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Case Report

“Sixteen and a half”: a rare neurological syndrome

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ABSTRACT: “Sixteen and a half” syndrome is a recently coined terminology for a novel pontine neuro-ophthalmological condition. It is characterised by “one and a half” syndrome with an additional ipsilateral seventh and eighth cranial nerve palsy ($1\frac{1}{2}+7+8=16\frac{1}{2}$). We hereby present a case with “sixteen and a half” syndrome, characterised by facial asymmetry, conjugate gaze palsy and unilateral deafness with vertigo. As demonstrated by magnetic resonance imaging, the responsible pontine lesion was a brainstem tuberculoma involving the ipsilateral abducens nucleus and the adjacent medial longitudinal fasciculus (MLF), along with facial and vestibulocochlear nerve. The location of the tuberculoma and the clinical presentation is unusual.

KEY WORDS: *Syndrome; Ipsilateral; Vertigo; Tuberculoma; Fasciculus*

INTRODUCTION

Two well known neuro-ophthalmological syndromes due to paramedian pontine infarction have been previously described. The “one and a half” syndrome, first reported and named by Miller Fisher in 1967¹, consists of horizontal gaze palsy to one side along with ipsilateral inter-nuclear ophthalmoplegia (INO) resulting in loss of all horizontal movements in the ipsilateral eye, except abduction of the contralateral eye. The lesion is within the ipsilateral pontine tegmentum. Anatomically, either involvement of combination of medial longitudinal fasciculus (MLF) plus abducens nerve nucleus or MLF plus Paramedian pontine reticular formation (PPRF) can cause “one and a half” syndrome. Involvement of facial nerve with “one and a half” syndrome has been termed as “eight and a half” syndrome by Eggen-Berger² who first reported three such cases. In continuation of this concept, “sixteen and a half” syndrome³ has been described with additional involvement of eighth cranial nerve in “eight and a half” syndrome. We describe a rare case of “sixteen and a half”

syndrome secondary to tuberculoma in the brainstem.

CASE DETAILS

A 55 year old male was admitted to our hospital with a four months history of headache, gradual progressive facial asymmetry, diplopia on left lateral gaze along with inability to look towards the right, decreased hearing in the right ear with vertigo and mild limb weakness on the left side. Clinical examination revealed loss of all horizontal eye movements except abduction of the left eye, associated with horizontal nystagmus of the left eye. Vertical eye movements and eyelid function were intact. Convergence was spared. Pupillary reflex was normal on both sides. He had right-sided facial weakness that involved the muscles of the entire right half suggestive of lower motor neuron facial palsy. Rinne’s test was positive in right ear with Weber’s test lateralising to left, suggestive of sensorineural hearing loss in the right ear. Power on right side was normal and on left was 4/5. Rest of the neurological examination was normal. All serum laboratory values were also normal. A magnetic resonance imaging (MRI) scan revealed T2 hypointense lesion involving right side of tegmentum of pons with extension to superior cerebellar peduncle and vermis with postcontrast enhancement and perilesional oedema (**Figure 1**). MR spectroscopy revealed reduced NAA peak with

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marginal elevation of lipid peak suggestive of tuberculoma. Pure tone audiogram was suggestive of sensorineural hearing loss in right ear with normal hearing on the left side. Cerebrospinal fluid (CSF) examination showed 113mg/dl proteins, 45mg/dl sugars, no polymorphs and 54 lymphocytes/ cu mm with no bacteria or fungi on special staining. Adenosine deaminase level was 24.3 (normal 0-6) suggestive of tuberculous meningitis. CSF culture for bacteria and fungi was negative. ELISA for HIV 1 antibody detected positive, CD4 count being 278. Toxoplasma antibodies in the serum and CSF for cryptococcal antigen were both negative. Chest radiograph and high resolution CT scan of the chest were normal. He was treated with steroids and four drug daily anti-tuberculosis therapy. Clinical improvement was noted in the form of decreasing headache and vertigo with an improvement in power after two weeks of therapy, but no improvement in eye movements was seen. Examination findings remained unchanged 1 month later.

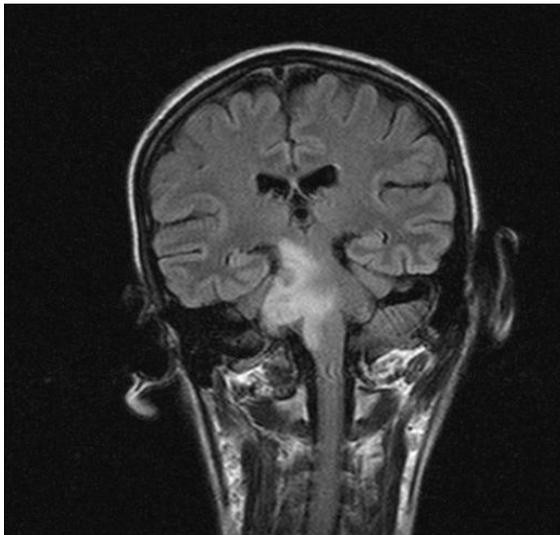


Figure 1: Coronal T2 weighted MRI image of the patient showing hypointense ring enhancing lesion in the tegmentum of pons with perilesional oedema

DISCUSSION

Tuberculosis is a commonly acquired infectious illness in developing country like India. Incidence of CNS tuberculosis among total tuberculosis cases is about 10% in immunocompromised and 2.5% in non immunocompromised patients.^{4,5} Tuberculomas were found in around 24% cases of CNS tuberculosis in one study.⁴ Pathogenesis involves extension of CSF infection into adjacent parenchyma via cortical veins or small penetrating veins or might result from haematogenous spread of systemic disease. CNS tuberculomas can have varied clinical presentation ranging from mild

headache, convulsions, hemiplegia, and also few rare presentations like panhypopituitarism⁶, unilateral or bilateral internuclear ophthalmoplegia, "one and a half" syndrome, and "eight and a half" syndrome have also been reported. Infratentorial tuberculomas are common in children and may present with brainstem syndrome, cerebellar manifestations and multiple cranial nerve palsies. Internuclear ophthalmoplegia is characterized by paresis or paralysis of adduction of the ipsilateral eye on attempted horizontal gaze to the contralateral side, horizontal jerky nystagmus in the contralateral abducting eye and intact convergence. Other associated findings are abnormalities in vertical smooth pursuit, optokinetic nystagmus; gaze evoked vertical nystagmus on upward gaze more frequent than downgaze if the lesion is bilateral and skew deviation. A unilateral INO is due to the interruption of the ipsilateral MLF after it has crossed the midline caudally in the pons from its site of origin in the contralateral abducens nucleus. In a review of 410 cases of INO, the commonest aetiology was infarction followed by multiple sclerosis and other unusual causes like trauma, tentorial herniation, vasculitis, tumour, infections (tuberculosis, brucellosis, neurocysticercosis, syphilis), lymphoma, aneurysm and AV malformation.⁷ "One and a half" syndrome is additional involvement of PPRF or sixth nerve nucleus with MLF resulting in complete loss of horizontal eye movements on ipsilateral side and adduction on the opposite side. As facial nerve fibres encircle abducens nucleus before leaving the pons, these fibres lie in close proximity to structures described above, they are also more prone to injury in this region causing additional seventh nerve involvement termed as "eight and a half" syndrome.

Our patient had an "eight and a half" syndrome plus additional ipsilateral hearing loss, this clinical entity has been named as "sixteen and a half" syndrome ($1\frac{1}{2} + 7+8=16\frac{1}{2}$). Hearing loss has not been previously described in association with either "one and a half" syndrome or "eight and a half" syndrome. Deafness is due to involvement of cochlear nucleus complex. "Sixteen and a half" syndrome therefore consists of "one and a half" syndrome plus ipsilateral facial nerve palsy and deafness. The cause of this syndrome in our patient was pontine tegmental tuberculoma which partially responded to anti-tuberculosis therapy and corticosteroids. The diagnosis was made on the basis of clinical history aided by investigations like MR spectroscopy and CSF ADA levels. One of the peculiarities of *M. tuberculosis* is the production of lipids which can be easily detected non-invasively by MR spectroscopy. The reduction of NAA peak and elevation of lipid peak is quite characteristic for a tuberculoma.⁸ Also, ADA has been considered as a marker of cell-mediated immunity and its

activity has been observed in various infections including tuberculosis. Various studies have showed that CSF-ADA estimation can differentiate tuberculomas from other neurological infections.⁹

CONCLUSION

This is a rare case of infranuclear facial weakness in association with the one and one-half syndrome and eighth cranial nerve involvement in an immunocompromised patient, the etiology being tuberculosis.

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