CASE REPORT

Progressive delayed hemidystonia following clinically mild traumatic brain injury

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SUMMARY

A 16-year-old boy presented with progressive left hemidystonia over 3 years. The possibilities of symptomatic hemidystonia due to focal lesions such as infarct (vasculitis), tumours, tuberculoma, arteriovenous malformations or hereditary degenerative disorders such as Wilson disease were considered. Imaging showed a peculiar scar involving right basifrontal region extending upto anterior, centromedian and dorsomedial nuclei of thalamus due to blowout fracture of roof of orbit. This scar was responsible for progressive left hemidystonia. On probing the history, it was revealed that patient had sustained a mild traumatic brain injury (mTBI) 3 years ago. Burke-Fahn-Marsden dystonia severity rating scale showed improvement from 19 to 6 after treatment with tablet trihexyphenidyl 16 mg and clonazepam 1 mg. A linear scar reaching upto thalamus due to blowout fracture of roof of orbit following clinically mTBI is unique. Delayed, progressive hemidystonia has been reported following severe head injury, however is less common following clinically mTBI.

BACKGROUND

Hemidystonia is dystonic posturing involving one side of body¹. According to recent consensus on classification of dystonia, our patient had childhood onset progressive hemidystonia (axis 1) and most probably secondary to focal brain lesion (axis 2).¹

It is generally under-recognised and under-reported.¹ Hemidystonia following severe head injury has been reported.² However, progressive hemidystonia following mild traumatic brain injury (mTBI) is rare.

Most of the times, haematoma or vascular lesions in caudoputamen area are reported. A clinically mTBI causing scar involving the thalamus resulting in delayed hemidystonia is unique.

CASE PRESENTATION

A 16-year-old previously healthy boy presented with history of dystonic posturing of left upper limb initially followed by lower limb. There was history of an accident 3 years ago. He fell down from a bicycle and sustained a minor abrasion over right forehead which was treated. His X-ray skull was normal and brain imaging was not done at rural place. There was only mild weakness in his left upper limb at the time of accident which resolved over 6–8 hours; however, there was no loss of consciousness, headache, vomiting, memory disturbances or behavioural changes.

One month later, parents noticed abnormal clenching of his left hand followed by difficulty in walking due to involuntary movements of his left foot. Three years later, he presented to our outpatient department with significant clenching of fingers of left hand with adduction, flexion of thumb and fingers with extensor posturing at metacarpophalangeal joints suggestive of thalamic hand dystonia (online supplementary video 1) and abduction, extension of left arm. Posturing used to worsen with action. He was dragging left lower limb with adduction and internal rotation at hip joint and inversion, plantar flexion of foot while walking. Burke-Fahn-Marsden dystonia severity score was 19 on presentation.

Rest of the central nervous system examination was normal. There was no Kayser-Fleischer ring. Birth and development history was normal.

INVESTIGATIONS

CT brain showed a subtle gliosis in right thalamus with fracture of roof of orbit (figure 1).

MRI brain showed a linear scar in the right basifrontal region extending to anterior, centromedian and dorsomedial nuclei of right thalamus along with a bone fragment due to a blowout fracture of the roof of orbit (figure 2).

Other blood parameters were normal—serum ceruloplasmin, and Peripheral Smear for acanthocytosis was negative.

DIFFERENTIAL DIAGNOSIS

Clinically from history and examination following possibilities were considered.

Figure 1 (A) Sagittal CT brain showing bone fragment in orbital roof and a linear scar extending from fracture site to right thalamus. (B) Axial CT brain showing gliotic region in right thalamus.

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Figure 2 (A) Axial T2-weighted MR images show peculiar linear scar extending from roof of orbit to thalamus on the right side; (B) coronal MRI image showing scar in right anterior, centromedian and dorsomedial nuclei of thalamic region.

1. As patient presented with progressive left hemidystonia gradually worsening since last 1 year possibility of symptomatic hemidystonia due to focal basal ganglia lesion such as infarcts (due to either vasculitis, antiphospholipid antibody syndrome, perinatal vascular insult), space occupying lesions such as slow growing tumours, infective lesions such as tuberculoma and arteriovenous malformations were considered as highly possible differential diagnosis.
2. There was a history of fall from bicycle 3 years ago, without any loss of consciousness, seizures and Glasgow Coma Scale was 15. It was a clinically mTBI. Such mTBI causing haematoma significant enough to cause hemidystonia was considered to be a less likely differential.
3. As patient was a young boy with hemidystonia, possibility of hereditary degenerative causes such as Wilson's disease, neurodegeneration with brain iron accumulation such as fatty acid hydroxylase-associated neurodegeneration (FAHN) and aceruloplasminemia were considered as differentials although less likely. Our patient had pure hemidystonia without any accompanying other movement disorder, extrapyramidal signs or behavioural changes which are more commonly seen with hereditary degenerative disorders.
4. Hemidystonia-hemiatrophy syndrome was also kept as one of the possibilities.

TREATMENT

Patient was treated with trihexyphenidyl 16 mg and clonazepam.

OUTCOME AND FOLLOW-UP

There was significant improvement in his posturing of left upper limb and was able to completely open the fist (online supplementary video 2). Burke-Fahn-Marsden dystonia severity scale improved from 19 to 6 after 2 months of treatment.

DISCUSSION

A 16-year-old male patient presented with progressive left hemidystonia over 3 years without any prior neuroimaging. In view of progressive hemidystonia, focal basal ganglia lesions such as infarct (due to either vasculitis, antiphospholipid antibody syndrome, or perinatal vascular insult), slow growing tumour, granulomas such as tuberculoma and AV malformations were considered highly likely possibilities.

Though it was a pure dystonia, possibility of hereditary degenerative disorders such as Wilson disease, neurodegeneration with brain iron accumulation such as FAHN and aceruloplasminemia

were also considered. Another differential diagnosis considered was hemidystonia-hemiatrophy syndrome.

On probing the history, patient briefly mentioned history of fall from a bicycle followed by minor forehead abrasion and transient left upper limb weakness which improved over 6–8 hours. There was no loss of consciousness, vomiting, memory or behavioural disturbances. As the GCS was 15/15 and X-ray skull was normal, brain imaging was not done. According to WHO, American Congress of Rehabilitation Medicine (ACRM) and Centre for Disease Control (CDC) criteria of severity of traumatic brain injury, our patient had clinically mTBI.³ Such an mTBI causing large haematoma and resulting hemidystonia was considered as a less likely differential. On evaluation, imaging showed an unusual linear scar in the right basifrontal region extending to involve anterior, centromedian and dorsomedial nuclei of thalamus along with a bone fragment due to a blowout fracture of roof orbit. This scar caused disruption in striato-thalamo-cortical loop, and it clinically correlated well with left hemidystonia and left thalamic hand.

In a study on natural history of acquired hemidystonia, of 158 patients, most were secondary to stroke (48.5%), trauma (17%) (usually severe), perinatal injury (8.8%), tumour (5%), infection (5.7%) and arteriovenous malformation (4%). In the above study, results of neuroimaging were available for 26 patients, out of which 25 cases one or more focal lesions. There was involvement of contralateral basal ganglia most commonly putamen (48%), thalamic (28%) and cortical (32%) and one in contralateral midbrain.⁴

Another study analysed the report of 62 cases of movement disorder with focal basal ganglia lesion.

Out of 33 patients with lesions confined to either posterior or posterolateral or paramedian part of thalamus presented with dystonia (10 hemidystonia, 5 hand dystonia, 1 segmental dystonia).⁵

It is suggested that 'delayed onset dystonia' is related to aberrant neuronal sprouting in central nervous system following a static lesion.⁶ Any lesion capable of disrupting the pathway between striatum, pallidum and thalamus while preserving corticospinal pathway could result in contralateral involuntary movements.⁷

Patient presented to our outpatient department with delayed, progressive, pure hemidystonia. There was a history of clinically mTBI 3 years ago. Such a mild trauma leading to blow out fracture of roof of orbit is a rare phenomenon. It usually tends to occur during high-impact craniofacial trauma.⁸ Orbital roof fractures associated with frontal lobe contusions, cerebrospinal fluid

Learning points

- Delayed, progressive and pure hemidystonia with remote history of mild traumatic brain injury (TBI) can still be attributed to it even if the injury was mild.
- Imaging is essential for evaluation of hemidystonia because most of the times it is secondary to a focal basal ganglia or thalamus lesion.
- Subtle imaging findings localising to appropriate anatomical structures need to be looked at carefully (in our case, scar in anterior, centromedian and dorsomedial nuclei of thalamus).
- Blowout fracture of roof of orbit can rarely occur with mild TBI and apart from contusion, pneumocephalus and CSF leak, a linear scar from the frontal lobe to the thalamus can also be one of its complications.

(CSF) leak, pneumoencephalus, acute subdural haematoma, subarachnoid haemorrhage have been well described.

Our patient developed blowout fracture of roof of orbit following mTBI and a linear scar due to penetrating injury with a bone fragment. Scar extended from basifrontal region upto thalamus.

Such extension of linear scar upto thalamus due to orbital roof fracture following a clinically mTBI is quite unique. This thalamic scar correlated well with left thalamic hand and left hemidystonia.

Patient responded to trihexyphenidyl 16mg and clonazepam 0.5mg with significant improvement in posturing of left upper limb with improvement of Burke-Fahn-Marsden dystonia severity score from 19 to 6.⁹ (online supplementary video 2)

Contributors SNN involved in patient evaluation and examination, planning and preparing a concept, review of literature, acquisition of the data, and preparation of manuscript, analysis and interpretation of data, acted as corresponding author. RTC: patient evaluation and examination, helping prepare a concept, assessment of manuscript, interpreting the data, supervised work, manuscript evaluation, help in editing the manuscript, assessment of all the images and videos, help understanding the learning points. KVK: review of manuscript, manuscript evaluation, supervised work, editing the videos and images, assessing the learning points. HB helped to evaluate and edit the manuscript, assessing the learning points, proofreading.

Competing interests None declared.

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