a boy aged 17, presenting with hearing impairment and recurrent vomiting for 18 months, weight loss for 12 months, dysphagia, dysarthria, hypophonia for 6 months, and gait unsteadiness for 5 months. He had bilateral 5th (motor) to 12th cranial nerve palsy, wasting and weakness of limbs, fasciculations, absent tendon reflexes, and positive Babinski's sign. Cerebrospinal fluid (CSF) showed raised protein and pleocytosis. Magnetic resonance imaging (MRI) showed extensive enhancing exudates in cisterns and post-contrast enhancement of bilateral 5th, 6th, 7th, and 8th nerves. Spine showed clumping with contrast enhancement of the cauda equina roots and encasement of the cord with exudates. Serum and CSF were positive for anti-*Brucella* antibodies. He showed significant improvement with antibiotics. At 4 months follow-up, MRI demonstrated near complete resolution of cranial and spinal arachnoiditis. It is important to recognize such rare atypical presentations of neurobrucellosis.

INTRODUCTION

Neurobrucellosis is an uncommon manifestation of generalized Brucellosis and the incidence is reported to be between 3% and 25%. Central nervous system (CNS) manifestations include meningitis, white matter lesions and demyelinating syndrome, intracranial granuloma, and transverse myelitis. Peripheral nervous system involvement generally manifests as an acute/chronic polyradiculopathy.^{1,2} Its effect on spinal cord is considered to be due to compression by an abscess or granuloma or involvement of the spinal roots.^{3–5} A single case of involvement of spinal roots by magnetic resonance imaging (MRI) in neurobrucellosis has been documented.⁶ In this report, we describe a boy who manifested unusually with an initial symptom of rapidly progressive hearing loss followed by facial weakness, bulbar palsy, and flaccid quadriparesis. The MRI demonstrated extensive spinal exudates plastering and encasing the entire cord leading to spinal arachnoiditis, along with thick basal meningitis and lower cranial nerve enhancement.

CASE REPORT

A 17-year-old boy hailing from Dharwad, Karnataka, India (endemic for Brucellosis), was evaluated during February 2017 with symptoms of progressive bilateral hearing impairment and recurrent intractable vomiting for 18 months, weight loss for 12 months, progressive dysphagia, dysarthria, hypophonia and complete deafness for 6 months and mild unsteadiness of gait for 5 months. No symptoms of fever, visual impairment, seizures or cognitive decline. He did not consume unpasteurized milk products nor had direct contact with cattle. Interestingly, this patient was referred by a young neurologist as juvenile motor neuron disease (Madras motor neuron disease/riboflavinopathy). On examination, there was severe

emaciation, the respiratory, cardiovascular, and gastrointestinal systems were normal. He was conscious and alert. He had severe bilateral 5th (motor) to 12th cranial nerve involvement (Figure 1A–C). He had symmetrical wasting and weakness of all limbs, muscle strength at girdles (grade 2+/5), distal (grade 3/5), minimal appendicular ataxia, absent tendon reflexes, and positive Babinski's sign. Hemogram showed lymphocytosis (49%), normal peripheral smear, and erythrocyte sedimentation rate of 74 mm/hour. Anti-nuclear antibody profile was normal, serum ammonia was 44 μ mol/L, and serum lactate was 23.7 mg/dL. Thyroid, liver, renal functions, and serum electrolytes were normal. Serum vitamin B12 level was 647 pg/mL. Serum human immunodeficiency virus, hepatitis B surface antigen, hepatitis C virus antibodies were negative. Visual evoked potentials revealed prolonged P100 latency of 124 ms (right) and 123 ms (left). Brainstem auditory evoked response was absent on both sides, somatosensory evoked potential from right tibial nerve (N45) was 74.9 ms (prolonged), audiometry revealed bilateral profound hearing loss with absent distortion-product otoacoustic emissions. Peripheral nerve conduction studies (motor and sensory) from upper and lower limb nerves were normal. There was absent “F” waves from median, ulnar, common peroneal, and posterior tibial nerves. MRI of brain and spine was performed: pre- and post-contrast fluid-attenuated inversion recovery images showed extensive exudates in the suprasellar, perimesencephalic cisterns and deep part of the sylvian fissures. The basal cisterns were filled extensively with exudates, and bilateral 7th–8th nerve complexes were enhancing. Post-contrast T1W magnetization-prepared rapid three-dimensional gradient-echo technique images showed enhancement of cisternal portions of both the abducent and trigeminal nerves (Figure 1G–I). Spine imaging demonstrated clumping of the cauda equina nerve roots with post-contrast enhancement and thick encasement of the entire cord with exudates (Figure 1D–F). Cerebrospinal fluid (CSF) was slightly hazy and showed 170 cells/mm³ (lymphocytes = 95%; polymorphonuclear cells = 5%), protein of 678 mg/dL, and glucose of 8 mg/dL. Ziehl–Neelsen staining did not reveal acid fast bacilli, and CSF venereal disease research laboratory test was nonreactive. Serum and CSF were positive for

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FIGURE 1. (A–C) Clinical photographs showing an emaciated boy with bifacial weakness, wasting of tongue, and small muscles of the hands. (D–F) Post-contrast T1 fat saturated sagittal and axial images of dorsal and lumbar spine showing encasement of the cord with exudates and enhancement of the cauda equina nerve roots (arrow) with clumping, respectively. (G, H) Pre- and post-contrast axial FLAIR images of brain at the level of suprasellar cistern showing extensive exudates in the suprasellar, perimesencephalic cisterns, and the deep part of sylvian fissures (arrow), respectively. (I) Post-contrast axial T1 MPR images showing enhancement of cisternal portions of bilateral trigeminal nerves and 7th–8th nerves (arrow), respectively. (J, K) Follow-up post-contrast images of the brain at the level of suprasellar cistern show decrease in the cisternal and cranial nerve enhancement as compared with the pretreatment imaging. (L, M) Follow-up post-contrast sagittal images of the spine show resolution of enhancement in the theca and nerve roots. The part figures (A, B, C) appear in color at www.ajtmh.org.

anti-*Brucella* antibodies by Rose Bengal plate test (RBPT). The serum was positive in 1:640 and CSF in 1:320 dilution for anti-*Brucella* antibody by STAT (Omega Diagnostics, Alva, Scotland). Both were also positive for immunoglobulin G and immunoglobulin M anti-*Brucella* antibody by enzyme-linked immunosorbent assay (ELISA) (NovaLisa from NovaTec GMBH, Dietzenbach, Germany). Blood and CSF cultures did not yield *Brucella* by conventional and MGIT automated method. Patient was treated with intravenous ceftriaxone, oral rifampicin, and doxycycline. At follow-up after 4 months during June 2017, patient had shown significant improvement. There was no vomiting, he was able to swallow better, had prominent nasal quality to speech, and muscle strength in limbs had improved by two grades; however, deafness persisted. Visual evoked potentials was still abnormal with P100 latency on left being 126 ms; right being 121.8 ms. CSF at this follow-up showed 24 lymphocytes, protein of 205 mg/dL, and glucose of 10 mg/dL. Serum and CSF remained positive for anti-*Brucella* antibody by RBPT, STAT, and ELISA, but the titers had reduced to 1 in 160 and 1 in 80 in serum and CSF, respectively, by STAT. Follow-up contrast imaging showed decrease in the cisternal enhancement and resolution of enhancement of the trigeminal and 7th–8th nerves. Post-contrast T1W images showed resolution of enhancement in the theca and along the spinal nerve roots (Figure 1J–M).

DISCUSSION

Brucellosis has a widespread geographic distribution and is labeled as regionally emerging zoonotic disease and has been

listed as a neglected tropical infection by the World Health Organization. It generally affects the gastrointestinal tract, hepatobiliary, and the skeletal system.⁷ CNS involvement is a rare, but important, complication and the occurrence rate of neurobrucellosis is reported to be < 5% among brucellosis cases.^{8,9} CNS affliction usually presents with serious complications including encephalitis, meningoencephalitis, radiculitis, myelitis, peripheral and cranial neuropathies, intracranial and subarachnoid hemorrhage, and psychiatric manifestations.^{8,10} The pathophysiology is possibly because of the direct effect of bacilli, cytokines, or endotoxins on the peripheral nerves, spinal cord, meninges, and brain.¹¹ The mortality rate of neurobrucellosis in the post-antibiotic era is 0–5.5%, but permanent neurologic deficits, particularly deafness, is common (20–30%).¹² Our patient had hearing impairment as the initial manifestation progressing to profound loss. The MRI revealed enhancing auditory nerves. In a case report, a 20-year-old German man with neurobrucellosis had permanent unilateral hearing loss secondary to eighth nerve involvement with vestibular dysfunction.¹³ However, the brain MRI did not reveal any abnormality.

In a large series of 18 cases of neurobrucellosis from Turkey, one patient had spinal arachnoiditis with features of subacute paraparesis, and MRI had shown diffuse myelitis and arachnoiditis.¹⁴ In another report on 19 cases of neurobrucellosis, six had proximal polyradiculoneuropathy, whereas spinal involvement was reported to be due to compression by abscess/granuloma or involvement of the spinal roots.³ In comparison, our patient had extensive arachnoiditis with

plastering/encasing of the entire cord with exudates and this feature has not been described earlier. Furthermore, follow-up MRI showed complete resolution of the profound arachnoiditis.

Neurobrucellosis and acute polyradiculoneuropathy resembling Guillain-Barré syndrome is reported earlier with spinal root thickening and diffuse enhancement along the distal cord and cauda equina on MRI. However, none had evidence of arachnoiditis.^{1,2,6}

Diagnosis of neurobrucellosis is usually confirmed by the detection of specific antibodies in blood and CSF by ELISA or Coombs' test, or less commonly by positive CSF cultures.¹⁵ Our patient was confirmed to have neurobrucellosis by regular testing methods viz., RBPT, STAT, and ELISA. Although bacterial culture is the gold standard for the diagnosis of Brucellosis, in the case of neurobrucellosis, it is associated with low sensitivity (< 20%) which could be either due to low bacterial load in CSF or due to usage of antibiotics.¹⁶ In a study of a large number of neurobrucellosis cases, *Brucella* culture was positive only in 28% of blood and 14% of CSF. Thus in the present report, diagnosis was based on correlation with clinical history followed by immunological tests along with CSF examination findings and MRI characteristics.

In conclusion, our report on this young boy with neurobrucellosis having thick basal meningitis and spinal arachnoiditis had presented clinically as a relentlessly progressive neurodegenerative disease with profound symmetrical deafness, cranial nerve palsies, and radicular involvement mimicking juvenile motor neuron disease with deafness. It would be imperative to consider neurobrucellosis in the differential diagnosis with the presence of multisystem involvement in patients hailing from endemic areas or having traveled to these regions.

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