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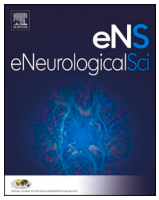

Case report

Distal oesophageal spasm in a patient with multiple system atrophy: A case report

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ABSTRACT

A 74-year-old man developed orthostatic syncope, a feeling of food stuck in his chest, and postprandial vomiting 3 years before presentation. Examination revealed severe orthostatic hypotension and cerebellar ataxia, and he was diagnosed with multiple system atrophy (MSA) with predominant cerebellar ataxia. Videofluoroscopic examination of swallowing showed lower oesophageal stricture and barium stagnation within the oesophagus. Oesophagogastroduodenoscopy revealed hypercontraction of the lower oesophagus, and high-resolution oesophageal manometry showed premature contractions of the lower oesophagus and decreased oesophageal peristalsis. The median integrated relaxation pressure in the lower oesophageal sphincter was normal, and achalasia was therefore excluded. Based on the Chicago classification version 4.0, his oesophageal dysmotility was classified as distal oesophageal spasm (DES). The stuck feeling in his chest and vomiting improved following endoscopic balloon dilation. This case suggests that DES can cause oesophageal food stagnation and postprandial vomiting in patients with MSA.

1. Introduction

Multiple system atrophy (MSA) is a neurodegenerative disorder that is characterized by the combination of cerebellar ataxia, Parkinsonism, and autonomic dysfunction. Some patients with MSA experience vomiting due to food stagnation within the oesophagus, leading to aspiration pneumonia or suffocation [1]. In addition, oesophageal dysmotility due to autonomic disturbance has also been reported in patients with MSA, including reduced oesophageal peristalsis [2] and impaired relaxation of the upper oesophageal sphincter (UES) [3]. Studies of oesophageal abnormalities in MSA however, are limited, and their pathophysiology is not fully understood. Here, we report a patient with MSA with predominant cerebellar ataxia who developed recurrent vomiting due to distal oesophageal spasm (DES).

2. Case presentation

A 74-year-old man presented to our hospital because of recurrent postprandial vomiting and syncope. At age 71 years, he had developed dizziness and syncope while standing, leading to frequent falls. At the same time, he noticed that food got stuck in his chest and he sometimes vomited after eating, leading him to choose to eat soft foods. He had no dysarthria or urinary symptoms. He had no history of hypertensive drug use. Examination revealed severe orthostatic hypotension (OH) (supine position: blood pressure (BP) 104/62 mmHg, heart rate (HR) 72/min; immediately after standing: BP 53/36 mmHg, HR 80/min). There was no compensatory increase in HR, suggesting neurogenic OH (ΔHR/Δsystolic BP $= 0.16 < 0.5$) [4]. Mild cerebellar ataxia was observed in the upper extremities. There was no muscle weakness, sensory disturbances, gait dysfunction, bradykinesia, or rigidity. Brain magnetic resonance imaging revealed cerebellar atrophy and enlargement of the fourth ventricle. Dopamine transporter single-photon emission computed tomography showed decreased striatal accumulation of ^{123}I-ioflupane bilaterally. According to the Movement Disorder Society Criteria for multiple system atrophy [5], he fulfilled the criteria for "clinically probable MSA with predominant cerebellar ataxia". Video-endoscopic examination of swallowing showed reduced pharyngeal contraction and residual saliva in the pyriform fossa. Vocal cord

Abbreviations: BP, blood pressure; DES, distal oesophageal spasm; HR, heart rate; MSA, multiple system atrophy; OH, orthostatic hypotension; UES, upper oesophageal sphincter.

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paralysis was not present on awakening. Videofluoroscopic examination of swallowing showed lower oesophageal stricture and barium stagnation within the oesophagus (Fig. 1). Oesophagogastroduodenoscopy showed normal mucosa and hypercontraction of the lower oesophagus (Fig. 2). High-resolution oesophageal manometry revealed elevated lower oesophageal pressure in synchronization with pharyngeal contraction (Fig. 3), with premature contractions in >20% of swallows. The median integrated relaxation pressure in the lower oesophageal sphincter was normal, suggesting that achalasia was unlikely [6]. There was also a decrease in oesophageal peristalsis. Based on the Chicago classification version 4.0 [6], we classified his oesophageal dysmotility as DES. Given that pharmacotherapy with agents such as calcium channel blockers or nitrates might worsen OH, we chose to perform endoscopic oesophageal balloon dilation, after which the stuck feeling in his chest and vomiting improved. He developed heartburn due to gastroesophageal reflux disease, but his symptoms were alleviated by antacids. At follow-up, he had no symptoms suggestive of recurrent oesophageal stricture and did not require further endoscopic dilation.

3. Discussion

We identified DES as an oesophageal dysfunction in a patient with MSA, using high-resolution oesophageal manometry. Previous studies of MSA demonstrated oesophageal dysfunction, including incomplete relaxation of the UES during swallowing[3], abnormal UES resting pressure, abnormal deglutitive proximal oesophageal contraction [7,8], and oesophageal hypomotility [2]. Although most studies have focused on upper oesophageal dysfunction, we identified DES by evaluating the entire oesophagus. Our findings suggest that oesophageal food stagnation in MSA can be induced by DES as well as by oesophageal hypofunction. Recognizing DES in MSA is important because, unlike oesophageal hypomotility, oesophageal dilation is required to treat food retention due to DES. This is a single case study and the relationship between MSA and DES cannot be determined. Therefore, similar cases should be sought to clarify the oesophageal characteristics of MSA.

The mechanism of DES in MSA is not completely understood, but it can be attributed to the dysfunction of inhibitory neurons. Oesophageal muscles are innervated by excitatory cholinergic neurons and inhibitory nitrogenic neurons, which are located separately in the dorsal motor nucleus [9]. Innervation by inhibitory neurons increases distally along the oesophagus, leading to a gradual increase in the latency of peristalsis. DES is generally caused by a loss of function of these inhibitory neurons, leading to synchronous oesophageal contractions [10]. A

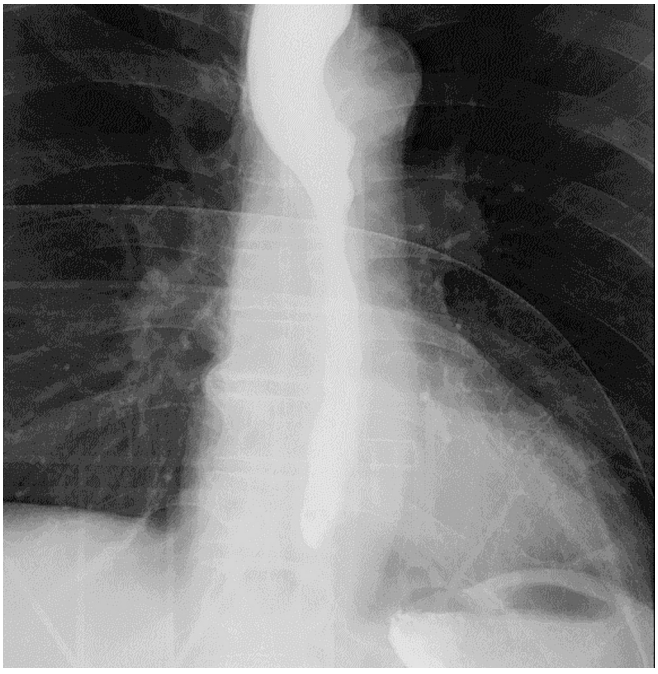

Fig. 1. Videofluoroscopic examination of swallowing.
Image showing lower oesophageal constriction and oesophageal barium stagnation due to DES.

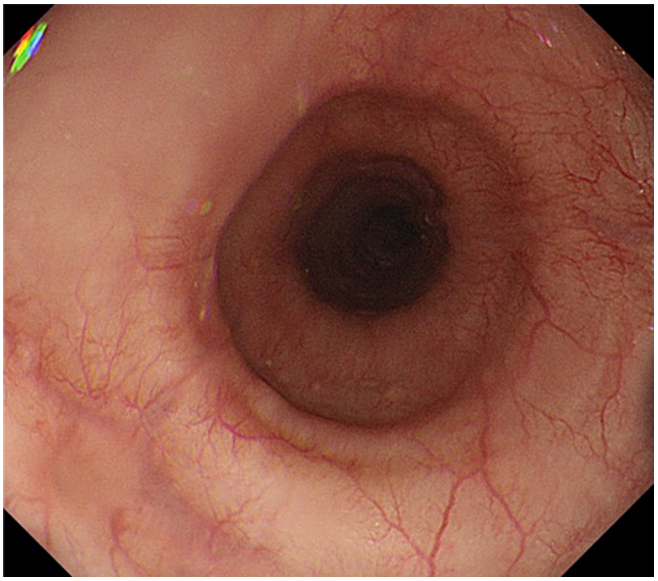

Fig. 2. Esophagogastroduodenoscopy.
Esophagogastroduodenoscopy showed hypercontraction of the lower oesophagus, suggesting oesophageal spasm. Mucosal lesions were not detected.

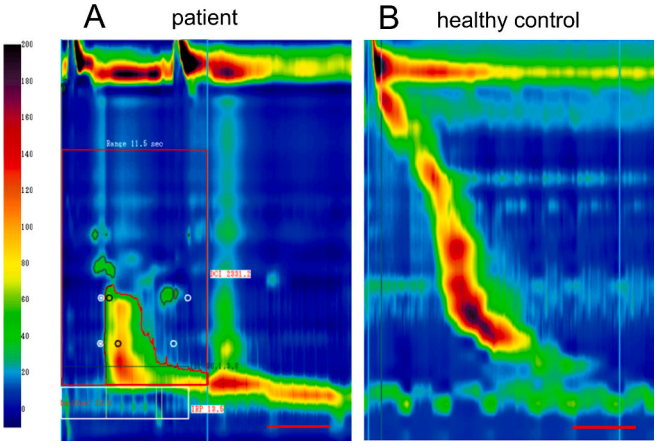

Fig. 3. High-resolution oesophageal manometry.
High-resolution oesophageal manometry in a patient with MSA (A) and in a healthy control (40-year-old man) (B). Red lines indicate 5 s. Each oesophageal pressure measurement was made when drinking 5 ml of water. The patient with MSA showed premature lower oesophageal contraction, occurring simultaneously with pharyngeal contraction. The median integrated relaxation pressure was within the normal range (13.5 mmHg, normal range: <15 mmHg). These findings suggest DES. Oesophageal peristalsis was decreased, as reported previously [2]. The distal contractile integral was normal (2331.2 mmHg·s·cm, normal range: 450–8000 mmHg·s·cm), which did not fulfil the criteria for jackhammer oesophagus characterized by repetitive prolonged contractions [6]. Relaxation of the UES during pharyngeal contraction was maintained. The UES was relaxed during pharyngeal contraction in a healthy control subject. Oesophageal peristalsis occurred from the pharynx to the stomach. There was deglutitive relaxation of the lower oesophageal sphincter. (For interpretation of the references to colour in this figure legend, the reader is referred to the web version of this article.)

previous study postulated a loss of excitatory cholinergic neurons as the cause of oesophageal dysmotility in MSA [7]. In contrast however, dysfunction of the inhibitory neurons was considered as the primary pathophysiology of DES in the present case. DES have also been reported in Parkinson's disease [11] and acute autonomic failure [12], supporting the involvement of autonomic dysfunction in DES in patients with MSA.

In conclusion, we report a patient with MSA who developed recurrent postprandial vomiting due to DES. High-resolution oesophageal manometry, in addition to videoendoscopic examination of swallowing, may be useful in patients with MSA with recurrent vomiting or a stuck feeling in the chest. Careful evaluation of oesophageal function can prevent aspiration pneumonia and suffocation in patients with MSA.

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Informed consent

Informed consent was obtained from the patient's guardian to publish this case report.

CRediT authorship contribution statement

Yoya Ono: Writing – original draft, Visualization. **Kenjiro Kunieda:** Formal analysis, Data curation, Conceptualization. **Jun Takada:** Investigation. **Takayoshi Shimohata:** Writing – review & editing, Supervision.

Declaration of competing interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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