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Case Report

CASE REPORT

Medial pontomedullary junctional infarction presenting vertigo, ipsilateral facial paresis, contralateral thermal hypoalgesia and dysphagia without lateral gaze palsy, curtain sign and hoarseness: a case presentation of a novel brain stem stroke syndrome with sensory disturbance-based dysphagia and review of the literature

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Abstract

In this report, we describe unilateral medial pontomedullary junction (MPMJ) syndrome as a novel brain stem stroke syndrome. A 68-year-old woman suddenly developed vertigo, ipsilateral facial paresis, contralateral thermal hypoalgesia (TH) and dysphagia without lateral gaze palsy, curtain sign and hoarseness. Magnetic resonance (MR) imaging showed a small infarction at the right MPMJ. MR angiography did not show vertebrobasilar arterial dissection, thrombosis or vasospasm. Finally, her dysphagia regressed over 4 weeks in synchronization with recovery of TH. To the best of our knowledge and based on a review of the literature, this MPMJ syndrome associated with the unilateral MPMJ infarction is a novel brain stem stroke syndrome different from Foville syndrome, Millard–Gubler syndrome, Wallenberg syndrome or Dejerine's syndrome. In the MPMJ syndrome, transient, albeit severe, dysphagia based on the TH-impaired swallowing reflex bothered the patient more than hemiparesthesia of TH did.

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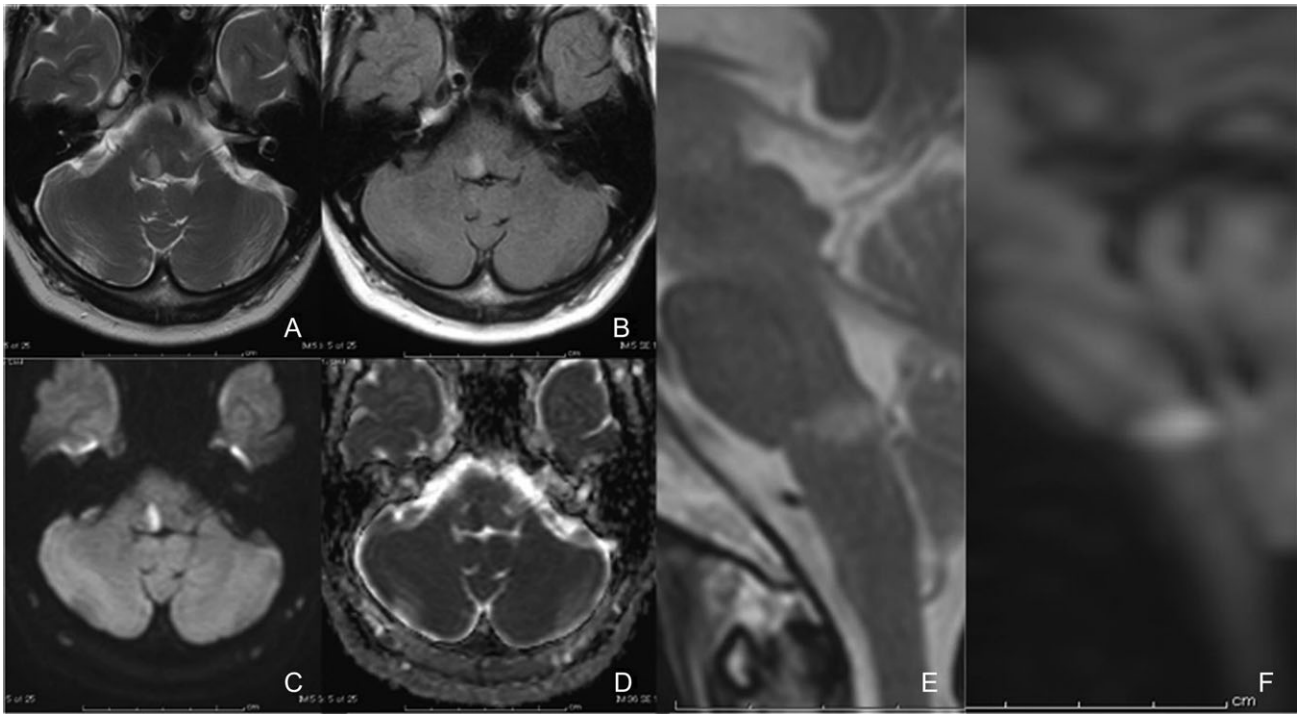

Figure 1: MR images show right medial pontomedullary junctional infarction: Each T2-weighted image (**A**), a fluid attenuation inversion recovery image (**B**), a diffusion weighted image (**C**) and an apparent diffusion coefficient map (**D**) indicate fresh ischemic lesion in axial sections. Sagittal sections of T2-weighted image (**E**) and diffusion weighted image (**F**) demonstrated, to the letter, pontomedullary junctional infarction

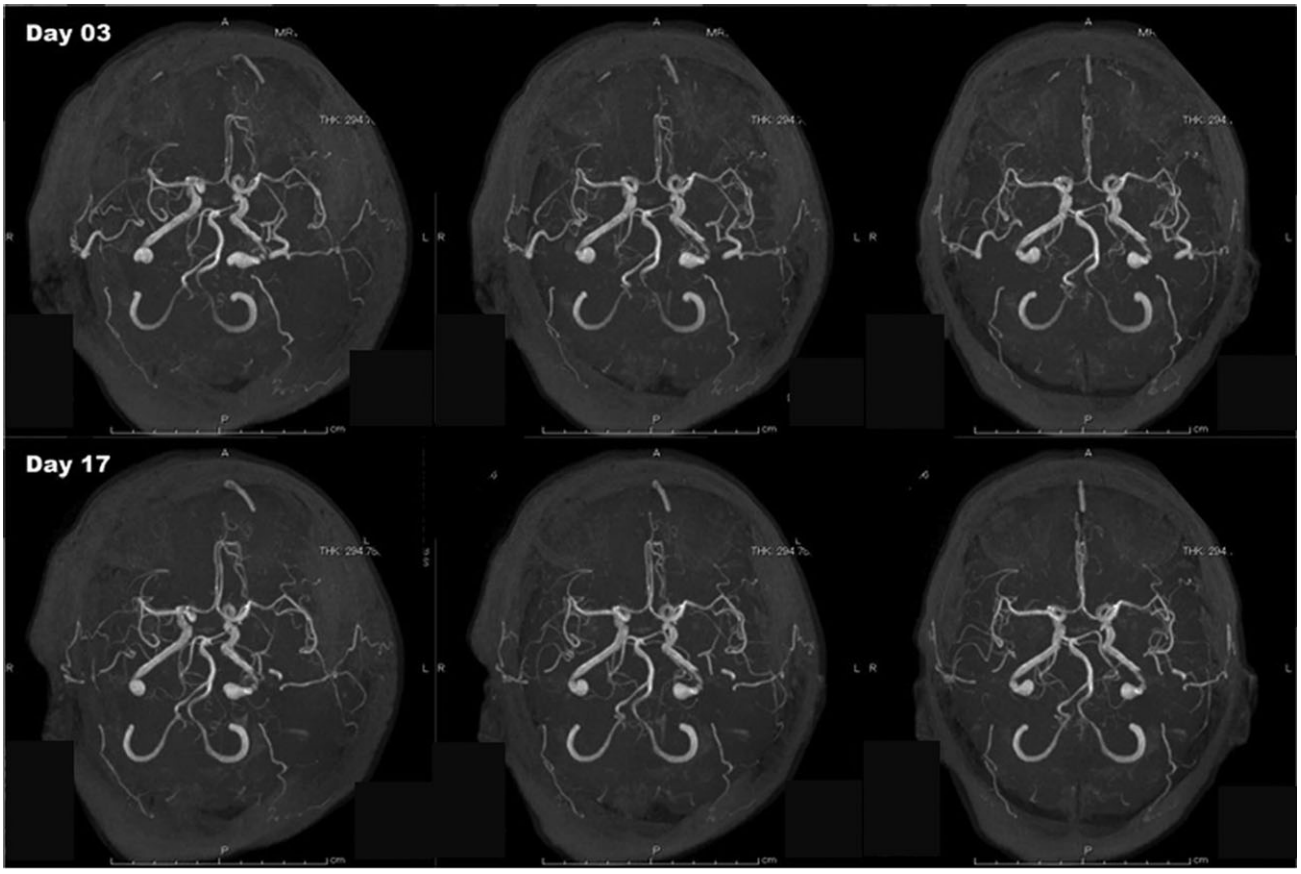

Figure 2: In comparison with the presymptomatic state (**A**), MR angiography shows no vertebrobasilar dissection or vasospasm/vasoconstriction but slightly stenotic change in the left vertebral artery on Day 03 (upper) with resolution on Day 17 (lower)

INTRODUCTION

Strokes in the distribution of the posterior circulation may present vertigo, imbalance and nystagmus [1]. A stroke involving the brain stem and cerebellum frequently presents acute vestibular syndrome [2]. Vascular vertigo is known to usually accompany other neurologic symptoms and signs [2]. Appropriate bedside evaluation is superior to MRIs for detecting central vascular vertigo syndromes [1]. Accurate identification of isolated vascular vertigo is very important because misdiagnosis of an acute stroke may result in significant morbidity and mortality [2].

In this report, we present a case of a localized small infarction in the right medial pontomedullary junction (MPMJ), manifested by ipsilateral facial paresis, contralateral hemiparesthesia of thermal hypoalgesia (TH) and dysphagia without curtain sign and hoarseness despite the lateral gaze being preserved bilaterally. To the best of our knowledge, through a survey of the literature, this MPMJ syndrome is a novel brain stem stroke

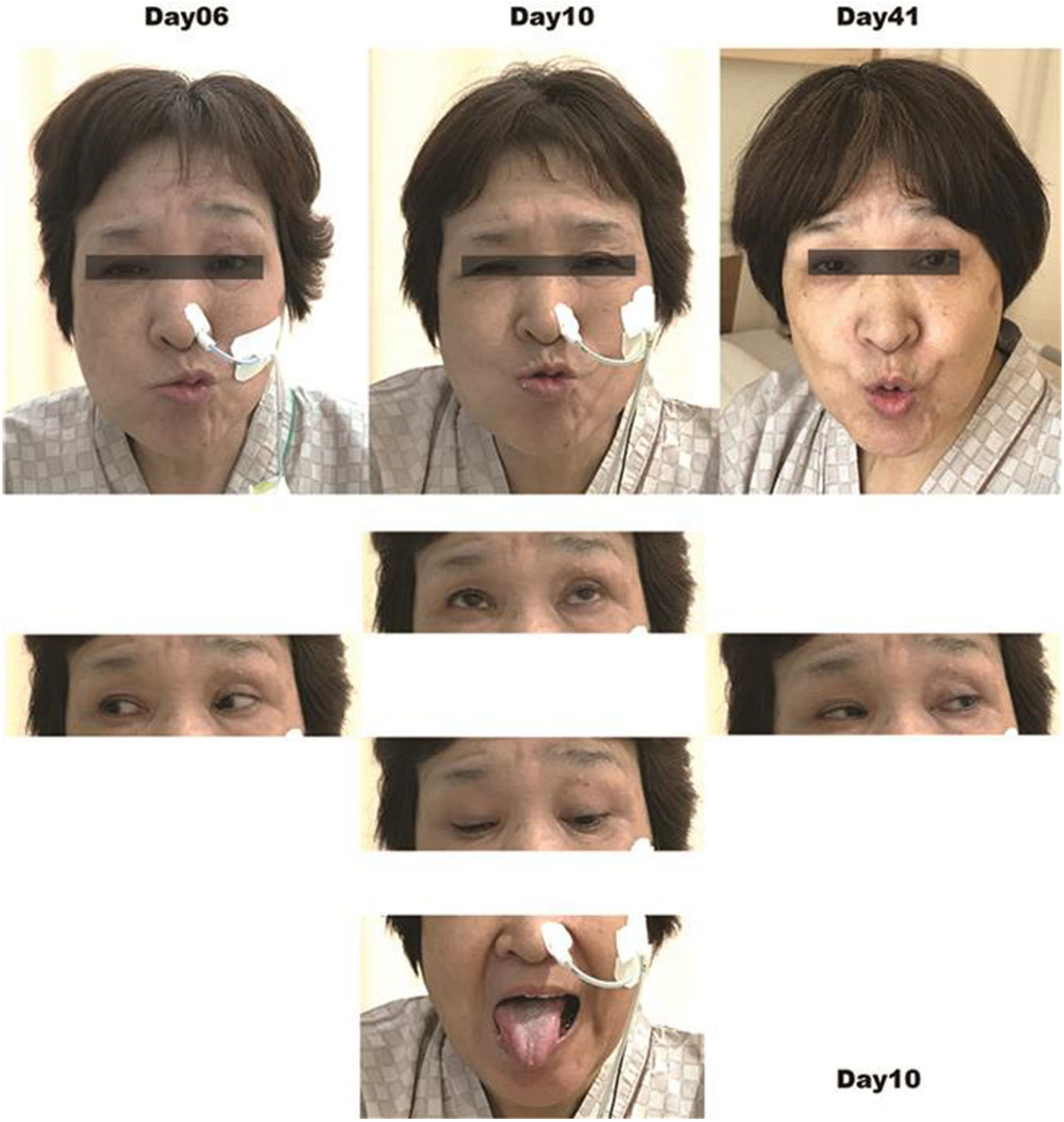

Figure 3: The right peripheral type facial paresis shows improvement gradually (upper). Extraocular movements show full range in all directions. The patient can protrude the tongue over the teeth without deviation from midline

syndrome, which is different from the previously reported brain stem stroke syndromes as discussed in detail later.

CASE REPORT

A 68-year-old hypertensive woman suddenly developed vertigo in the early morning when she went back to bed after micturition. She was brought to our hospital by ambulance. The medical history of the patient was unremarkable in spite of hypertension. On arrival, she was alert, cooperative, oriented, complaining of vertigo and suffering from nausea. A physical examination revealed right peripheral type facial paresis and left-beating nystagmus, despite no paresis nor ataxia of extremities. Horner's syndrome was not detected. The patient denied experiencing newly subjective hearing disturbance after onset of the vertigo. She noted the laterality of pain between the right arm and the left arm when her blood was being drawn through a sampling needle: less pain on the left arm. On the third hospital day, after settlement of vertigo and nystagmus, she confirmed TH on the left side of the body and face and severe dysphagia. Her extraocular movement was intact with full range. She complained of no diplopia. Curtain sign of the soft palate, as well as vocal cord paresis, was absent. Gag reflex was preserved. The tone of her voice was the same as it was before these symptoms developed. Magnetic resonance (MR) imaging showed a small infarction at the MPMJ (Fig. 1E and F). MR angiography did not show vertebrobasilar arterial dissection, severe thrombosis, nor apparent vasospasm (Fig. 2). Although her facial paresis ameliorated over the first week (Fig. 3, upper), her dysphagia did not. This right MPMJ infarction (Fig. 1E and F) caused right transient peripheral type facial paresis (Fig. 3, upper), left side TH of the body and face and severe dysphagia. The infarction (Fig. 4) did not cause lateral gaze palsy as right abducents palsy (Fig. 3, lower), hoarseness as glossopharybngeal nerve palsy, Horner's syndrome/curtain sign as vagus nerve palsy, or tongue deviation as hypoglossal nerve palsy (Fig. 3, lower), or left hemiparesis. Videofluorographic examinations showed consistent recovery from dysphagia caused by cricopharyngeal sphincter achalasia, evidenced by her improved ability to swallow (Fig. 5). Finally, through swallowing rehabilitation, her dysphagia regressed over 4 weeks in synchronization with the recovery of TH. She left our hospital with full independent activity of daily living despite the fact that her mild left TH-hemidysethesia continued.

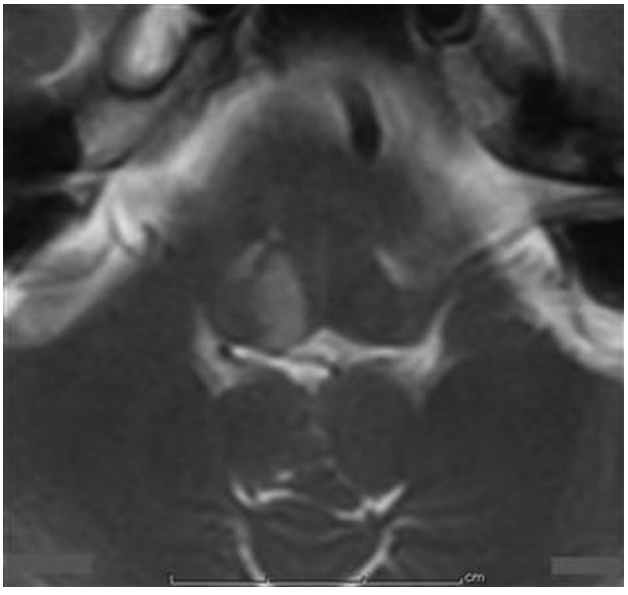

Figure 4: A magnified T2-weighted axial image shows localized infarction in the right pontomedullary junction

DISCUSSION

MPMJ syndrome, described in this report, is a unique syndrome of a brain stem stroke. The initial symptoms of unilateral MPMJ syndrome are acute onset vertigo, ipsilateral peripheral type facial paresis, dysphagia and contralateral hemidysesthesia (TH: deficits in pain and temperature sensation), all affecting the face as well as the body.

This unilateral MPMJ syndrome is not associated with hemiparesis, hemiataxia, conjugate gaze palsy, nor lower cranial nerve (IX–XII) palsy. These clinical manifestations distinguish the MPMJ syndrome from the previously reported pontomedullary syndrome. These manifestations include Millard–Gubler (facial palsy and contralateral hemiparesis) [3], Foville (facial palsy, conjugate gaze paralysis and contralateral hemiparesis) [3], Raymond–Cestan (internuclear ophthalmoplegia and contralateral hemiparesis) [3], Gasperini syndrome (ipsilateral impairment of the VI, VII and occasionally VIII cranial nerves and contralateral sensory loss) [4], Wallenberg (crossed hemisensory disturbance: sensory loss on the ipsilateral face and poor pain and temperature sensation on the contralateral body, ipsilateral Horner syndrome, vestibular symptoms: vertigo and nystagmus and ipsilateral cerebellar signs) [5], Dajuren (hypoglossal nerve palsy and contralateral hemiparesis) [6], Reinhold (ipsilateral hypoglossal nerve palsy, contralateral hemiparesis sparing the face and Wallenberg's syndrome) [7, 8], Babinski-Nageotte syndrome (contralateral hemiparesis sparing the face and Wallenberg's syndrome) [8–11] and Cestan-Chenais syndrome (all symptoms of the Babinski-Nageotte syndrome with the exception of the ipsilateral cerebellar hemiataxia) [11].

MPMJ syndrome should be distinguished from an incomplete form of Gasperini syndrome. Gasperini syndrome is a rare crossed brain stem syndrome characterized by ipsilateral impairment of the VI, VII and occasionally VIII cranial nerves and contralateral sensory loss [4]. The most frequent cause is the occlusion of the long circumferential branch of the anterior inferior cerebellar artery (AICA). The syndrome was initially described by Ubaldo Gasperini in 1912 [12, 13]. Gasperini syndrome results from a lesion of the caudal pons tegmentum [4]. Its core neurological signs are peripheral facial nerve palsy and abducens palsy of the affected side [14]. Thus, it is appropriate to differentiate the MPMJ syndrome from Gasperini syndrome despite the possible common cause of the branch occlusion of the AICA.

Although unilateral MPMJ syndrome does not accompany lower cranial nerve (IX–XII) dysfunction nor Horner's syndrome, videofluorographic examinations showed dysphagia caused by cricopharyngeal sphincter achalasia in the acute phase and its recovery in the subacute-chronic phase (Fig. 5, lower row). Since the patient's dysphagia regressed over four weeks in synchronization with recovery of TH, TH may have been related to cricopharyngeal sphincter achalasia and subsequent dysphagia (Fig. 5) in this case. Cricopharyngeal achalasia is a failure of the cricopharyngeus muscle to relax in response to a food bolus. In our case, the unilateral spinothalamic tract and the ipsilateral medial lemniscus were involved and recovered their function with concurrent recovery of TH. The neural impulse or inappropriate conductivity to or from the central nervous system may indirectly

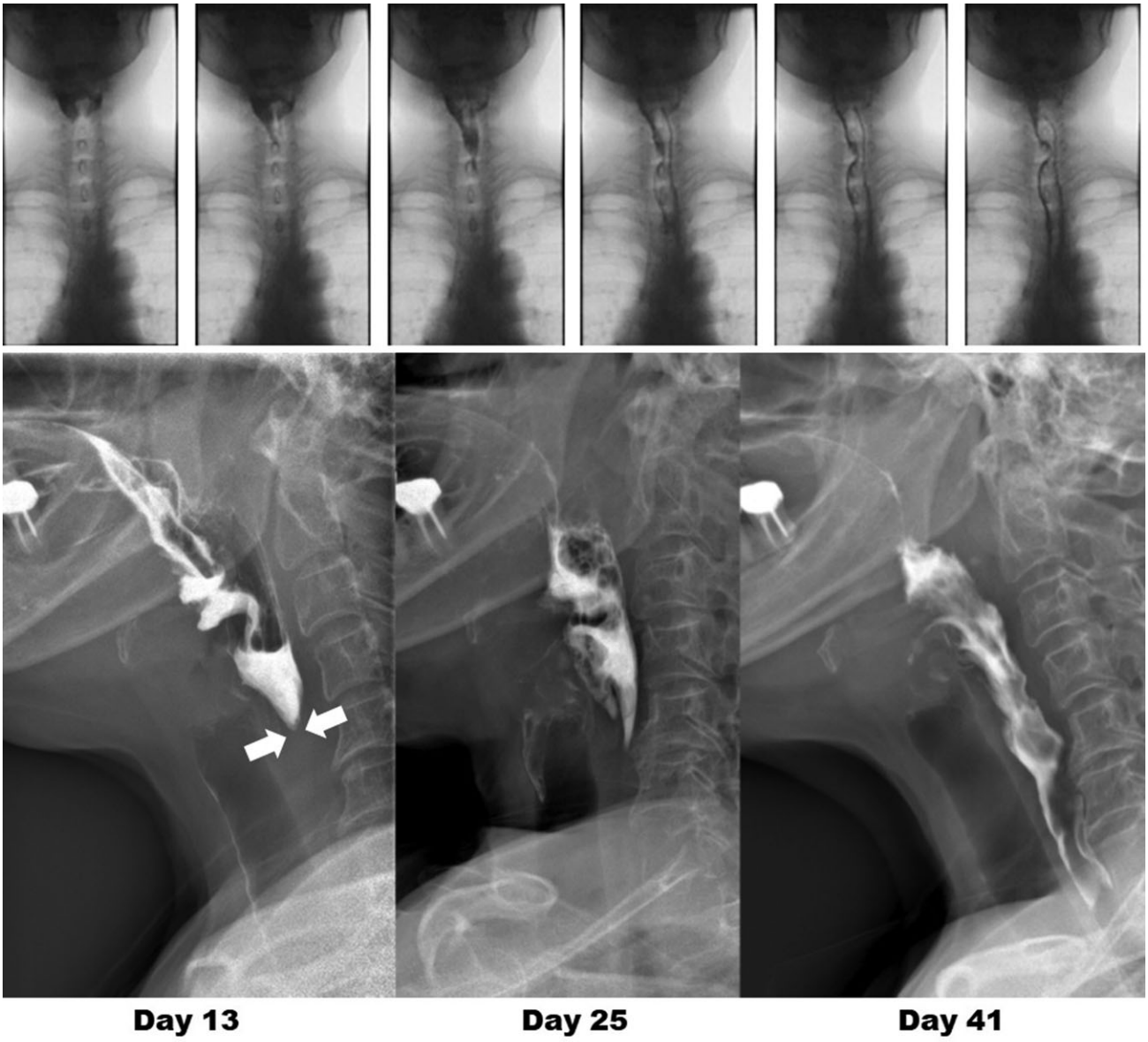

Figure 5: Anteroposterior view pictures of a videofluorography (50 flames per second, upper row) on the 25th day after onset show the laterality of the hypopharyngeal pyriform sinus during swallowing. They clearly show hypopharyngeal dysfunction in the left side. Cricopharyngeal achalasia (arrows) regresses over 4 weeks in synchronization with the recovery of thermal hypalgesia (lower row)

affect healthy peripheral striated muscular system [15]. This may be the same for the muscles involved in hyolaryngeal elevation [15]. Cricopharyngeal dysfunction (CPD) refers to incoordination of the cricophyngeal muscle either due to a primary functional disorder or as a result of an underlying neurological or medical condition [16]. CPD may have led to severe dysphagia and aspiration [16]. Symptoms of CPD can range from a globus sensation to oropharyngeal dysphagia manifested by regurgitation, coughing choking and recurrent aspiration [17]. The sensory stimuli that trigger and modulate swallowing include tactile stimuli (light and heavy pressure, air puffs, different bolus volumes and viscosities), chemical stimuli (water, other solutions, cations and anions), thermal stimuli and combined stimulus modalities [18–21]. Tactile-thermal application is a therapy technique designed to enhance the swallowing response in persons with dysphagia [22]. Latency to swallow-specific activity is significantly shorter following mechanical + cold + gustatory condition compared to no stimulation [22]. The clinical course of our case reflects the effectiveness of tactile-thermal application.

In unilateral MPMJ syndrome, ipsilateral facial sensation is preserved so that the spinal tract nucleus of the trigeminal nerve is spared. The ipsilateral peripheral type facial paresis is transient so that the nucleus of the facial nerve is spared. Hoarseness is unobservable so that the nucleus of the glossopharyngeal nerve is spared. Curtain sign, as well as Horner's syndrome, is absent so that the nucleus of the vagus nerve is spared. Tongue deviation is unaccompanied by MPMJ syndrome so that the nucleus of the hypoglossal nerve is spared. The contralateral hemidysesthesia (TH: deficits in pain and temperature sensation) has gradually improved so that the spinothalamic tract is involved but partially spared. These symptoms helped us to locate the damaged region (Fig. 4). Literally,

'MPMJ' indicates the responsible focus of the MPMJ syndrome (Fig. 4) and its presumable clinical manifestation.

In the unilateral MPMJ infarction, it was TH-related impairment of the swallowing reflex, rather than hemiparesthesia of TH, causing transient, albeit severe, dysphagia bothering the patient. As the TH got better, however, dysphagia improved more in this unilateral MPMJ infarction.

In conclusion, this MPMJ syndrome associated with the unilateral MPMJ infarction, including ipsilateral peripheral type facial paresis, contralateral TH of the body and face, severe dysphagia, but not ipsilateral abducens palsy nor contralateral hemiparesis, is a novel brain stem stroke syndrome different from those of Foville syndrome, Millard–Gubler syndrome, Wallenberg syndrome, Dajuren syndrome, Gasperini syndrome or others that were previously reported. Severe dysphagia is probably based on impaired swallowing reflex related to sensory dysfunction and bothered the patient more than TH-hemiparesthesia did. The facial paresis is transient as is vertigo.

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CONFLICT OF INTEREST STATEMENT

There are no conflicts of interest to declare.

FUNDING

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ETHICAL APPROVAL

No ethical approval was needed

CONSENT

Informed consent has been obtained from the patient.

GUARANTOR

Yuichiro Yoneoka

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