CASE REPORT

Isolated paroxysmal dysarthria caused by a single demyelinating midbrain lesion

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SUMMARY

Paroxysmal dysarthria is an unusual condition characterised by brief episodes of dysarthria with the sudden onset and frequent recurrence. It has been mainly reported in multiple sclerosis and an association with midbrain lesions has been claimed; however, most of the reported patients had multiple brain alterations so it was difficult to associate this symptom with a specific lesion site. We illustrate the cases of two patients with an isolated demyelinating midbrain lesion presenting paroxysmal dysarthria as the only symptom; both participants had oligoclonal bands in the cerebrospinal fluid and an unremarkable follow-up. Both patients had benefit from carbamazepine treatment, similarly to previously reported cases. Our report confirms that a demyelinating midbrain lesion is sufficient to provoke paroxysmal dysarthria. It is noteworthy that an erroneous diagnosis of psychogenic disorders was initially made in both cases, highlighting the importance not to underestimate isolated paroxysmal symptoms in clinical practice.

BACKGROUND

Paroxysmal dysarthria is an unusual condition characterised by brief episodes of dysarthria with the sudden onset and frequent recurrence, and it is typically associated with multiple sclerosis^{1–9} even if it has been seldom reported in other diseases.^{10–12} It has been suggested a role of midbrain lesions in the genesis of paroxysmal dysarthria^{7–8}; however, since most of the reported cases focused on multiple sclerosis patients with widespread brain alterations, it has been difficult to ascribe certainly this symptom to a single lesion. Moreover, there are only two reports of participants with isolated paroxysmal dysarthria and a solitary upper brainstem lesion: a patient with a demyelinating lesion⁶ and a case of midbrain infarction.¹¹ Thus, the hypothesis that an isolated demyelinating midbrain lesion could be sufficient to provoke paroxysmal dysarthria is supported by only few data. Moreover, to our knowledge, a correlation between the extension of the lesion and the severity of paroxysmal symptoms has never been previously supposed.

It's worthy of mention that in most of the reported cases of paroxysmal dysarthria the patients also presented additional symptoms, signs or clinical history clearly suggesting an underlying neurological disease. When paroxysmal symptoms are the only manifestation in a previously healthy subject, it could be more difficult to recognise them promptly and avoid delays in the diagnosis of neurological disorders.

CASE PRESENTATION

Case 1: A 33-year-old woman with no previous medical history was evaluated at our hospital for short episodes (3–10 s) of inability to speak fluently, recurring 10–30 times a day, associated with feelings of 'weight in the eyes' and 'jaw tension', which had appeared 2 weeks prior. During the first evaluation no episodes were observed and the neurological examination was normal, as well as a brain CT scan, so the patient was dismissed with a diagnosis of psychogenic disorder. Owing to the persistence of the symptoms, she came back to our hospital the day after and a prolonged episode was observed by chance, so the patient was admitted to our department.

Case 2: A 46-year-old man without any relevant pathology was evaluated at our hospital for brief episodes of inability to speak fluently, impaired coordination in balance with 'rigidity' and feeling of 'shock into the head', which appeared about a month prior. The episodes started and finished abruptly, recurred several times per day and could be evoked by postural changes. At the moment of the first evaluation, the neurological examination was normal as well as the head CT scan, so the patient was dismissed with the diagnosis of psychogenic disorder. Owing to persistence of the symptoms, he came back to our hospital 2 weeks later and, at that time, the neurological examination revealed a bilateral Babinski sign so the patient was admitted to our department.

INVESTIGATIONS

Case 1: The brain MRI documented a paramedian lesion in the lower midbrain below the red nucleus level and without gadolinium enhancement (figure 1A–C). No other lesions were disclosed in the brain or in the spinal cord. Cerebrospinal fluid (CSF) examination showed normal cell count and protein levels, negative PCR for common viruses, but the presence of oligoclonal bands. Multimodal evoked potentials showed no abnormalities. During the EEG recording an episode analogous to the patient attacks occurred, with no paroxysmal abnormalities associated. Extensive laboratory tests revealed only a slight positive IgM titre for Epstein-Barr virus and influenza A.

Case 2: The brain MRI disclosed an isolated midbrain paramedian lesion (figure 1D–F) without gadolinium enhancement and in the absence of other lesions. CSF analysis showed normal cell count and negative PCR for common viruses, but increased protein levels and presence of oligoclonal bands. During the EEG recording an episode analogous to the patient attacks occurred, with no EEG

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Figure 1 MRI showing the isolated paramedian midbrain lesions of the two patients. For the first patient described, a coronal fluid-attenuated inversion recovery (FLAIR) (A), an axial T2 (B) and a sagittal T2 (C) scans are shown and a small rounded hyperintense lesion in the parasagittal midbrain can be seen. For the second patient described, a coronal FLAIR (D), an axial FLAIR (E) and a sagittal T2 (F) scans are shown and a large rounded hyperintense lesion in the midbrain can be seen.

changes. In the extensive laboratory tests only a monoclonal gammopathy of uncertain significance was found.

DIFFERENTIAL DIAGNOSIS

An epileptic origin of the episodes could have been supposed, even if the manifestation was not typical. In both patients, the absence of changes in the EEG recorded during episodes occurrence was a strong element against this hypothesis.

Paroxysmal symptoms have also been reported in episodic ataxias. However, the clinical picture of our patients did not match the typical features of these syndromes, since episodic ataxia type 1 has usually a childhood onset, episodic ataxia type 2 is usually characterised by long-lasting (hours) episodes and other episodic ataxias are often associated with further neurological signs or symptoms.¹⁰

TREATMENT

Case 1: The patient was given a high-dose intravenous steroid therapy but paroxysmal episodes persisted, so carbamazepine was introduced up to 400 mg/day, with disappearance of the symptoms in about 1 week.

Case 2: The patient was given a high-dose intravenous steroid therapy without any relevant benefit, so carbamazepine was introduced gradually up to 400 mg/day, with gradually disappearance of the symptoms during the titration.

OUTCOME AND FOLLOW-UP

Case 1: At the last follow-up, 1 year later, no new symptom had occurred; the brain and spinal cord MRI were negative for new lesions and carbamazepine withdrawal brought no relapses of the episodes.

Case 2: At the last follow-up, 2 years later, a brain and a spinal cord MRI revealed no new lesions, carbamazepine withdrawal brought no relapses and no new symptoms had appeared.

DISCUSSION

We presented two patients with paroxysmal dysarthria as isolated neurological symptom due to a single paramedian midbrain demyelinating lesion, oligoclonal bands in the CSF and unremarkable follow-up. Our report confirms that an isolated midbrain lesion is sufficient to provoke paroxysmal dysarthria in demyelinating disorders. Interestingly, the patient with truncal ataxia associated with dysarthria presented a larger lesion compared to the patient with only dysarthric symptoms, suggesting a correlation between the extension of the lesion and the severity of the symptoms.

There is little evidence about the best symptomatic treatment for paroxysmal dysarthria. A few reports suggest certain effectiveness of carbamazepine^{7–9 11 12} even if it seems not to be useful in all patients.¹³ A stable disappearance of symptoms has also been reported after carbamazepine withdrawal out of the acute phase.^{8 9 12} Our report confirms that carbamazepine is useful and that drug suspension can be considered during the follow-up.

A mechanism by which a midbrain lesion can cause paroxysmal dysarthria has been hypothesised. Ephaptic activation of axons within a demyelinating lesion is commonly accepted as cause of paroxysmal attacks in multiple sclerosis,^{8 9} and the same mechanism can be reasonably supposed for other demyelinating disorders. There is only one study of functional imaging in paroxysmal dysarthria and ataxia, involving a patient with a

lateralised midbrain stroke, and a hypoperfusion of the parietal lobe contralateral to the midbrain lesion was disclosed during the paroxysmal attack.¹¹ The parietal lobe is profusely interconnected with the cerebellum and ephaptic transmission originated from the lesion could impair the cerebellotalamocortical pathway. This could provoke a functional depression of the parietal lobe resulting in a disruption of the descending corticopontocerebellar pathway with subsequent paroxysmal dysarthria and ataxia symptoms.

It is noteworthy, finally, that an erroneous diagnosis of psychogenic disorder was initially made in both cases. Our report reminds clinicians that paroxysmal dysarthria can be the first and unique manifestation of a brain lesion, avoiding delays in the diagnosis of central nervous system diseases.

Learning points

- ▶ A single demyelinating midbrain lesion is sufficient to provoke paroxysmal dysarthria.
- ▶ The severity of the paroxysmal symptoms could be directly proportional to the extension of the lesion.
- ▶ Carbamazepine therapy could be effective at improving symptoms in paroxysmal dysarthria, and suspension of the symptomatic treatment may be considered during the follow-up.
- ▶ Isolated paroxysmal symptoms can be the first manifestation of a brain lesion, without any other signs or symptoms and even in patients with no previously known neurological disease.

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