

[ CASE REPORT ]

## Multiple System Atrophy Showing Portomesenteric Venous Gas with Pneumatosis Intestinalis on Abdominal Computed Tomography Associated with a Cause of Death

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### Abstract:

This manuscript describes the first known case of a patient with multiple system atrophy whose parasympathetic dominant disturbance might have been associated with the relative constriction of the superior mesenteric artery, leading to nonocclusive mesenteric ischemia and subsequent portomesenteric venous gas with pneumatosis intestinalis on abdominal computed tomography approaching death.

**Key words:** multiple system atrophy, portomesenteric venous gas, pneumatosis intestinalis

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### Introduction

Early autonomic failure in multiple system atrophy (MSA) is not only associated with a shorter survival (1) but is also an independent risk factor for sudden death (2). Furthermore, portomesenteric venous gas (PMVG), the presence of gas in the portal and mesenteric venous systems (3), coupled with pneumatosis intestinalis (PI), the presence of gas within the wall of the gastrointestinal tract (4), is strongly associated with acute bowel infarction (5). We herein report an MSA patient whose parasympathetic dominant disturbance might have been associated with the relative constriction of the superior mesenteric artery (SMA), leading to nonocclusive mesenteric ischemia and subsequent PMVG with PI approaching death. To our knowledge, this is the first report in which PMVG with PI was associated with a cause of death in MSA.

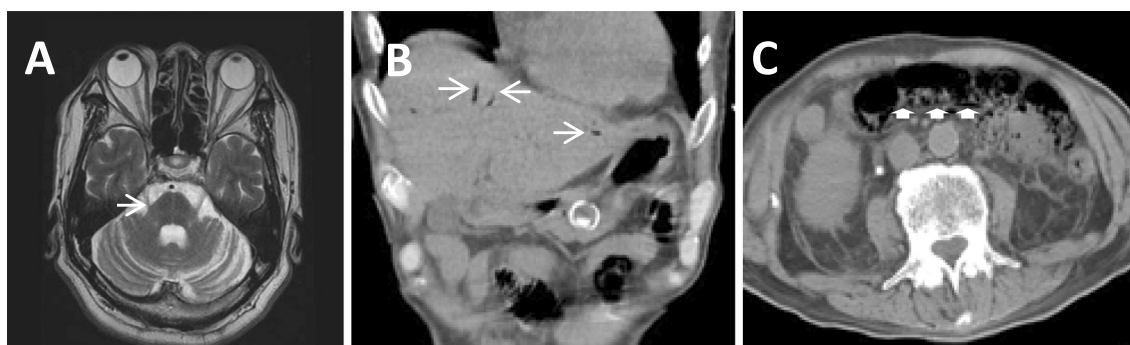
### Case Report

A 65-year-old man with a diagnosis of MSA was admitted to the hospital for bilateral pneumonia in February 2012. The patient's history included clumsiness of the left upper extremity in September 2008, and frequent falls were ob-

served starting in April 2009. Severe episodes of constipation began in July 2009 followed by orthostatic hypotension and urinary disturbance. Cranial magnetic resonance imaging (MRI) in July 2010 demonstrated slitlike high signal intensities at the posterolateral putaminal margin and a "hot cross bun" sign at the pons (Figure A) on T2-weighted sequences without obvious cerebellar atrophy. On admission the patient had preexisting unintelligible speech, severe dysphagia, wheelchair dependence, and a urinary catheter. The patient was drowsy and showed mild saccadic eye movements. There were cogwheel rigidities in all extremities without sensory disturbance. The deep tendon reflexes of the upper extremities were hyperreflexic, while those of the lower extremities were normal with bilateral Babinski signs. Diadochokinesis was difficult to examine because of his severe rigidity.

The laboratory findings were unremarkable except for inflammation due to pneumonia, and bacterial cultures of the sputum were positive for alpha-hemolytic *Streptococcus* and *Candida spp.* On the first day, cefmetazole and clindamycin were administered for bilateral pneumonia. Tracheostomy was performed on the third day, and on the fourth day, percutaneous endoscopic gastrostomy was performed to improve malnutrition. Cefmetazole and clindamycin, which were discontinued following clinical improvement, were

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**Figure.** Brain T2-weighted MRI (A) at 19 months before admission, and coronal (B) and axial (C) abdominal CT images on the last day. (A) A “hot cross bun” sign (arrow) was shown on brain T2-weighted MRI at 19 months before admission. (B) PMVG was revealed as multiple areas of intrahepatic gas (arrows) in coronal abdominal CT on the last day. (C) PI was shown as gas within the wall of the gastrointestinal tract (arrows) in axial CT images on the last day.

started again on the 21st day because of recurrent pneumonia. On the 38th day, the patient developed acute respiratory distress due to recurrent pneumonia, for which mechanical ventilation was required for three days with ampicillin/sulbactam for seven days. The patient's condition then improved, and the SpO<sub>2</sub> (saturation of peripheral oxygen) on the 42nd day was 97% on room air. On the 60th day at midnight, the patient's temperature increased suddenly to 39.5°C, and the SpO<sub>2</sub> fell to 70-80%. The ventilator was restarted along with ampicillin/sulbactam and clindamycin administration. As anisocoria was observed without other neurological symptoms, glycerol was administered for suspected cerebrovascular disease; brain computed tomography (CT) showed no obvious intracranial lesions. Abdominal guarding was observed with constipation. Abdominal CT demonstrated PMVG (Figure B) with PI (Figure C) and air in the superior mesenteric vein (SMV), while chest CT findings were unchanged. The systolic blood pressure decreased and was unresponsive to dopamine, dobutamine, and high-dose methylprednisolone. The patient subsequently died on the 61st day with a total clinical course of 3 years and 8 months.

## Discussion

Early autonomic failure in MSA is not only associated with a shorter survival (1) but is also an independent risk factor for sudden death (2), the leading cause of death in MSA (6). In our case, severe episodes of constipation followed by orthostatic hypotension and urinary disturbance progressed 11 months from the onset. Patients with combined motor and autonomic involvement within three years of the onset, as seen in our case, have a significantly increased risk of a low survival (7). High degrees of anisocoria and complex pupil response impairments observed in MSA suggest pupillary imbalance and predominantly involve the parasympathetic branch of the autonomic nervous system (8). Despite no intracranial lesions being detected in our patient's brain on CT, the possibility of cerebral infarction remained.

PMVG is a rare radiological feature and is occasionally associated with PI (4). Although CT findings indicative of PMVG or PI are not pathognomonic for acute bowel infarction, the combination of both entities is strongly associated with this problem (5). The clinical consequence of patients with intestinal ischemia and these CT findings seems to depend predominantly on the severity of their underlying diseases (3). Although no MSA patients have been reported to have PMVG, a 60-year-old man suffering from MSA with PI was reported. The presence of chronic idiopathic intestinal pseudo-obstruction due to severe dysautonomia and a longstanding bed-ridden state may have been the cause of PI in that patient (9). Our patient was in a state of septic shock on the 60th day due to recurrent intractable pneumonia, which resulted in a shift of the blood supply from the central to peripheral regions; this may have resulted in the subsequent relative loss of blood volume, including that in the SMA, towards the intestine. The SMA supplied arterial blood to the intestinal region, from which the venous blood returned through the SMV. In this case, PMVG with PI and air in the SMV were observed on abdominal CT on the 61st day just before his death. Acute bowel ischemia might have developed in the intestine supplied by the SMA, followed by the destruction of the intestinal wall, which might have induced the reflux of air in the SMV observed in this case.

Nonocclusive mesenteric ischemia was reported in a dialysis patient who showed PMVG with PI and extensive vascular calcification of the SMA (10). This nonocclusive mesenteric ischemia may be induced by prolonged hypotension during hemodialysis treatments that reduce blood flow to the small bowel and massive vascular calcification that negatively affects the compliance of the SMA. The present patient with MSA showed orthostatic hypotension, severe constipation, and urinary disturbance as symptoms of autonomic failure; parasympathetic dominant disturbance was suggested by the presence of anisocoria (8). As sepsis alone without dead bowel is an infrequent cause of PMVG (11), it was unlikely that acute bowel infarction with PMVG was induced by septic shock. In addition to prolonged hypotension

and relative loss of blood volume in SMA by septic shock, the development of nonocclusive mesenteric ischemia might also be affected by the relative constriction of the SMA, which may be provoked by the parasympathetic dominant disturbance observed in this case. Another possibility was acute bowel ischemia caused by occlusion of the SMA (12).

In conclusion, although there was no pathological evidence, the parasympathetic dominant disturbance might have been associated with the relative constriction of the SMA, leading to nonocclusive mesenteric ischemia and subsequent PMVG with PI, which was the cause of death in this patient with MSA.

**The authors state that they have no Conflict of Interest (COI).**

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