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Case report

Solitary sclerosis presenting as isolated spontaneous paroxysmal dysarthria

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ABSTRACT

Paroxysmal dysarthria and ataxia (PDA) is a rare syndrome characterized by brief, stereotyped episodes of slurred speech, clumsiness with extremities, or vertigo. It is usually observed in young patients suffering from multiple sclerosis with numerous lesions. PDA is challenging to identify in those presenting with atypical patterns. Here, a non-ataxic variant of PDA in an otherwise neurologically healthy elderly man is presented who had a single midbrain lesion. A broad diagnostic workup illustrates the challenges of identifying PDA. Teaching points emphasize the significance of the midbrain lesion and response to anti-epileptic medication.

1. Introduction

Paroxysmal dysarthria and ataxia (PDA) is a rare syndrome characterized by brief, stereotyped episodes of slurred speech, clumsiness with extremities, or vertigo [1]. First described by Parker and Störring in the 1940s, PDA has been largely associated with multiple sclerosis (MS) and is one of the least common types of MS-related paroxysms [1,2], with Klaas et al. reporting only 57 published cases in their 2013 review [1]. No treatment guidelines exist but sustained response to carbamazepine has often been described and is considered nearly diagnostic for the syndrome.

Most cases of PDA in the reported literature have additional disseminated neurological lesions or established neurological disorders. Few reports describe patients with single lesions or who are otherwise neurologically normal [3]; such patients present a diagnostic challenge that can delay diagnosis. Here, we report a case of spontaneous paroxysmal dysarthria in an otherwise healthy individual and discuss the lessons learned from this presentation of "spontaneous paroxysmal dysarthria" (term used when no obvious signs of ataxia are present).

2. Case report

A 74 year-old man presented with 5 months of difficulty speaking and episodes of sudden and unprovoked seconds-long "wave[s] of dizziness" immediately followed by speech difficulty as though his "tongue [was] held down". These episodes lasted 15–20 s and occurred up to 30× per hour. In the embedded Video, our patient is seen before, during, and after a period of dysarthia. Other bulbar functions were preserved.

Initial magnetic resonance imaging (MRI) demonstrated a 1.4 cm area of increased T2 intensity in the caudal midbrain that remained stable over serial images (Fig. 1A-B). Broad serologic, infectious, and metabolic work up was unrevealing but cerebrospinal fluid (CSF) exhibited two well-defined gamma restriction bands.

Following initiation of carbamazepine (CBZ), the paroxysm frequency and severity dramatically declined. Two months after presentation, a repeat CSF demonstrated a persistence of oligoclonal bands. Additionally, a repeat MRI had shown redemonstration of the T2 hyperintense and T1 hypointense midbrain lesion (Fig. 1C-D). Subsequent positron emission tomography (PET) and skeletal survey scans were unrevealing, effectively ruling out malignancy or other systemic inflammatory processes. The patient received high dose methylprednisolone and a 6 month follow-up MRI showed interval lesion regression on T2-weighted imaging although some persistence of enhancement remained, albeit decreased. (Fig. 1E-F). The patient's dysarthria has since completely resolved with CBZ.

3. Discussion

This patient is an otherwise healthy elderly man who presented with sudden onset spontaneous paroxysmal dysarthria associated with a single midbrain lesion. The accompanying video provides a dramatic example of what occurs throughout an episode and highlights the isolated dysarthria. The patient's favorable symptomatic response to CBZ and interval lesion improvement after high dose steroids was felt to support paroxysmal dysarthria secondary to an isolated inflammatory lesion such as solitary sclerosis.

Multiple factors in this case made it a diagnostic challenge including

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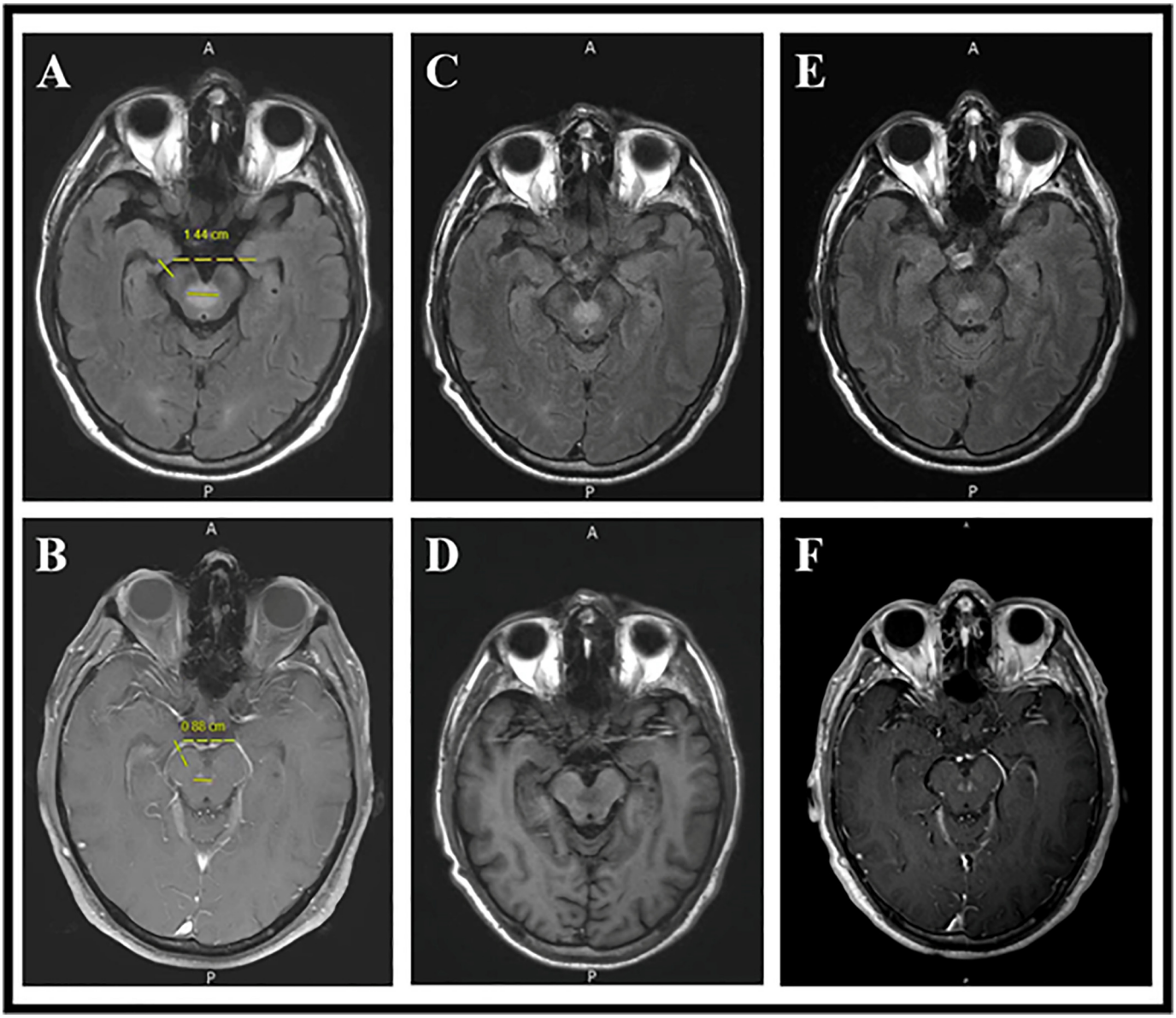

Fig. 1. T1 and T2-weighted MRI of the case over time.
MRI images 2 months (A,B), 5 months (C,D), and 10 months (E,F) after symptom onset. The patient presented at 5 months to our institution and received high dose steroids in month 8. (A,C,E) show serial axial T2 FLAIR images of the hyperintense midbrain lesion that is slightly reduced in (E). (B) is an axial T1 post-contrast FFE image of the hypointense lesion with punctate enhancement while (D,F) are axial T1 post-contrast FLAIR images.

patient age, persistence of MRI contrast enhancement, and lesion morphology. Much of the literature has described PDA in the context of MS, which usually involves a younger age of onset and disseminated disease [1]. Indeed, the persistence of oligoclonal bands supported a demyelinating process such as MS although the patient lacked clear evidence of dissemination in time and space. A neoplastic etiology was also pursued since imaging initially suspected a tumor. However, the lesion's regression with steroids and stability over time (Fig. 1E-F) suggested a demyelinating origin.

Individuals with PDA often have a characteristic midbrain lesion close to the red nucleus and the few reports of patients with single lesions have supported this finding [2–5]. Our patient's lesion corroborates those select reports. Matsui et al. previously speculated that the midbrain is key to PDA pathogenesis through disturbances in the cerebello-thalamo-cortical pathway, which connects cerebellum and cortex via the superior cerebellar peduncle (SCP) [6]. Further, isolated dysarthria has been suggested to be secondary to smaller lesions while larger lesions, which may encompass more of the SCP, lead to ataxic symptoms [3].

Finally, this patient's response to CBZ therapy supports an ongoing theory of ephaptic spread [6]. Ostermann & Westerberg first described transversely spreading, ephaptic communication of axon potentials in partially demyelinated axons [7]. They reported how ion channel dysfunction can alter adjacent fibers and generate axon potentials without synapses. Successful treatment of PDA with CBZ supports this theory since CBZ affects an entire axon membrane and therefore blocks transverse ephaptic conduction and episodes of PDA [4]. Additionally, other antiepileptic medications have been reported to be effective as well including phenytoin, lamotrigine, acetazolamide, and levetiracetam [1,2].

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Declaration of interest

None.

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