Annals of Clinical Case Reports

Case Report
Published: 11 Dec, 2018

Juxtaposition of One and a Half Syndrome with Pseudo One and a Half Syndrome

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Abstract

Conjugate gaze palsy with contralateral internuclear ophthalmoplegia or One and a Half Syndrome, is a rare neurological condition that was originally described in 1967. The authors present two patients, one with true one and a half syndrome caused by a CVA and another with pseudo one and a half syndrome caused by myasthenia gravis. Clinical presentation of these syndromes can be similar with minute differences appreciated on physical exam; however, the clinical relevance is quite large as the management is entirely different.

Abbreviations

INO: Internuclear Ophthalmoplegia; MLF: Medial Longitudinal Fasciculus; PPRF: Para Median Pontine Reticular Formation; CT: Computed Tomography; MRI: Magnetic Resonance Imaging

Introduction

"One and a half syndrome" was originally described in 1967 as conjugate gaze palsy with a contralateral Internuclear Ophthalmoplegia (INO) [1]. The syndrome is typically due to a single unilateral lesion of the Paramedian Pontine Reticular Formation (PPRF) or the abducens nucleus, and the adjacent Medial Longitudinal Fasciculus (MLF) [2,3]. Damage to the former manifests as the ipsilateral conjugate gaze palsy. Damage to the latter interrupts the internuclear fibers crossing midline to the contralateral medial rectus resulting in contralateral adduction failure on opposite gaze [2]. The side of the INO refers to the direction in which the previous adductor palsy manifests and is contralateral to the gaze palsy. The affected adductor muscle in the INO is able to adduct on convergence as this action bypasses the MLF. In addition, the unaffected abducting eye in an INO, during gaze in the direction of the INO, will exhibit nystagmus in the opposite direction of the INO [2,4]. Damage to these areas is most commonly seen in strokes or multiple sclerosis. However, other etiologies have been described including traumatic and malignant causes [5]. We report a case of "one and a half syndrome" and juxtapose it to a case with nearly identical findings that have been coined "pseudo one and a half syndrome".

The Case of One and a Half Syndrome

A 53-year-old man with a past history of diabetes mellitus, hypertension, and hyperlipidemia presented to the Emergency Department complaining of worsening diplopia, headache, dysarthria, nausea and left sided weakness. These symptoms had waxed and waned the prior evening of admission but the patient delayed seeking care. Blood pressure was elevated at 184/107 mmHg. Stroke was suspected but tissue Plasminogen Activator (tPA) was not administered as the patient presented outside of the required time window [6]. Computed Tomography (CT) of the head without contrast showed no acute intracranial disease. CT angiography of the brain showed various segments of moderate to severe stenosis in the anterior circulation but unremarkable vertebral and basilar arteries. Detailed neurological examination revealed right sided facial droop, left arm, leg weakness, left dysmetria and dysarthria. Ophthalmologic examination revealed full elevation and depression of both eyes, right gaze palsy (complete restriction of abduction in the right eye and adduction of the left eye), full abduction in the left eye, inability to adduct the right eye on left gaze, but intact adduction of right eye on convergence. There was right nystagmus of the left eye on left gaze. MRI revealed a right parapontine infarct extending from the ventral pons (basis pontis) to the dorsal pons (pontine tegmentum) just anterior to the fourth ventricle (Figure 1). The MLF and abducens nuclei rest in the dorsal pontine area and a true "one and a half syndrome" was diagnosed. Cardiac work up was unremarkable and patient was discharged to rehabilitation with medical

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Received Date: 14 *Nov 2018*
Accepted Date: *04 Dec 2018*
Published Date: *11 Dec 2018*

Citation:
Chai E, Cardoza S, Rosado I, Manes P, Rivero H, Bernstein A. Juxtaposition of One and a Half Syndrome with Pseudo One and a Half Syndrome. Ann Clin Case Rep. 2018; 3: 1565.

ISSN: 2474-1655

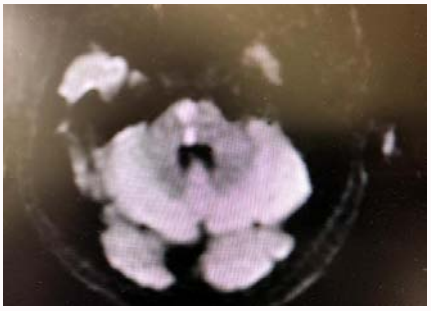

Figure 1: MRI Diffusion Weight Imaging (DWI) showing an acute stroke in the right paracentral pons.

management where the left INO began to resolve but the right gaze palsy remained unchanged after one week.

The Case of Pseudo One and a Half Syndrome

A 48-year-old male presented to the Emergency Department complaining of right eye ptosis and diplopia for two weeks. The symptoms were associated with numbness and weakness in the first digit of the upper extremity bilaterally and weakness on raising both upper extremities beyond 90 degrees. These symptoms began nearly simultaneously, had no alleviating factors, and were exacerbated as the day progressed. Physical exam revealed left gaze palsy and left adductor palsy on right gaze. The left eye was unable to adduct on conversion. The right eye was able to abduct on right gaze and there was mild left nystagmus. It was believed that the patient had a lesion affecting the left abducens nucleus (due to the left gaze palsy) and right MLF (manifesting with a right INO) in spite of the inability for the left eye to adduct on conversion. CT imaging of the head was significant for encephalomalacia in the right frontal anterior lobe superiorly. Magnetic Resonance Imaging (MRI) of the brain revealed chronic microvascular ischemic changes but no acute infarct. Diagnoses of stroke or multiple sclerosis were ruled out. Based on the generalized symptoms and fatigability, myasthenia gravis was considered and serology was examined. CT imaging of the chest revealed an equivocal 0.7 cm × 0.8 cm × 0.7 cm ill-defined mass in the thymus that suggested a thymoma. A trial of pyridostigmine 2 mg was initiated and the patient's diplopia and ptosis improved dramatically. Continued improvement was seen upon initiating a regimen of pyridostigmine 60 mg every eight hours. Serology showed was negative for acetylcholine receptor blocking (<15%) but acetylcholine receptor binding antibody level of 13.95 nmol/L (normal range <0.4 nmol/L). The patient successfully underwent an operative thymectomy. Pathology revealed no thymoma and only hyperplasia. The ocular symptoms remained during the post-operative period. This was expected as remission rates with surgery in the first year are approximately 20% [7].

Myasthenia gravis is a disease of the neuromuscular junction where antibodies either directly bind to and destroy, or adversely affect receptors in the post synaptic junction interfering with neuromuscular transmission. The result is weakness or paralysis of the affected muscles. Ptosis and diplopia are common symptoms as involvement of the extra-ocular muscles is found in approximately 90% of patients either as a presenting symptom or secondary symptom [8]. 20% to 50% of myasthenia gravis patients manifest with only ocular symptoms of ptosis or diplopia [9]. The propensity to affect the extra-ocular muscles is multifactorial. Reduced post synaptic folding in extra-ocular muscles may result in greater susceptibility to receptor destruction [10]. Higher firing frequencies of extra-ocular motor units and their unique histology may also contribute. The nature of diplopia also makes it the most conspicuous [11]. Pupil constriction is mediated through muscarinic acetylcholine receptors and is not affected by myasthenia which affects the nicotinic acetylcholine receptors.

Despite the near ubiquity of ocular findings in myasthenia gravis, no clear clinical pattern has yet to be described. Past reports reveal myasthenia can present as apparent singular cranial nerve palsy [12], Internuclear Ophthalmoplegia (INO) [13] or conjugate gaze palsy [14]. Levator palpebrae, medial recti and superior recti are traditionally thought to be the extraocular muscles most commonly affected [8]. Subsequent studies suggested ptosis or ophthalmoplegia were present in 80% of myasthenic confirming the frequency of ocular findings. However, the ocular elevators; superior rectus and inferior oblique, were the most commonly affected, followed by the lateral and medical rectus. The high incidence of elevator involvement may have been artificially inflated by the high incidence of baseline up-gaze palsy found in the elderly population. Isolated extra-ocular muscle weakness was found in only 15.4% of the cases. Bilateral extra-ocular muscle involvement was very common (72%) as well as combined vertical and horizontal involvement (59%) [15]. Recent study suggested inferior oblique involvement was found in almost 2/3 of the cases and lateral rectus in almost 1/3 of the cases [16].

Discussion

Our case represents the wide spectrum of eye findings from ocular myasthenia gravis that can be easily be mistaken as a central nervous system issue. We describe a case of a true "one and a half syndrome" of an ischemic etiology in order to introduce a case of "pseudo one and a half syndrome" caused by myasthenia gravis. The apparent INO found in a patient with myasthenia gravis has been termed a "pseudo-INO". Instead of an abducens/PPR nuclear and MLF lesion, there was a neuromuscular junction blockade pattern that mimicked the pattern of weakness in a true "one and a half" syndrome. The presence of nystagmus in both cases was indicative of an expected central compensatory response to ophthalmoplegia regardless of the underlying etiology of the ophthalmoplegia [11]. The clinical symptom differentiating between the two diagnoses was the presence or absence of adduction of the affected medial rectus on conversion as this was found only in the true "one and a half" syndrome.

Ocular syndromes can be difficult to assess; however, one must be keenly aware of the symptomatology as well as common causes. It has been established that when presented with gaze palsies and internuclear ophthalmoplegia, as we have described, an ischemic process such as a stroke or multiple sclerosis should be the primary differential diagnosis [3]. However, when common causes have been properly excluded, further studies should be administered and symptom constellations should guide the diagnostic algorithm.

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