Multiple cranial palsy in pasein with cerebral cavernoma malformations on the brain stem: A rare case report

Wulandari
Neurology Resident of Neurology Department, Universitas Airlangga – Dr. Soetomo Teaching Hospital, Surabaya, Indonesia

Achmad Firdaus Sani
Teaching Staff of Neurology Department, Universitas Airlangga – Dr. Soetomo Teaching Hospital, Surabaya, Indonesia

Dedy Kurniawan
Teaching Staff of Neurology Department, Universitas Airlangga – Dr. Soetomo Teaching Hospital, Surabaya, Indonesia

Abstract—Facial palsy is a condition that is often encountered in neurological practice. The weakness of closing the eyes can be found in cases of central facial paralysis. Facial palsy is characterized by acute, unilateral, partial, or complete facial paralysis. Cerebral cavernous malformation is one rare form of lesion, accounting for about 5 to 13% of blood vessel anomalies in the central nervous system. Brain stem cavernomas are rare and less than 20% intracerebral cavernomas. Most patients with cerebral cavernoma of the brain stem have symptoms of acute neurological deficits. The inferior cerebellar peduncle, vestibular nucleus, trigeminal nucleus of the spine, and ambiguous nucleus are usually affected. Clinical signs include vestibulocerebellar symptoms such as vertigo, falling to the side of lesions, diplopia, multidirectional nystagmus, ipsilateral Horner syndrome, hiccups, contralateral body weakness, pain and sensation of temperature, motion, dysphonia, dysphagia, dysarthria, and decreased gag reflex (Sciaccata et al., 2019). A 26-year-old man with a right eye cannot close, the right face cannot be moved since 4 months, the right side face feels thick from the right cheek to the right chin. The patient has no previous history of stroke. In the examination obtained hipestesi NV2 and NV3 right, facial palsy peripheral right. MRI head with impressive contrast cavernous angioma pedunculus medulla oblongata right. Cerebral Cavernous Malformations has symptoms of cerebral hemorrhage, seizures, headaches and neurological deficits. In patients, rare symptoms of cerebral cavernous are peripheral facial palsy accompanied by paresis nervus
V2 and V3 and diagnosed as peripheral facial palsy. From imaging obtained cavernoma in the brain stem.

**Keywords**—Facial palsy, Cerebral cavernoma malformation, Brain stem.

**Introduction**

Facial palsy is a condition frequently encountered in everyday neurological practice. Central and peripheral facial palsy is easy to differentiate with the presence of upper facial weakness involved (Hebant et al., 2020; Lin et al., 2017). Peripheral facial palsy is characterized by acute, unilateral, partial, or complete facial paralysis. Peripheral facial palsy occurs in peripheral nerve weakness (Hebant et al., 2018; Hebant et al., 2020). Weakness may be partial or complete, and may be associated with mild pain, numbness, increased sensitivity to sound, and altered taste. Additional symptoms of peripheral facial palsy may include mild pain in or behind the ear, oropharyngeal or facial numbness, impaired tolerance to usual noise levels, and impaired taste in the front part of the tongue.

Cerebral cavernoma or cavernous angiomas is a cerebral cavernous malformation (CCM) that is a rare form of the lesion. According to autopsy reports, CCM occurs in 0.1% to 0.4% of the population and 8% to 15% of all cases of vascular malformation, 70 to 80% of which occur in the supratentorial. Most CCMs occur sporadically, but can be inherited in an autosomal dominant manner (Tumturk et al., 2018). Brainstem cavernomas are rare, accounting for less than 20% of intracerebral cavernomas. Approximately 57% of cavernomas occur in the pons, followed by the midbrain (14%), the pontomedullary junction (12%), and the medulla (5%). The incidence of rebleeding in patients with brainstem cavernoma is reported to occur in 30% of patients per year. A 20% mortality rate for patients with conservative management is reported, while the postoperative mortality rate is 0 to 1.9%. However, surgical therapy is technically associated with an additional risk of brainstem injury (Menon et al., 2011). Annual bleeding and recurrent bleeding occur in approximately 0.3% to 6.3% of non-brainstem cavernoma patients, and in approximately 2.8% to 32.3% of brainstem cavernoma patients (Menon et al., 2011; Taslimi et al., 2016). In addition, the outcome of untreated brainstem cerebral cavernoma may result in poor patient condition, as it can potentially lead to recurrent neurologic deficits and tend not to recover after multiple rheumatic attacks (Tumturk et al., 2018; Abla et al., 2011; Garcia & Ivan, 2015; Hauck et al., 2009).

Cerebral Cavernous Malformations are generally congenital lesions that occur at any age from the neonatal period to adulthood. Familial lesions are inherited in an autosomal dominant manner. The clinical symptoms usually include seizures (in 40-70% of patients), focal neurologic deficits (in 35-50% of patients), unspecified headaches (in 10-30% of patients), and cerebral hemorrhage (in 41% of patients) (Faria et al., 2004).
Cerebral Cavernous Malformations located in the brainstem almost never cause seizures, and most patients experience typical brainstem symptoms such as diplopia, ataxia, facial or body sensory impairments, or ataxia.

Cerebral Cavernous Malformations can occur in different regions of the brain, vary in size, and have different clinical features. Brainstem malformations have been very challenging for neurosurgeons. Due to its deep location and high surgical risk, conservative therapy has been the mainstay of treatment for decades (Tumturk et al., 2018).

**Case Report**

A 26-year-old man came to the neurology department with a complaint that his right eye could not close for the previous four months. The patient also complained that his right face could not move since the previous six months, the right side of the face felt thick starting from the right cheek to the right chin since October 2020, and all sounds that enter his right ear sound "shrill" since the previous four months. The patient also complained that he experienced difficulty speaking since the previous two months. The patient had no history of stroke, DM, HT, brain infection, and head trauma. The patient has been treated with mecobalamin and pain medication but experienced no improvement. He had a BP of 143/79 mmHg, a Pulse Rate of 84 x/minute, a RR of 20x/minute, a T of 36°C, and a GCS of 456. He also had lingual dysarthria, negative meningeal sign, nuchal rigidity, round pupil isocor of 3mm/3mm, light reflex +/+ , corneal reflex +/+ , hypesthesia nevus V2 and NV3 dextra, facial palsy dextra peripheral type, no lingual palsy, an upper motor of 5/5 and a lower motor of 5/5, normal sensory, physiological reflexes BPR +2/+2 TPR +2/+2 KPR +2/+2 APR +2/+2, Babinski pathological reflex -/- Chaddock -/-, Hoffman tromner -/-, normal cerebellar sign, and normal autonomic system. According to the MRI, the head with contrast indicated a right cavernous angioma pedunculus medulla oblongata (Fig. 1).

![Figure 1. MRI and MRA of the head with contrast coronal and axial sections](image-url)
The patient underwent conservative or nonsurgical therapy because of the deep position of the cavernoma and the high surgical risk.

**Discussion**

Cerebral Cavernous Malformations are generally congenital lesions that occur at any age from the neonatal period to adulthood. Familial lesions are inherited in an autosomal dominant manner (Zakaria et al., 2012). The clinical symptoms usually include seizures (in 40-70% of patients), focal neurologic deficits (in 35-50% of patients), unspecified headaches (in 10-30% of patients), and cerebral hemorrhage (in 41% of patients). Cerebral hemorrhage, epilepsy, headache, and neurological disorders are the main manifestations of this disease, but in our case report, there are other symptoms that are rarely complained of by cavernous hemangioma patients, namely facial palsy of the peripheral type and accompanied by paresis of the V2 and V3 nerves.

Cerebral Cavernous Malformations range in size from millimeters to centimeters. Little data have been found regarding the size of this malformation. Most cavernous malformations are small but can reach significant sizes. Giant cavernous malformation (GCM) has a diameter greater than 6 cm and is relatively rare. The clinical symptoms of GCM are not different from those of CCM.

Facial palsy is a condition frequently encountered in neurological practice. Differentiating peripheral facial palsy (PFP) from central facial palsy (CFP) is relatively easy and depends on the involvement of the upper face. Indeed, upper and lower facial weakness suggests a diagnosis of PFP, whereas isolated involvement of the lower face suggests a diagnosis of CFP. However, contrary to basic knowledge, upper facial involvement can also be encountered in cases of CFP, as in peripheral type facial palsy. This dissociation must be due to the separate origins of the corticofacial projections. The nervous systems that affect involuntary movements originate from the caudal cingulate motor cortex, whereas, those that affect involuntary movements can originate from the main motor cortex. It should be noted that in cases of facial palsy with upper facial dysfunction, dominant involvement of the lower face, pointing towards CFP rather than PFP, is required for brain imaging (Hebant et al., 2020).

The non-hemorrhagic focal neurologic deficit is defined as focal neurologic signs or symptoms without evidence of bleeding. Chronic headache is common in patients with Cerebral Cavernous Malformation, but in the absence of acute bleeding associated with imaging, chronic headache is not considered a symptomatic sequela of Cerebral Cavernous Malformation requiring surgery (Flemming et al., 2020).

**Conclusions**

Cerebral cavernoma malformations can show a variety of clinical symptoms on the cranial nerves, and one of the rarely encountered symptoms in the brainstem is peripheral facial palsy, thus requiring a careful clinical examination supported by complete supporting examinations. These procedures are performed to allow
early diagnosis and avoid misdiagnosis so that patients receive the right treatment options immediately.

References


