

Hemifacial spasm caused

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CASE REPORT

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Hemifacial spasm caused by tortuous verteobasilar artery: a case report

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Abstract

Background: Hemifacial spasm is a rare movement disorder. Prevalence estimates worldwide was 14.5 per 100,000 women and 7.4 per 100,000 men. Hemifacial spasm generally caused by compression of blood vessels at the root entry zone of the facial nerve in the brainstem, tortuous anteroinferior cerebellar artery (AICA) and posteroinferior cerebellar artery (PICA). Direct compression by verteobasilar dolichoectasia (VBD) with coincidence cavernoma is extremely rare.

Case report: A 50-year-old woman with right hemifacial spasm for 1 year, with a history of hypertension for 10 years, did not take medication regularly. MRI MRA was performed showing suspicious dolichoectasia in the verteobasilar artery and cavernoma in the left basal ganglia. Then digital subtraction angiography was performed, it was found that the tortuous verteobasilar junction artery with a curve to the right caused right hemifacial spasm.

Conclusion: Vascular imaging examination is important to do to find the cause of hemifacial spasm suspected to be due to vascular causes. The finding of two types of intracranial vascular malformations should be explored further. Therefore, the selection of therapy and management becomes more appropriate.

Keywords: Hemifacial spasm, Verteobasilar tortuous, Verteobasilar dolichoectasia, Cavernoma, 8th nerve compression

Background

Hemifacial spasm is a rare movement disorder, characterized by involuntary tonic or clonic contractions of the muscles innervated by the peripheral facial nerves [1–4]. Hemifacial spasm is divided into primary hemifacial spasm and secondary hemifacial spasm [1, 2, 4, 5]. Primary hemifacial spasm caused by compression of blood vessels or blood vessels ectasis aberrant the facial nerve in the root entry zone, which is most often anteroinferior cerebellar artery (AICA) and the superior cerebellar artery (SCA) and vertebral artery [1, 2, 4, 6]. Meanwhile, secondary hemifacial spasm, can be caused by a space-occupying lesion (SOL) at the cerebellopontine angle, can

also be caused by lesions in the brainstem such as stroke and multiple sclerosis [1, 2, 4]. Although hemifacial spasm is not life-threatening, this disorder causes psychosocial stress for the sufferer, so correct diagnosis and selection of appropriate management are important [2].

Hemifacial spasm, tinnitus, and other brainstem compression syndromes can be caused by tortuous arteries in the posterior circulation, commonly caused by the verteobasilar arteries, known as VBD [7]. VBD is part of intraarterial dolichoectasia. Intraarterial dolichoectasia can be acquired or hereditary [8]. Risk factors for acquired intraarterial dolichoectasia include aging, male gender and hypertension [8]. Intraarterial dolichoectasia can be caused by hereditary conditions, one of which is accompanied by a cavernous malformation [8].

To the best of our knowledge, we report the first case of hemifacial spasm and 8th nerve abnormality caused by compression of the verteobasilar tortuous artery,

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with coincidence finding cavernoma in basal ganglia. We discuss about the etiology, pathophysiology, whether the two vascular anomalies in this patient are hereditary or coincidental and the best treatment for our case.

Case report

A 50-year-old woman came to the neurology polyclinic of Dr. Soetomo Teaching Hospital Surabaya with complaints of right facial twitching in the last 1 year ago. Twitch movements occurred around the eyes, cheeks, and mouth; facial movements occurred spontaneously. The twitching improved during sleep and got worse when stressed, emotional and when talking. In the last 3 months, the patient also complained of vertigo, tinnitus, and decreased hearing. In the last 8 months, the patient also complained of weakness on the right side of the body, facial rot. The patient has a history of hypertension for 10 years ago and does not take medication regularly. The patient does not smoke and does not consume alcohol.

From the neurological examination, it was found that the *brow lift sign* or Babinski sign II was positive, right hemiparesis, right facial palsy of central type and right lingual palsy of central type. We had used GE Magnet Monitor 3, produced by GE Healthcare Technologies made in USA 2012 for the MRI and MRA. From the MRI examination with contrast and MRA examination, dolichoectasia was suspected from the verteobasilar junction artery to the basilar artery and a cavernoma in the left basal ganglia. In Fig. 1, T2W1 MRI in axial section



Fig. 1 Contact between facial nerve root entry zone and tortuous verteobasilar artery (arrow)

shows the root entry zone of the facial nerve in contact with the tortuous verteobasilar artery. Moreover, Fig. 2 shows the verteobasilar artery curves to the right (arrow) and with the bow is the area of contact between the verteobasilar artery and the facial nerve. On the coronal section in Fig. 2, the orange arrow shows the 8th nerve on the patient that also be compressed by tortuous verteobasilar artery, therefore, it causes tinnitus and vertigo.

Cavernoma in the left basal ganglia causes focal neurological deficits in the form of right hemiparesis, central type right facial nerve paresis. Then a cerebral DSA, used Phillips Allura FD10 model Xper table standard type: AD7 made in Holland 2018, was performed to explore the suspicion of dolichoectasia and also cavernoma on the MRI MRA image. From the results of the cerebral DSA, it appears that the tortuous verteobasilar artery junction with a curve to the right causes right hemifacial spasm. In Fig. 3, cerebral DSA shows a tortuous verteobasilar artery junction with a curve to the right (bow) causing right hemifacial spasm, arrow indicate AICA in this patient is not the cause of hemifacial spasm.

This patient received therapy with donazepam 2 mg twice daily, haloperidol 0.5 mg once daily, amlodipine 10 mg once daily and neurotropic. With the administration of conservative therapy for 1 month the patient experienced a significant improvement in complaints

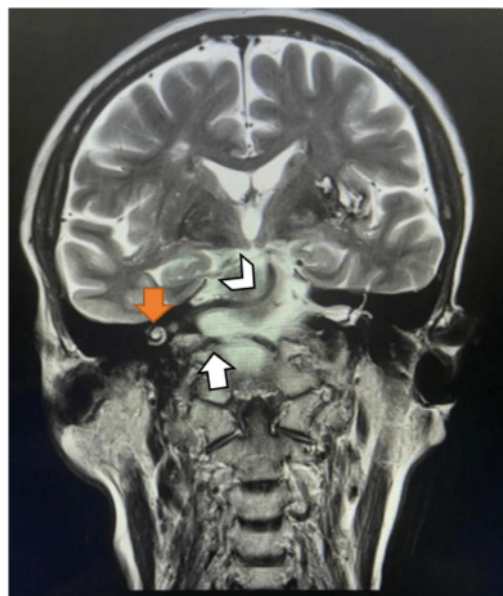


Fig. 2 The VBD (bow) and its compression (arrow), 8th nerve compression (orange bow)



Fig. 3 Tortuosity vertebral artery (bow) and AICA (arrow)

and a decrease in the frequency of movement disorder. Complaints of tinnitus and vertigo also improved with conservative therapy and blood pressure control. For cavernomas, although the lesions are symptomatic, observation is the preferred management strategy considering the absence of signs of bleeding in the cavernoma and the location of the cavernoma deep and in the eloquent cortex. Hemiparesis in the patient improved with medical rehabilitation therapy. Regular follow-up for complaints, increasing neurological deficits in patients and signs of bleeding. As a long-term follow-up, we planned repeat vascular imaging as a consideration for more aggressive management.

Discussion

HFS is a rare case of movement disorders [9]. Prevalence estimates worldwide HFS was 14.5 per 100,000 women and 7.4 per 100,000 men, with tendency of women 2:1 compared to men [1, 2]. HFS is most often caused by compression of blood vessels at the root entry zone of the facial nerve in the brainstem, such as that often leads to compression is AICA, SCA and posteroinferior cerebellar artery (PICA) which tortuous [1, 2, 4]. Direct compression by VBD is extremely rare, reported 0.7% of 1642 cases of HFS [6, 10].

Tortuous arteries often occurs in the cerebral basilar artery, communicating artery, the artery at the anterior and posterior cerebral, as well as in the arterioles in the white matter [10, 11]. Cerebral arteries may develop a tortuous condition due to increased flow associated

with elastin degradation or malformations [10, 11]. Severity of hypertension is associated with the development of tortuous arteries [10]. Increased age and male sex are also associated as risk factors for tortuous cerebral arteries [10–12]. Arteries are elongated and tortuous can cause disturbances in brain structure, especially in the pons and medulla, also cranial nerves. Therefore, it causes irritating symptoms such as hemifacial spasms and trigeminal neuralgia as well as tinnitus [10, 11].

Tortuous vertebral arteries are a form of the condition we refer to as vertebrobasilar dolichoectasia [8, 12]. VBD is a rare case. The incidence of VBD is known to vary between 0.06 and 5.8% [8, 12]. Clinical manifestations of VBD vary from asymptomatic, clinical manifestations due to VBD compression and clinical manifestations of ischemia [8, 12]. Systematic review on 375 patients VBD mention the 5-year risk for TIA by 10.1%, 17.6% for ischemic stroke, 10% to 5% compression of the brain and for the brain hemorrhage [8, 12].

Inherited factors have been implicated in the pathogenesis of dolichoectasia, including defects in the tunica media and damage to the elastic lamina, Ehlers Danlos syndrome, Marfan syndrome, tuberous sclerosis, and Moyamoya disease, in addition to acquired factors such as hypertension [8]. Cavernomas are caused by incorrect differentiation in the second stage of development of cerebral vessels in the embryological process [13]. During the second stage of development, arteries and veins are formed by the fusion of small vascular structures [13]. Dolichoectasia may be due to incorrect differentiation during the second stage of the same stage as cavernoma and therefore its etiology may be like that of cavernoma [13]. The cavernous hemangioma in our patient was in the basal ganglia, presents as a solitary lesion, and without a previous family history; whereas, dolichoectasia occurs at a location distant from the basal ganglia namely the vertebral artery. Therefore, the association of vertebral artery dolichoectatic and cavernous hemangioma in our patient may be the result of coincidence factors rather than hereditary factors. Therefore these become the difference with the previous case report that reported by Kanemoto and colleagues and also Al-Sharshah and colleagues [13, 14].

In the early stages of HFS patients when symptoms are still mild, and spasms occur infrequently/not often, effective oral medication should be considered [2, 4]. Benzodiazepine drugs such as clonazepam and carbamazepine have been reported to be effective [4]. Botulinum toxin injection is another safe and effective treatment option for hemifacial spasm [2]. Botulinum toxin is injected subcutaneously or intramuscularly into the involved facial muscles with an onset of action of 3–5 days and lasting

up to 3–6 months. After 3–6 months, botulinum toxin injection can be repeated [4, 6].

Surgical therapy is aimed at the compressive lesion causing HFS. Because surgical access in the posterior fossa is risky and adjacent to neovascular structures, the management of decompression in VBD is challenging [4]. Microvascular decompression technique for the management of hemifacial spasm has been reported to have good outcomes [4]. The MVD is the definitive treatment but carries the risk of facial nerve paresis and hearing loss, either temporary or permanent [4, 6]. In our case report, the patient improved with clonazepam and antihypertensive drugs. Control of blood pressure as a risk factor for tortuous vertebrobasilar artery in these patients also improved the symptoms of hemifacial spasm and also 8th nerve compression.

Conclusion

HFS can be caused by various causes divided into primary and secondary, which in our case was primary HFS caused by malformations of blood vessels or arteries that are tortuous or dolichoectasia. HFS accompanied by other symptoms of brainstem compression should be suspected due to a vascular malformation of the brainstem. Vascular imaging examination is important to do to find the cause of hemifacial spasm suspected to be due to vascular causes.

The findings of two types of vascular malformations simultaneously in the intracranial need to be explored whether due to hereditary or acquired factors, so that the involvement of vascular malformations in extracranial vessels can be explored further, so that unexpected complications can be prevented early. Furthermore, so that the selection of therapy and management becomes more appropriate.

Abbreviations

VBD: Vertebrobasilar dolichoectasia; DSA: Digital subtraction angiography; HFS: Hemifacial spasm; TIA: Transient ischemic attack; AICA: Anteroinferior cerebellar artery; SCA: Superior cerebellar artery; MRI: Magnetic resonance imaging; MRA: Magnetic resonance angiography; SOL: Space occupying lesion; PICA: Posteroinferior cerebellar artery; MVD: Microvascular decompression.

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Author contributions

MIT, DDY, PN, HAM was directly involved in the care of the patient, FIR performed DSA. MIT co-wrote the manuscript and DDY and FIR supervised and critiqued the final draft. All authors read and approved the final manuscript.

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Declarations

Ethics approval and consent to participate

We confirm that ethical clearance was not required for publication of this case report. Written informed consent in Indonesian language has been obtained from the patient before performing the procedures as a standard care in Teaching Hospital Dr. Soetomo Surabaya, East Java, Indonesia.

Consent for publication

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Competing interests

The authors declare they have no competing interests.

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