Cystic Progression of a Cavernous Malformation at the Level of the Trigeminal Root Entry Zone Presenting With Sudden Onset of Trigeminal Neuralgia

Carlo Giacobbo Scavo, MD, Raffaelino Roperto, MD, Guglielmo Cacciotti, MD, and Luciano Mastronardi, MD, PhD

Background: Cavernous malformations (CM) in the cerebellopontine angle (CPA) are rare, and most of them are solid and extend from the internal auditory canal into the CPA. In contrast, cystic CM arising in the CPA and not involving the internal auditory canal and dura of the skull base are extremely rare. The authors present an uncommon large cystic progression of a cavernous malformation at the level of the trigeminal root entry zone evolving to severe trigeminal neuralgia and brainstem compression.

Methods: A 62-year-old female presented a sudden onset of left trigeminal neuralgia, caused by a large cystic lesion at the level of the root entry zone of the left 5th nerve. On neurological examination, she showed slight gait ataxia and hypoesthesia on the left hemiface (on the first and second trigeminal branches). Other cranial nerves were in order. Magnetic resonance imaging showed a large cystic intracranial mass, with a small solid portion, leading to brainstem compression.

Results: Microsurgical removal of the lesion was performed via retrosigmoid approach, with intraoperative monitoring of somatosensory evoked potentials, facial, and cochlear nerves. The posterior-medial portion of the lesion was solid, whereas the main portion was cystic, containing xanthochromic fluid. The small solid lesion continued with a thin capsule of a large cyst adherent to brainstem, cerebellar hemisphere, and trigeminal nerve entry zone. A big draining vein arising from the solid part of the lesion runned parallel to brainstem. The mass was piecemeal totally removed.

After surgery the patient recovered both left trigeminal neuralgia and hypoesthesia; ataxia was significantly relieved too. Postoperative magnetic resonance imaging confirmed the total removal. Histopathological features were consistent with a CM. At 6-month follow-up, patient's symptoms at the presentation had resolved.

Conclusion: The authors present a very rare patient of large cystic cavernous malformation at the level of the trigeminal root entry zone presenting with sudden onset of trigeminal neuralgia. Even if it has not established imaging features, a cystic cavernoma of the

From the Division of Neurosurgery, San Filippo Neri Hospital, ASL Rome, Rome, Italy.

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Address correspondence and reprint requests to Luciano Mastronardi, MD, PhD, San Filippo Neri Hospital, Via Reno 14, 00198 Rome, Italy; E-mail: mastro@tin.it

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cerebello-pontine angle may be suspected when a cystic mass is present, not involving the internal acoustic meatus nor the skull base dura mater. Careful microneurosurgical technique and monitoring of cranial nerves allow good long-term results.

Key Words: Brainstem cyst, cavernous malformation, retrosigmoid approach, root entry zone, trigeminal neuralgia

C avernous malformations (CM) or cavernomas represent the second most common intracranial cerebrovascular lesion, accounting for 10% to 15% of all vascular malformations of the central nervous system.^{1–3}

About 40% of the CM are silent.³ When symptomatic, the clinical picture includes intracerebral hemorrhage (36%-56%), epilepsy (20%-50%), focal deficits (20%-45%), and headache (6%-34%).¹⁻³ Cranial nerves or nerve roots involvement by CM is very rare.¹⁻¹¹

Intracranial CM are commonly located in the supratentorial region, basal ganglion, brain stem, and cerebellar hemispheres. Cavernous malformations arising in the cerebellopontine angle (CPA) are rare and the majority of them are solid lesions arising from the internal auditory canal, extending to the CPA.^{1–11} Among them, cystic CM in the CPA are really uncommon.^{4,6,8,10,11}

Trigeminal neuralgia is commonly caused by a microvascular conflict with the fifth cranial nerve in the CPA, at the level of its root entry zone.^{12,13} Occasionally, it is secondary to compression of the fifth nerve by intra- or extra-axial pathologies such as tumors or vascular lesions.^{2,5}

We report the exceptional case of a patient who presented with sudden onset of left trigeminal neuralgia, facial numbness, and ataxia caused by a large cystic cavernous angioma with small solid portion, at the level of the root entry zone of the fifth cranial nerve, with marked brainstem compression, which was treated surgically with complete resolution of preoperative symptoms.

PATIENT PRESENTATION

A 62-year-old female was admitted in our department for the sudden onset of severe left trigeminal neuralgia (mainly 1st and 2nd branches). On neurological examination, she showed a slight gait ataxia and hypoesthesia on the left hemiface (except for the third trigeminal branch). Other cranial nerves were in order.

The c.e. cerebral magnetic resonance imaging (MRI) (Fig. 1A and B) showed a large cystic intracranial mass in the left side of brainstem, at the level of the entry zone of left trigeminal nerve, with a small solid portion that appeared hyperisointense on T1WI and hyper-hypo-intense on T2WI, leading to brainstem compression. Hemosiderin greater susceptibility sequence (Gradient Echo Sequences) depicts high degree of hemosiderin deposits indicating probable CM. The lesion did not appear to extend into the internal auditory canal, and no dural tail was present.

Microsurgical removal of the lesion was performed by left retrosigmoid approach, with the patient in lateral position; intraoperative somato-sensory evoked potentials, facial, and cochlear nerves monitoring were recorded. The posterior-medial portion of the lesion was solid and showed signs of cystic changes, whereas the main portion of the lesion was cystic, containing xanthochromic fluid. The small solid lesion appeared to be reddish, well margined, firm, vascular, continuing with a thin capsule adherent to brain stem, cerebellar hemisphere, and trigeminal nerve.

In particular, the cystic part was attached to the trigeminal nerve root, between the cisternal and intra-axial portions of the nerve. In addition, the lesion was in strict contact with facial and cochlear nerves too. A big draining vein arising from the solid part of the lesion ran parallel to the brainstem in the caudal direction.

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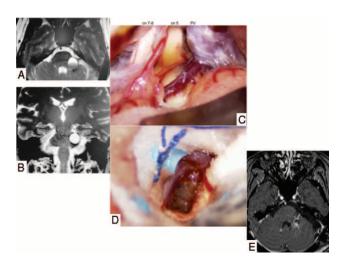


FIGURE 1. (A) Preoperative MRI axial image. (B) Preoperative MRI coronal image. Large cystic lesion with deep solid part in the left CPA, at the level of trigeminal root entry zone, compressing brain stem and cerebellum. The maximum diameter of the lesion was about 3 cm. The solid component showed hypo- and iso-intensity on T1WI images, without significantly enhancement after contrast medium administration. (C) Intraoperative view: trigeminal nerve (cn 5) and its root entry zone, facial and vestibular-cochlear bundle (cn 7–8), and petrosal vein (PV) were crossed by a longitudinal vein on the brainstem. (D) Intraoperative view: reddish membrane of the large cyst in the midbrain found after medial mobilization of the longitudinal vein. (E) Postoperative MRI confirmed the removal of the large cystic lesion and the persistence of the satellite vein surrounded by a small caput medusae. CPA, cerebellopontine angle; MRI, magnetic resonance imaging.

Mobilizing the vein, the cystic wall of the lesion was exposed. After the detachment of the cyst wall from the adjacent brainstem nervous tissue and cranial nerves entry-exit zones with the microneurosurgical technique, it was piecemeal totally removed, including the solid portion (Fig. 1C and D).

On histological examination the solid part showed to be a cavernous malformation.

Postoperative course was uneventful and both left trigeminal neuralgia and hypoesthesia recovered; ataxia improved significantly too.

Postoperative MRI demonstrated the complete resection of the lesion and the persistence of the satellite vein surrounded by a small caput medusae (Fig. 1E). At 6 months follow-up, the patient was symptom-free.

DISCUSSION

Cavernomas are low-flow vascular malformations that can occur single or multiple.^{1,3} Classically CM are described as congenital lesions resulting from an alteration in the development of arteriole-capillary network, but modern studies have shown this to be dynamic lesions, which can vary in size.^{1,3}

Cavernomas consist of dilated vascular channels of different sizes, which are lined by a thin and weak epithelium,³ lacking in muscular and elastic layers, which predisposes to bleeding. They may have internal calcifications, but its main histological feature is that there is no brain parenchyma between them, unlike capillary telangiectasia.³

Cavernous malformations are commonly located in supratentorial areas and occasionally in the brain stem, cerebellum, on cranial nerves, dura mater, and venous sinuses.^{1,2,3,5} However, the occurrence of CM in the CPA is rare, and most of those reported to date are solid and extend from the internal auditory canal into the CPA.^{1,3,5–9,11} In contrast, CM in the CPA not involving the internal auditory canal and dura of the skull base are extremely rare.^{1,6,10,11} In particular, cystic CM of the CPA are exceptional: including the present case, in the international literature only 6 patients are described in detail.^{4,6,8,10,11}

The most common cause of idiopathic trigeminal neuralgia is microvascular compression of the fifth nerve at the level of its entry zone in the brainstem.^{12,13} As described in detail by Xia et al¹² in their systematic review, usually the trigeminal nerve is compressed by the superior cerebellar artery and, less frequently by the anterior-inferior cerebellar artery or by a vein, or by a combination of these vessels.^{12,13} Secondary causes of trigeminal neuralgia include multiple sclerosis, schwannomas of trigeminal or vestibular nerves, meningiomas, epidermoid cysts, brainstem gliomas, lipomas, arachnoid cysts, metastases, lymphomas, arteriovenous malformations, aneurysms, vertebrobasilar ectasia, chordomas, and Chiari I malformations.¹³ Only a few reports have described trigeminal neuralgia related to a CM or even more rare to a cystic CM.^{2,5,7}

Huang et al⁴ reported 1 patient of cystic CM in the CPA with review of the literature, analyzing a total of 5 patients.^{6,8,10,11} According to them, the causes of cystic degeneration of CMs remain unknown.⁴ Recurrent minor hemorrhage of internal vascular sinuses or neo-capillaries within CM could be involved in this process. When bleeding episodes occur within a CM, the osmotic pressure across the membranes of intralesional vessels could change and lead to gradual fluid accumulation within the CM evolving to cystic degeneration, followed by CM growth.^{4,6,8,10,11} Consequently, the CM may show various MRI features of cystic degeneration.^{3,4,6,8,10,11}

Diagnosis of cystic CM should be suspected when a cystic mass with no involvement of internal auditory canal and skull base dura is found in the CPA. 3,4,6,8,10,11

Magnetic resonance imaging is the main technique for the diagnosis of cavernomas.^{1,3} Its radiographic appearance is variable depending on the stage of hemorrhage. The classical image in uncomplicated CM is known as "popcorn," with a central area of heterogeneous signal on T1- and T2-WI, surrounded by a ring of hemosiderin with less intense signal in T2WI.³ The most sensitive sequence to detect cavernomas is the gradient echo T2, because of magnetic susceptibility of products generated by degradation of hemoglobin.^{1,3}

The present case confirms that, when possible, total excision of symptomatic brainstem cavernomas is the best treatment.^{1,2,4,6,8,10,11} Even if extremely rare, it is noteworthy to consider CM at the level of the root entry zone of fifth cranial nerve as a possible cause of trigeminal neuralgia. In particular, a cavernoma should be suspected when a cystic lesion in the CPA is found on MRI.

CONCLUSION

The intra-axial compression of the trigeminal nerve by a brainstem cavernoma can be the cause of trigeminal neuralgia. We present an exceptional large cystic progression of a cavernous malformation at the level of the trigeminal root entry zone evolving to severe and acute trigeminal neuralgia and brainstem compression. A CM should be suspected when a cystic lesion is present in the cerebello-pontine angle, not involving internal acoustic meatus nor the skull base dura mater. Careful microneurosurgical technique and intraoperative monitoring of cranial nerves allow good short- and long-term results.

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