Brainstem ischemia

To The Editor: In a recent systematic review of 140 CT angiography studies of the V3 segment of the vertebral arteries (VAs), Yamaguchi et al. (Yamaguchi S, Eguchi K, Kiura Y, et al: Posterolateral protrusion of the vertebral artery over the posterior arch of the atlas: quantitative anatomical study using three-dimensional computed tomography angiography. J Neurosurg 9:167–174, August 2008) found anatomical variations and anomalies in this arterial segment. In 23 (8.2%) of 280 VAs examined, the authors observed posterior inferior cerebellar arteries (PICAs) originating from the V3 segment, whereas Stafford13 found absence of PICAs from the V4 segment in 24% of 150 brains examined (on the right side in 15, left side in 6, and both sides in 3). The knowledge of these anatomical variants or anomalies in the V3 and V4 segments of the VAs has enormous clinical and neurological importance.8,12 Recently, we placed omental tissue on the upper cervical cord (at the Cl–2 level) for the treatment of primary occipital neuralgia and on the posterior surface of the medulla oblongata for the treatment of sporadic olivopontocerebellar atrophy (OPCA). In both cases, the patients’ pre- and postoperative status was recorded on videotape.

Case 1. This 51-year-old man had a 21-year history of unilateral or bilateral occipital neuralgia, which occurred in 4 episodes with a duration of 3–10 weeks each. During the first 3 episodes, the pain was of low intensity and was alleviated with nonsteroidal anti-inflammatory drugs (NSAIDs). The fourth episode, however, was more severe and had lasted 6 weeks at the time of the patients’ admission to the hospital. The pain usually began on the left lateral area of the neck, occasionally on the right side. The onset of pain was insidious and the pain progressed rapidly, with radiation to the mastoid, temporal, parietal, and orbital regions as well as the forehead of the affected side. The pain spread and intensified, and within minutes it had become intense and constant, involving both of the orbits, the eyes, the forehead, and temporal regions; it was associated with photophobia, watering of the eyes, and nausea. The pain was reported as initially aching, burning, or pricking, and some minutes later, in the forehead and bitemporal regions as throbbing, explosive, or constrictive. The intense pain lessened with NSAID treatment but did not disappear. The patient had a 30-year history of smoking.

The neurological examination demonstrated no abnormalities. Preoperative CT revealed atherosclerosis in the V4 segment of the VAs, basilar artery, and supraventricular segment of the internal carotid arteries. During surgery (in January 2007) we found: 1) atherosclerosis in both VAs; 2) unilateral hypoplasia of the V4 segment of the right VA; and 3) hyperplasia and tortuosity of the left VA. To date (April 2009) the patient has had no occipital neuralgia since the immediate postoperative period.

Case 2. This 58-year-old right-handed woman was admitted to a local hospital in Mexico City with a 5-year history of early OPCA. Five years before her admission, she presented with progressive difficulty in pronouncing certain words and 2 years later with problems swallowing her saliva. The symptoms had an variable but progressive course (periods of clinical improvement at sea level or following hyperbaric oxygenation therapy alternated with those of worsening symptoms). The patient was a biochemistry teacher who had worked with organic solvents for several years. There was no family history of any similar condition.

The neurological examination revealed moderate dysarthria, a hypotonic uvula, and severe impairment of the patient’s gag reflex due, essentially, to damage to the motor fibers in the ninth and tenth cranial nerves. The rest of the cranial nerves were normal. The patient also had cerebellar ataxia, spasticity and hyperreflexia in the lower limbs, and slight dysmetria and dysdiadochokinesia on both upper limbs. She did not have truncal ataxia or the Romberg or Babinski sign. Preoperative CT and MR imaging revealed: 1) moderate hypotrophy at the anterior lobe of the cerebellum; 2) hypoplasia at the V4 segment of the left VA; and 3) atherosclerosis at the supraclinoid carotid arteries and basilar artery. During surgery (June 2008), we made 4 important observations: 1) moderate atherosclerosis in the V4 segments of both VAs; 2) moderate hypoplasia in the V4 segment of the left VA; 3) origination of the left PICA (diameter < 1 mm) from the V3 segment of the VA; and 4) several exsanguinated and collapsed perforating arteries over the posterior surface of the medulla oblongata. On the first postoperative day, the dysarthria and gag reflex improved 80%, and 3 days later, voluntary movement of the upper extremities was normal. At present, 10 months after the surgery, the patient’s voice and swallowing are normal and her lower-limb motor function is 80% of normal.

Based on our wide experience with omental transplantation to the encephalon8,11 and spinal cord9,10 in patients with ischemic injury, we have observed that there is more neurological improvement during the first weeks after the surgery than in the following months. Thus, we believe that the disappearance of pain in Case 1 was due to a revascularization of ischemic neu-
rons in the caudal portion of the subnucleus caudalis (cell column of 12 mm) in both of the spinal trigeminal nuclei. Because previous studies have shown that ischemia and chemical or electrical stimulation in these subnuclei can provoke painful hypersensitivity and/or spontaneous paroxysms of pain.\textsuperscript{2,3,10} So then, the omentum improvement is due to revascularization of both the nucleus ambiguus and the olivary nuclei because the patient experienced a rapid improvement of her speech and motor coordination as well as reduction of spasticity of the lower limbs. In this respect, lesions in the anterior lobe (paleocerebellum) of the cerebellum are associated with an increase in extensor muscle tone. Therefore, moderate atrophy in the anterior lobe and superior surface of the cerebellum in extensor muscle tone. Therefore, moderate atrophy in the anterior lobe and superior surface of the cerebellum on our patient was caused by degeneration and loss of Olivocerebellar fibers, especially of climbing fibers that originated from the dorsal accessory olivary and principal inferior olivary nuclei (dorsal layer). The history and physical findings in Case 2 are similar to those previously reported by other authors,\textsuperscript{1,14} and, moreover, support the findings of Gilman and colleagues\textsuperscript{5,6} that OPCA is associated with a progressive hypoperfusion and hypometabolism in the brainstem and cerebellum. On the other hand, omental tissue placed on the upper cervical cord and medulla oblongata promotes neoangiogenesis, including omental penetrating vessels, into the underlying medulla oblongata and produces an increase in blood flow, transporting oxygen, neurotransmitters, neurotrophic factors, cytokines, and omental stem cells.\textsuperscript{4,7,9,11}

In summary, based on these anatomical, clinical, and neurosurgical observations in patients with ischemia in the upper cervical cord and medulla oblongata, my colleagues and I believe that several vascular lesions of the craniovertebral junction can be successfully treated, especially in some forms of OPCA. The neurological improvement obtained in the patient with early OPCA after surgery suggests that the primary cause of this disease is of microvascular origin, secondary to anatomical anomalies and/or atherosclerosis in the \( V_1 \) or \( V_2 \) segments of the VAs.

Hernando Rafael, M.D.
Universidad Nacional Autónoma de México
Mexico City, Mexico

References


Response: We would like to thank Dr. Rafael for his interest in our article. As Dr. Rafael has mentioned in his letter, the knowledge of anatomical variants and anomalies of the VA around the craniovertebral junction have significant importance for neurosurgeons and spinal surgeons. The \( V_2 \), \( V_3 \), and \( V_4 \) segments attract many researchers’ concern because of their unique and 3D course. Thus, we tried to quantify this unique anatomical course of the VA, and observed anatomical variations around the craniovertebral junction, based on images obtained from 140 CT angiographic studies. In our study, 8.2% of the PICAs originated from the extracranial portion of the VA. As Dr. Rafael pointed out, the prevalence of extracranial-originating PICAs in our study was lower than in cadaveric and conventional angiographic studies.\textsuperscript{2} Although the cause of this lower prevalence in our article is unclear, the limitations of spatial resolution of CT angiography may have hindered finding these anomalies of small vessels.

In the literature, omental transplantation has been performed for various diseases in the CNS, including moyamoya disease, temporal lobe epilepsy, and chronic spinal injuries.\textsuperscript{1,3,4} However, its clinical utility has not been determined yet. In his letter, Dr. Rafael reports on transplantation of the omentum to the upper cervical cord and medulla oblongata for the treatment of medically intractable occipital neuralgia and OPCA, respectively. It is a great surprise to us to learn that occipital neuralgia and symptoms derived from cerebello medullary dysfunction improved immediately after the operation. In the letter, the primary cause of the disease was speculated to be microvascular origin, secondary to anatomical anomalies and/or atherosclerosis in the \( V_1 \) or \( V_2 \) segments of the