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Case Report

CASE REPORT

Case report of a patient with 'one-and-a-half plus syndrome: nine syndrome'

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Abstract

This case talks about 'One-and-a-half plus syndrome', a clinical syndrome affecting binocular vision and facial nerve. One-and-a-half plus syndrome is a less known clinical syndrome which constitutes of a conjugate horizontal gaze palsy in one direction and an internuclear ophthalmoplegia in the other direction. Despite the known association between ischemia, autoimmune disorders, multiple sclerosis, with mono neuritis multiplex resulting in extra ocular movement disorder, one-and-a-half plus syndrome is rarely considered in the differential diagnosis of eye ball movement disorders, as many clinicians are not able to diagnose such a case as ' the eyes don't see what the mind doesn't know'. Our report aims to raise awareness about connective tissue disorders presenting as neuro-ophthalmological syndrome, as early recognition can accelerate diagnosis and decrease the morbidity.

INTRODUCTION

The one-and-a half syndrome (1 and ½, OAHS) is a neurological disorder resulting in disorganization of conjugate eye movement: exotropia, conjugate horizontal gaze palsy (CHGP) in one direction and internuclear ophthalmoplegia (INO) in the other [1–5].

Conjugate Horizontal gaze palsy (CHGP) and internuclear opthalmoplegia/nystagmus is present if there is extensive lesion involving the centers and tracts (medial longitudinal fasiculus) controlling binocular vision.

The lesion may be infarction, atherosclerosis, hemorrhage in the basilar artery territory, demyelination, tuberculoma or any other SOL.

CASE DESCRIPTION

A 16-year-old female was referred from a tertiary care hospital for steady gait, tendency to fall on right side,vertigo right sided facial deviation, disorientation to persons and time and also had a 'static left eye' The family gave history of intermittent high grade fever, with profuse vomiting.

On examination, patient was restless and irritable: she had blood pressure of 110/60 mmHg, with no postural drop; pulse 90 b/min, respiratory rate 22 b/min and temperature 37.8°C. Oral ulcers were present. During neurological examination she had right side hemiataxia although there was not any focal motor deficit. Cerebellar examination was unremarkable. Gag reflex present on both sides. Brudzinski's sign was positive and cranial nerve examination revealed that the left eye could not move superiorly, inferiorly, medially nor laterally and there was right eye lateral gaze nystagmus. Left side facial palsy was also present.

A clinical diagnosis of 'One-and-a-half syndrome along with left side facial palsy' was made and keeping in view of the possible etiologies, she was planned to be worked up extensively, i.e on lines of viral, bacterial, infectious and autoimmune etiologies.

Nonspecific routine Laboratory tests revealed: hemoglobin 9.1 mg/dl, low HCT 31%, MCV 64 fl, MCHC 39 g/dl, MCH 19 pg, ESR 32 mm/h, platelets 331 × 10^9/l, WBC 7.6.

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Features of acute disseminated encephalomyletis seen on MRI (Fig. 1).

The diagnosis of MS was ruled out by, the location of lesions, and their enhancement pattern was not typical of MS and CSF analysis showed no oligoclonal bands.

CSF report revealed marginally elevated CSF glucose and CSF protein, polymorphs and lymphocytes were all within normal range.

Viral markers, i.e. CSF PCR for HSV1, HSV2 and VZV were reported negative.

MRA and MRV were unremarkable to show any CT disease association.

All required labs including connective tissue profile ANA, Anti-DsDNA, ENA profile sent and CSF examination including PCR for VZ, and HSV sent to Agha Khan lab.

She was empirically started on IV Acyclovir 750 mg TDS, Inj Dexamethasone, 4 mg QDS, IV Mannitol 100 ml × BD and antibiotic cover of IV Ceftriaxone 2 g BD. She was also started on oral Anti Tuberculous drugs, however, only continued with that treatment for 2 days and was abandoned on further thought process and discussion by the medical team. Autoimmune workup revealed U1-RNP antibodies significantly positive 14.18U/ml and SS-B/LA antibodies marginally positive 1.20U/ml along with negative Anti dsDNA and ANA.

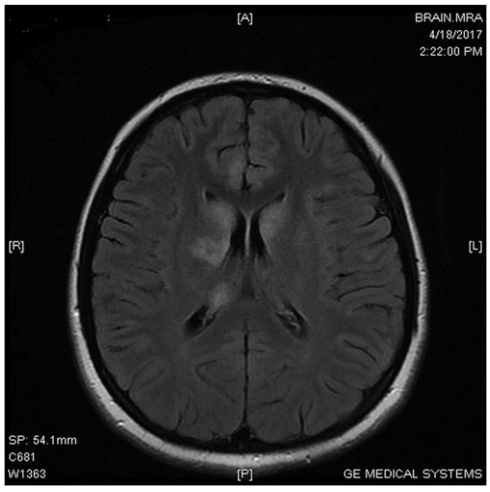

Figure 1: MRI brain (with gadolinium contrast) of patient on presentation showed multifocal hyper intensities in cortical and subcortical location of parietal, temporal, frontal lobes, basal ganglia, thalami, pons and left side midbrain causing localized swelling, mild compression upon frontal horns of lateral ventricles. Ring like enhancements in some lesions of basal ganglia, thalami and pons

Final diagnosis of one-and a-half plus syndrome was made, i.e. nine syndrome.

After 1 week treatment course (IV. Acyclovir, IV. Dexamethasone, Right eye horizontal nystagmus disappeared, movement of left eye (that with complete opthalmoplegia) improved in all other gazes, and left facial palsy also significantly improved. The patient could not walk without support but tendency to fall reduced significantly.

MRI scan after 1 week showed reduction in size as well as mass effect of previously noted hyperactive intensities of brain and brain stem (Fig. 2).

Patient was discharged from the hospital after 2 weeks course of antiviral on low dose steroidal therapy. She was no longer irritable, facial palsy, generalized weakness had improved and left eye lateral movement was marginally better. Counseling was done in detail.

Patient visits the outdoor department every 4 weeks. She has showed complete recovery. No signs of facial palsy, left eye gaze palsy in any direction nor generalized weakness present. Currently she is on steroid sparing immunomodulators, i.e. Azathioprine 100 mg/day, Hydroxychloroquine 200 mg BD and Vitamin B6 along with physiotherapy. Regular follow up advised.

DISCUSSION

OAHS, Clinical classification based on neuro-ophthalmological manifestations

(Can be classified if any one of the following fulfills the criteria with the patient)

OAHS, Type 1	a. CHGP and INO (horizontal nystagmus) b. CHGP and preserved abduction in one eye

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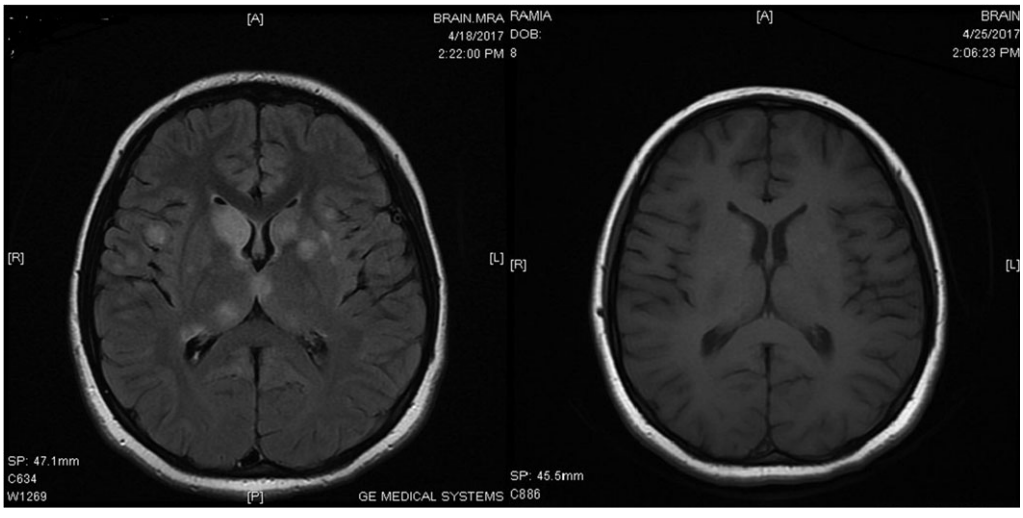

Figure 2: Comparison of MRI before and after 1 week treatment.

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OAHS, Type II	a. CHGP and normal adduction in one eye b. Adduction nystagmus and preserved bilateral abductor c. CHGP and bilateral abduction palsy d. Any combination plus bilateral miosis
OAHS, Type III	a. CVGP and asymmetrical vertical eye movements b. CVGP and vestibular nystagmus c. Vertical and horizontal gaze nystagmus at the same time

AN = Abducent nucleus.
CHGB = Conjugate horizontal gaze palsy.
CVGP = Conjugate vertical gaze palsy.

One-and-half plus syndromes

Eight-and-half syndrome
OAHS with facial palsy due to posterior circulation syndrome, Eric Eggenberg [4] added cranial nerve 7 to OAHS (1½ + 7) and he named the combination as eight-and-half (8½) syndrome.

Other causes of this combination syndrome autoimmune causes, hemorrhagic stroke, giant cell arteritis and pontine tuberculoma.

More combinations are made by adding numbers of the cranial nerves involved, e.g.:

Five-and-a-half syndrome
(1½ + 4-throclear nerve): OAHS and fourth nerve palsy.

Seven-and-a-half syndrome
(1½ + 6-abducens): OAHS and sixth nerve palsy.

Nine syndrome
Eight-and-a-half syndrome and an additional lesion in the midbrain tegmentum/red nucleus causing contralesional hemiataxia [6].

11½ Syndrome
(1½ + 7-facial nerve + 3-oculomotor nerve).

15½ Syndrome
(Bilateral seventh nerve palsy and one-and-a-half syndrome: 7 + 7 + 1½).

16 Syndrome
(Bilateral seventh nerve palsy + one-and-a-half syndrome + hemiparesis) [7].

If simple arithmetic combinations adds confusion than clarity, then adding the term 'plus' with typical OAHS, 'Type I–II–III' may define the extent of illness.

CONFLICT OF INTEREST STATEMENT

No conflict of interest.

FUNDING

No sources of funding.

ETHICAL APPROVAL

No approval required.

CONSENT

A verbal and written informed consent was taken from the patient for writing of her case report.

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