Carotid-cavernous fistula with brainstem congestion mimicking tumor on MRI

Article abstract—A 65-year-old woman presented with a left abduction deficit and "red eye," mild proptosis, chemosis, arterialization of the conjunctival vessels, intention tremor, and bilateral pyramidal signs. MRI showed significant left-sided brainstem involvement that mimicked a tumor. Right hemiplegia ensued 1 week later. Venous congestion of the brainstem with hemiplegia resulting from shunting of blood flow from both carotid arteries is an extremely rare complication of carotid-cavernous fistula.

NEUROLOGY 2000;55:1929-1931

Shuzo Shintani, MD; Shin Tsuruoka, MD; and Tatsuo Shiigai, MD

Dural carotid-cavernous fistula (CCF) is an abnormal communication between the dural branches of the internal or external carotid arteries and the cavernous sinus. This communication commonly results in chemosis, proptosis, a bruit,¹ and, less commonly, ophthalmoplegia.²⁻⁵ However, venous congestion of the brainstem causing hemiplegia is extremely rare as a complication of CCF.

Case presentation. A 65-year-old woman with an 8-month history of chemosis and "red eye" (figure 1) and a 2-month history of vertigo was admitted to our hospital with a chief complaint of diplopia. Medical and family history were unremarkable. On admission, her blood pressure was 135/82 mm Hg; her heart rate was 76/minute with a regular rhythm. The patient was alert and exhibited no motor or sensory deficits in any extremity. Neurologic abnormalities included a left abduction deficit with a partial bilateral horizontal and upward gaze palsy (see figure 1), bilaterally increased deep tendon reflexes, bilateral Babinski signs, and a left-sided intention tremor evident on performing the finger-to-nose test.

MRI on admission showed a mildly hypointense pontine lesion on T1-weighted images that became hyperintense on T2-weighted images. This lesion had an indistinct border and was located predominantly on the left side (figure 2A). Contrast-enhanced T1-weighted images revealed marked enhancement, demarcated by the pontine raphe, on the left side of the pons (figure 2B).

The following laboratory examinations on admission revealed no abnormalities: erythrocyte sedimentation rate, C-reactive protein, complete blood cell counts, urinalysis, blood chemistry, blood sugar concentration, coagulation tests, and immunologic studies.

One week after admission, the patient developed right hemiplegia. Cerebral angiography showed no abnormalities involving the vertebrobasilar arteries. However, a left carotid angiogram (figure 3A) showed a left-sided CCF, and a right carotid angiogram (figure 3, B and C) revealed that shunted blood flow in the CCF was supplied predominantly from the right carotid siphon. The CCF was associated with venous drainage into the inferior petrosal sinus.



Figure 1. A 65-year-old woman presented with a left abduction deficit with a partial bilateral horizontal and upward gaze palsy, mild proptosis, chemosis, and arterialization of the conjunctival vessels (spiral vessels) on admission. Top to bottom: left gaze, right gaze, forward gaze, upward gaze, and downward gaze.

From the Departments of Neurology (Dr. Shintani), Neurosurgery (Dr. Tsuruoka), and Internal Medicine (Dr. Shiigai), Toride Kyodo General Hospital, Toride City, Ibaraki, Japan.

Received February 9, 2000. Accepted in final form August 17, 2000.

Address correspondence and reprint requests to Dr. Shuzo Shintani, Department of Neurology, Toride Kyodo General Hospital, 2-1-1 Hongoh, Toride City, Ibaraki 302-0022, Japan; e-mail: dw4s-sntn@asahi-net.or.jp



Follow-up MRI revealed that the lesion gradually involved the medulla oblongata in the T2-weighted images.

Three months later, the patient manifested CO_2 narcosis (PaO₂ 67.9 mm Hg and PaCO₂ 73.8 mm Hg on room air) and became comatose. Mechanical ventilation was performed for 2 weeks, but blood pressure dropped despite IV dopamine administration. She died of respiratory and circulatory failure before an embolization procedure could be performed. An autopsy was refused.

Discussion. MRI in the current patient showed a sizable pontine lesion with an indistinct border on T2-weighted images. Yet she had only vertigo as a brainstem symptom at the time of admission, with diplopia. The clinical and radiologic findings supported a tentative diagnosis of brainstem tumor.

However, the brainstem lesion was mildly hypointense on noncontrast T1-weighted images, and showed marked enhancement, demarcated by the pontine raphe, on contrast-enhanced T1-weighted images. The medial border on the postcontrast T1 images is highly atypical for tumor. These features indicated that the pontine lesion was likely to be of Figure 2. Axial noncontrast MRI on admission showed a prominently hyperintense lesion (arrowhead) with an indistinct border and predominantly leftsided location on a T2-weighted image (A). A contrast-enhanced T1-weighted image showed marked enhancement (arrowhead) of the left side of the pons, demarcated by the pontine raphe (B).

vascular origin, and this appearance is in accord with previously reported findings in dural arteriovenous fistula (DAVF) associated with venous congestion.^{6,7}

The initial report describes two cases of DAVF with convergence of venous drainage into the basal vein of Rosenthal or cortical veins of the posterior fossa, resulting in pontine venous congestion.⁶ The second report is of two patients with DAVF of the cavernous sinus with venous congestion of the brainstem who showed significant improvement of a pontine lesion after transvenous embolization, as seen on T2-weighted images.⁷

These previously reported CCF cases included chemosis and ophthalmoplegia, but lacked hemiplegia or brainstem dysfunction despite a sizable pontine lesion apparent on T2-weighted images.^{6,7} Our case was similar in its initial presentation.

As described in figure 1, there was a partial bilateral horizontal and upward gaze palsy with a marked left abduction deficit. The limitation of eyeball movements is thought to be caused by mechani-



Figure 3. Lateral view in a left carotid angiogram (A) showed a left-sided carotid-cavernous fistula (arrowheads). Posterior-anterior view of a right carotid angiogram (B) showed shunting of blood flow to the left cavernous sinus. The abnormal flow originated predominantly from the right side (paired arrowheads). Lateral view (C) showed the fistula (paired arrowheads) to drain into the inferior petrosal sinus (triple arrowheads).

1930 NEUROLOGY 55 December (2 of 2) 2000

cal compression of III, IV, and VI nerves in the cavernous sinus, ischemia of the nerve nutrition vessels, or the involvement of these cranial nerve nuclei in the brainstem in this patient.

Our patient died of respiratory and circulatory failure 3 months after admission, before embolization could be performed. Follow-up MRI revealed that the lesion gradually involved the medulla oblongata in the T2-weighted images. We suggest that the expansion of the lesion to the medulla resulted in respiratory and circulatory failure.

Mechanisms of venous congestion in CCF remain unknown. CCF-related pontine lesions seen by MRI are potentially reversible by embolization,⁷ supporting a pathogenetic hypothesis involving vasogenic edema rather than irreversible ischemic change. In the current case, cerebral angiography revealed bilateral shunting of blood via the left-sided CCF, with normal vertebrobasilar vessels. Because of bilateral shunting, our patient may have had a greater overall amount of shunting than others with unilateral shunting. We believe that venous hypertension and passive congestion due to an exceptionally large flow volume shunted via the CCF resulted in the hyperintense pseudoneoplastic lesion seen on T2-weighted images.

References

- 1. Newton TH, Hoyt WF. Dural arteriovenous shunts in the region of the cavernous sinus. Neuroradiology 1970;1:71-81.
- Kosmorsky GS, Hanson MR, Tomsak RL. Carotid-cavernous fistulae presenting as painful ophthalmoplegia without external ocular signs. J Clin Neurol Ophthalmol 1988;8:131–135.
- Selky AK, Purvin VA. Isolated trochlear nerve palsy secondary to dural carotid- cavernous sinus fistula. J Neuro-Ophthalmol 1994;14:52–54.
- Watanabe S, Takahashi A, Mochizuki H, Takase S, Itoyama Y. Cavernous sinus dural arteriovenous shunts presenting with isolated trochlear nerve palsy as an initial sign. Neurol Med (Tokyo) 1999;51:79-84.
- Inoue H, Udaka F, Kameyama M. Carotid-cavernous fistula presenting with abducens nerve palsy. Neurol Med (Tokyo) 1995;43:262-264.
- Uchino A, Kato A, Kuroda Y, Shimokawa S, Kudo S. Pontine venous congestion caused by dural carotid-cavernous fistula: report of two cases. Eur Radiol 1997;7:405-408.
- Takahashi S, Tomura N, Watarai J, Mizoi K, Manabe H. Dural arteriovenous fistula of the cavernous sinus with venous congestion of the brain stem: report of two cases. AJNR Am J Neuroradiol 1999;20:886–888. 2000

Identical dysferlin mutation in limbgirdle muscular dystrophy type 2B and distal myopathy

Article abstract—Limb-girdle muscular dystrophy type 2B (LGMD2B) and Miyoshi myopathy (MM) are autosomal recessive disorders caused by mutations in the dysferlin gene on chromosome 2p13. The authors studied a large Russian family with both LGMD2B and MM. All affected individuals, as well as one preclinical boy with dystrophic changes on muscle biopsy, were found to be homozygous for a novel dysferlin mutation, TG573/574AT (Val67Asp). This finding supports the view that additional factors (e.g., modifier genes) contribute to the phenotypic expression of causative mutations in dysferlinopathies.

NEUROLOGY 2000;55:1931-1933

S.N. Illarioshkin, MD, PhD; I.A. Ivanova–Smolenskaya, MD, PhD; C.R. Greenberg, MD, CM; E. Nylen; V.S. Sukhorukov, MD, PhD; V.V. Poleshchuk, MD; E.D. Markova, MD; K. Wrogemann, MD, PhD

Thus far, eight genetic loci have been identified in autosomal recessive muscular dystrophies, with causative genes known for seven forms.¹ Clinical presentations in most of these forms correspond to a phenotype of limb-girdle muscular dystrophy

Received February 22, 2000. Accepted in final form August 2, 2000.

(LGMD). However, a locus on chromosome 2p13 was shown to be associated with two distinct clinical phenotypes, either LGMD type 2B² or Miyoshi myopathy (MM),³ the latter disease being a rare form of distal myopathy with predominant involvement of the calf muscles. Furthermore, two consanguineous families were reported in which different family members homozygous for the same 2p13 haplotypes exhibited either LGMD or distal myopathy.^{4,5}

Mutations in a novel gene, dysferlin, were shown to be associated with different phenotypes of 2p13linked muscular dystrophies.^{6,7} In the large Canadian kindred with both LGMD and MM, all affected members were reported to carry the same missense mutation in the homozygous state, which resulted in similar reductions of dysferlin expression in both types of patients.⁸ No clear explanation exists for the remarkable inter- and intrafamilial variation of dys-

From the Department of Neurogenetics (Drs. Illarioshkin, Ivanova–Smolenskaya, Poleshchuk, and Markova), Institute of Neurology, Russian Academy of Medical Sciences; Department of Pathomorphology (Dr. Sukhorukov), Institute of Paediatrics and Child Surgery, Moscow, Russia; and the Departments of Biochemistry and Medical Genetics (Drs. Greenberg, Nylen, and Wrogemann), and Pediatrics and Child Health (Drs. Greenberg and Wrogemann), University of Manitoba, Winnipeg, Canada.

Supported by grants from the Muscular Dystrophy Association, the Medical Research Council of Canada, and the Children's Hospital Foundation of Manitoba.

Address correspondence and reprint requests to Dr. Sergei N. Illarioshkin, Department of Neurogenetics,, Institute of Neurology, Russian Academy of Medical Sciences, Volokolamskoye Shosse 80, Moscow 1233671, Russia; e-mail: neurogen@online.ru



Carotid-cavernous fistula with brainstem congestion mimicking tumor on MRI Shuzo Shintani, Shin Tsuruoka and Tatsuo Shiigai *Neurology* 2000;55;1929-1931 DOI 10.1212/WNL.55.12.1929

Updated Information & Services	including high resolution figures, can be found at: http://www.neurology.org/content/55/12/1929.full.html
References	This article cites 5 articles, 1 of which you can access for free at: http://www.neurology.org/content/55/12/1929.full.html##ref-list-1
Permissions & Licensing	Information about reproducing this article in parts (figures,tables) or in its entirety can be found online at: http://www.neurology.org/misc/about.xhtml#permissions
Reprints	Information about ordering reprints can be found online: http://www.neurology.org/misc/addir.xhtml#reprintsus

This information is current as of December 26, 2000

Neurology [®] is the official journal of the American Academy of Neurology. Published continuously since 1951, it is now a weekly with 48 issues per year. Copyright . All rights reserved. Print ISSN: 0028-3878. Online ISSN: 1526-632X.

