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CASE REPORT

Thirteen-and-a-Half syndrome in 14 year old female

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ABSTRACT

Background: Thirteen-and-a-half syndrome consists of a one-and-a-half syndrome with an ipsilateral facial and trigeminal nerve palsy. This is due to lesions that affects the ipsilateral paramedian pontine reticular formation (PPRF) or the ipsilateral abducens nerve nucleus (VI), the contralateral medial longitudinal fasciculus (MLF), the facial nerve (VII), and the trigeminal nerve (V) .
Objectives: This is a case of Thirteen-and-a half syndrome and stress the importance of a proper neurologic exam to aid in the localization of lesions in the brain.
Methodology: This patient was monitored during her admission. She underwent a plain cranial CT scan to confirm the suspected hemorrhage and supportive management was done to relieve her symptoms.
Results: A 14-year-old female patient presented with a one-day history of right-sided hemiparesis. There was associated binocular diplopia, dizziness, slurring of speech, dropping of the left lip, and three episodes of spontaneous projectile vomiting. Plain cranial CT scan showed a left-sided pontine hemorrhage, and she was then advised admission. There was exotropia of the right eye on primary gaze with -4 on adduction, abducting nystagmus on the right eye, horizontal gaze palsy on the left eye, no convergence, left facial weakness, and decreased left facial sensation with minimal improvement during her admission. On the 1-year follow up, there was significant improvement with full motility on the right eye and a -1 on abduction on the left eye.
Conclusion: This is a case of a Thirteen-and-a-half syndrome in a young female patient. A complete neuro-ophthalmological exam is paramount as it is both sight-saving and life-saving.

Introduction

Conjugate horizontal gaze is a binocular eye movement that allows the eye to move together toward one direction of gaze. These are supranuclear in origin with control arising from the cerebrum and brainstem. The horizontal gaze center is specifically located at the paramedian pontine reticular formation (PPRF) at the pons. It sends a signal towards the ipsilateral abducens nerve as well as the contralateral medial longitudinal fasciculus (MLF). Injury to this area will result in the one-and-a-half syndrome with the "1" being the ipsilateral gaze palsy and the "1/2" being the internuclear ophthalmoplegia (INO). Clinically, this presents as an adduction deficit of the eye toward the affected side with a horizontal nystagmus on abduction of the contralateral eye. Furthermore, cranial nerves 5,7, and 8 may be affected due to their proximity as they are either found or in proximity to the pontine tegmentum.[3,5,9].

Thirteen-and-a-half syndrome consists of a constellation of findings primarily including one-and-a-half syndrome with ipsilateral facial palsy (VII) and ipsilateral trigeminal nerve palsy (V). This is due to a lesion that affects the ipsilateral PPRF or the ipsilateral abducens nerve nucleus (VI), the contralateral MLF, the fascicle of the facial nerve (VII), and the trigeminal nerve (V) [5]. This syndrome can occur from a lesion to the dorsal pons through an ischemic stroke, intracranial hemorrhage, demyelinating conditions, and even mass lesions [1,2,5,9].

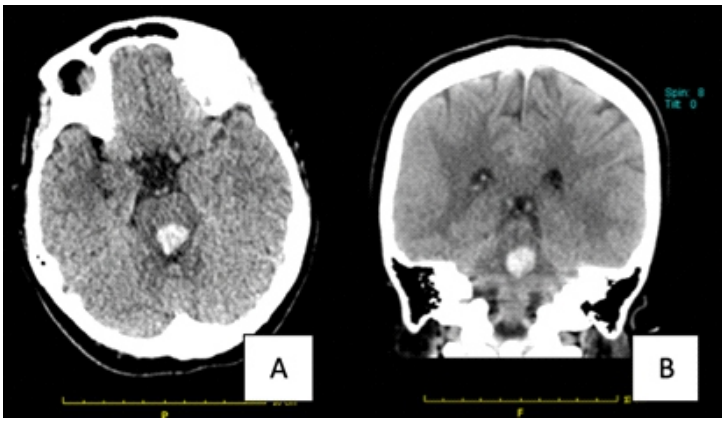
There have been 2 case reports of thirteen-and-a-half syndrome to date. In 2016, Albon and La Hood reported a case of a 50-year-old man with a One-and-a-half syndrome associated with an ipsilateral facial and trigeminal nerve palsies (1+5+7) as a result of a post-transplant lymphoproliferative disorder which affected the pons as seen on magnetic resonance imaging [1]. Another case in 2021 by Rardin, Ahmed, and Martin, noted a case of a 72 year old woman with left facial weakness, esotropia of the left eye, left gaze palsy, and decreased corneal sensitivity associated with a left dorsal pontine hemorrhage due to uncontrolled hypertension [5].

This case report aims to discuss a rare Thirteen-and-a-half syndrome in a young Filipino female which is the first case to be reported in a young patient. The objective of this report is to discuss the rarity, as well as the presentation of this syndrome, which demonstrates the importance of a comprehensive neurologic and ophthalmologic examination.

Methodology

Results

A healthy 14-year-old female, with no noted comorbidities but with a family history of a cerebral cavernous malformation from her paternal grandfather came into the outpatient department with a one-day history of right-sided weakness with stiffening of the extremities. There was associated binocular diplopia, dizziness, three episodes of spontaneous projectile vomiting, slurring of speech, and dropping of the left lip. The following day, the patient sought to consult with the Ophthalmology outpatient department due to diplopia, but was advised to seek an emergency department (ED) consult. At the ED, the patient underwent a plain cranial CT scan which showed a left-sided pontine hemorrhage (Figure 1).



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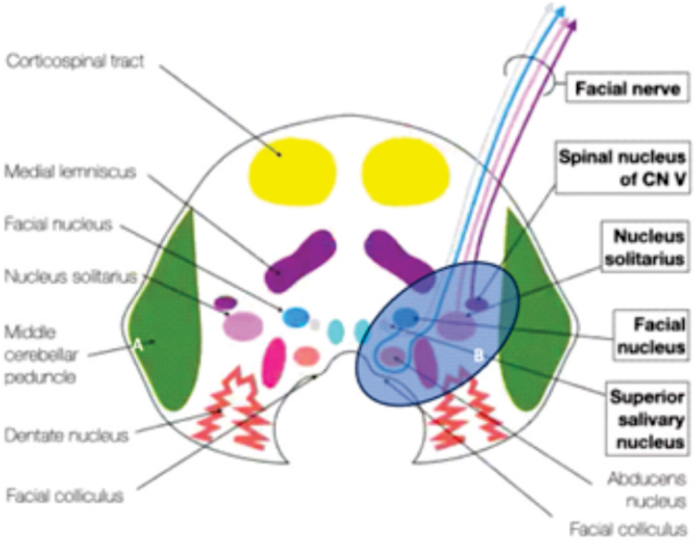

Figure 1. Plain Cranial CT Scan. [A] Axial View [B] coronal View [C] Pontine anatomy demonstrating the proximity of the nucleus of cranial nerve 5, 6, and 7 (Hacking, 2020).

The patient was then subsequently admitted to the intensive care unit. A complete neurologic and ophthalmologic examination was done wherein there was noted exotropia of the right eye on primary gaze with -4 on adduction and abducting nystagmus, -4 on abduction and adduction of the left eye, loss of convergence, left facial weakness, and decreased left facial sensation as seen in image A and B of figure 2. There was a noted right sided hemiparesis 4/5 for both the upper and lower extremifies. The rest of the neurologic examination was unremarkable. The patient also was noted to have good visual acuity at 20/20 for both eyes, 12/12 on Ishihara color plate testing, and her pupils were noted to be 2mm equally and briskly reactive to light with no relative afferent pupillary defect (RAPD). This constellation of findings is in line with a Thirteen-and-a-half syndrome that can be localized to the dorsal pontine area.

During the course of her admission, neurology service deduced that the pontine hemorrhage was due to a pontine cavernous hemangioma because of the constellation of symptoms, it being the most probable cause in young patients [6]. She was prescribed leviteracetam for seizure prophylaxis and mannitol for intracranial pressure control. Patient was advised to do an alternate patching of her eyes to relieve the horizontal diplopia. On discharge, there was noted improvement in the extraocular motility, but still with binocular horizontal diplopia.

A year after post-consultation, the patient sought a follow-up consultation at the outpatient department where she was noted minimal binocular horizontal diplopia only on lateral gaze with no noted headaches, nausea, vomiting, nor weakness. The patient still had a best corrected visual acuity of 20/20 for both eyes with 12/12 on Ishihara color plate testing, pupils were noted to be 3mm equally and briskly reactive to light with no RAPD, but with nystagmus on both eyes. On extraocular motility testing; there was a noting of -1 abduction deficit on her left eye as seen in image C in Figure 2. The patient was advised to have a yearly follow up for monitoring of her neurologic status and overall well-being, as there is an increased risk for re-bleeding; it will also serve as an assessment of her siblings due to the strong family history present in her case.

Discussion

Here is a case of a 14 year old female with right exotropia, left horizontal gaze palsy, an ipsilateral internuclear ophthalmoplegia, an ipsilateral facial nerve palsy, and an ipsilateral trigeminal nerve palsy that had no known underlying conditions. On imaging, a pontine hemorrhage was noted, which was due to a pontine cavernous hemangioma. These findings fit with the anatomic location of the hemorrhage (Figure 1). There was note of an exotropia on the right eye which was termed paralytic pontine exotropia in an article by Sharpe, *et al.*, in 1974. Sharpe noted that there is a paralytic lateral

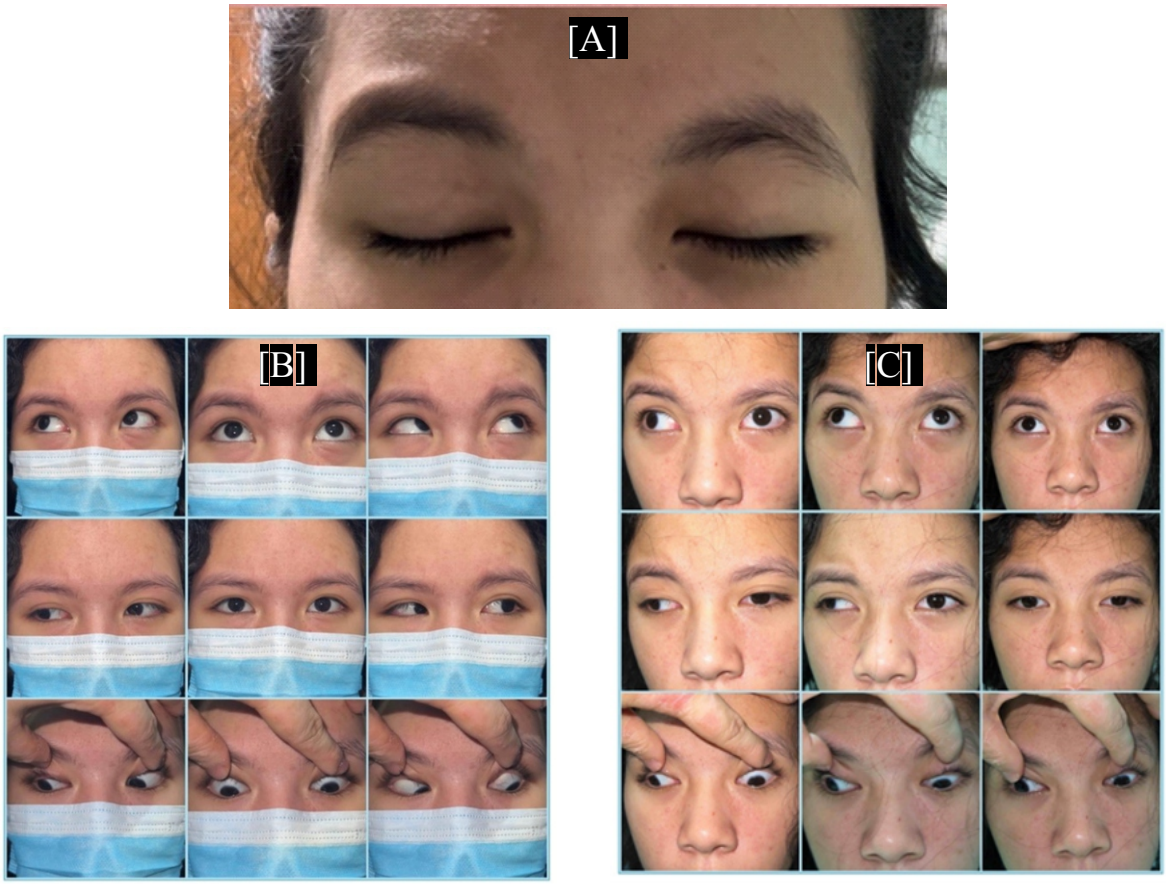

Figure 2. Extraocular motility testing during the admission and on 1 year follow up. [A] Left facial weakness presenting with asymmetric eye brow elevation, right more than the left. [B] On admission, ocular motility testing showing aright exotropia, horizontal gaze palsy on the left eye, and an adduction deficit on the right eye [C] 1 year post insult, Ocular motility testing showing a -1 abduction deficit on the left eye

deviation of one eye and the contralateral eye has a horizontal gaze palsy. This denotes the lesion to be at the pontine area due to damage to the PPRF and MLF on the side opposite the deviated eye. This finding is due to the tonic contralateral ocular deviation of the contralateral eye [7,8]. Up to 25% of cerebral cavernous malformations (CCM) affect children less than 18 years of age. The 5 year risk of hemorrhage in these patients is at 20% with the re-bleeding risk at 7.1% if with the following risk factors: family history of CCM and finding of a brainstem cavernous malformation (BSCM) [6].

Cavernous hemangiomas are diagnosed based on the combination of radiologic and neurologic findings [6]. The imaging of choice for cavernous hemangioma are T1 and T2-weighted magnetic resonance imaging (MRI). There is a pathognomonic "popcorn-ball" appearance due to the loculated hemorrhages, hemosiderin deposition, and gliotic reaction which is seen on MRI [4]. Hemorrhages from this lesion tend to exert pressure on the surrounding cranial nerve nuclei and tract which result in deficits in 60% of patients [4]. Comparing the plain cranial CT scan as well as the representative pontine anatomy, it can be noted that the pontine hemorrhage is located in the pontine tegmentum where the 5th, 6th and 7th cranial nerve nuclei are found [4].

This is a rare case that shows a thirteen-and-a-half syndrome in a young female. It is important to always consider ischemic and hemorrhagic causes in the young despite their lack of comorbidities. This is due to CCMs being one of the most common causes of intracranial hemorrhage in the young and monitoring is crucial as there is a high rebleeding risk in these patients [6].

To date there are two published case reports on thirteen-and-a-half syndrome. One case was published in 2022 by Gosal, *et al.*, a case of a 20 year old male who presented similarly to this case. That patient also had a intracranial hemorrhage which was due to a cerebral cavernous malformation as seen on MRI [10]. The second case was published in 2016 by Albon, *et al.,* about a 50 year old male who also had similar findings, but the etiology was due to a mass specifically a cerebral lymphoma [1]. In comparison to these two cases, the first case noted minimal improvement with persistence of the left horizontal gaze palsy after 2.5 years, while no date on long term outcomes for the second case. This is different compared to this patient as there was almost complete resolution in this case [1,10]. Furthermore, the similarities in etiologies in the 14 year old female and 20 year old male, show that intracranial hemorrhages from cerebral cavernous malformations are indeed the most common cause of these types of hemorrhages in the young, while mass lesions are more likely in the elderly as seen in the case of the 50 year old man [1,6,10].

When considering the findings of a left horizontal gaze palsy, left facial weakness, and decreased facial sensation on the left, localization to the left dorsal pontine area is possible. This case serves to highlight importance of a comprehensive neurologic and ophthalmologic exam to localize the lesion which aids in timely and proper management.

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