Tic douloureux caused by primitive trigeminal artery or its variant

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Primitve trigeminal artery (PTA) is an extremely rare cause of tic douloureux. None of the reports on PTA variant, which is an anomalous cerebellar artery arising from the internal carotid artery without anastomosis to the basilar artery, has suggested the possibility of this vessel causing tic douloureux. Eight cases of tic douloureux are reported in which a PTA or PTA variant was found during microvascular decompression (MVD). These cases were derived from a series of 1257 patients treated with MVD for tic douloureux. In one patient, the neuralgia was caused by a combination of vessels: a PTA, the superior cerebellar artery, and the anterior inferior cerebellar artery. In the other seven cases, a PTA variant was compressing the root entry zone of the trigeminal nerve. All eight patients gained excellent pain relief after MVD of the root entry zone. The significance of PTA’s and PTA variants as the cause of tic douloureux and the effectiveness of MVD in the management of such cases are discussed.

Key Words • tic douloureux • primitive trigeminal artery • trigeminal neuralgia • microvascular decompression

Primitve trigeminal artery (PTA), which is the most common persistent carotid-basilar artery anastomosis, is seen in 0.1% to 0.2% of angiograms.14 In 1948, Sunderland21 suggested that this artery might be the cause of tic douloureux on the basis of its close relationship to the fifth cranial nerve. Although numerous reports of PTA’s have been published,19,22 there have been only three reported cases in which tic douloureux was associated with this anomaly.10,13,18

Recently, there have been several reports of an atypical cerebellar artery originating from the internal carotid artery (ICA) without anastomosis to the basilar artery.1,2,7,8,16,20,23-25 This anomalous vessel is assumed to be a variant of a PTA,7 but thus far no author has suggested that this vessel may cause trigeminal neuralgia. A series of eight cases of tic douloureux associated with PTA or its variant is presented.

Summary of Cases

Clinical Material

Between 1981 and 1987, our service performed microvascular decompression (MVD) surgery on 1257 patients with medically intractable trigeminal neuralgia. In eight of these patients, exploration of the posterior fossa revealed an abnormal artery emerging from Meckel’s cave or from an isolated dural foramen at the dorsum sellae in close proximity to the trigeminal nerve. All of these patients were females and their average age at presentation was 66 years (range 53 to 77 years). All eight cases presented with classical trigeminal neuralgia. Symptoms involved the left side in five patients and the right side in three. The pain was distributed over the second and third divisions of the trigeminal nerve in four cases, over the second division in three cases, and over the first and second divisions in one case (Table 1). Two patients had undergone percutaneous gasserian ganglion block at other institutions and suffered slight facial hypesthesia. No other neurological deficits were detected, and computerized tomography (CT) scans demonstrated no abnormality in any patient. Preoperative angiography was not performed in any of the eight cases and the anomalous vessels were found unexpectedly during the MVD procedure.

Operative Technique

Surgery was performed with the patient in a lateral position. The trigeminal nerve root and offending vessels were exposed through a small retromastoid opening. All of the vessels in contact with the nerve were mobilized and held off the root entry zone of the fifth cranial nerve by means of shredded Teflon pledgets. Extreme care was taken to avoid kinking the transposed vessels or stretching the perforators. When an artery
TABLE 1
Clinical summary in eight cases of tic douloureux caused by a PTA or its variant*

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Age (yrs)</th>
<th>Sex</th>
<th>Side</th>
<th>Distribution of Pain†</th>
<th>Duration of Symptoms (yrs)</th>
<th>Gasserian Ganglion Block</th>
<th>Offending Vessels</th>
<th>Interposition Between Roots‡</th>
<th>Follow-Up Period (mos)</th>
<th>Operative Complications</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>75, F</td>
<td>rt</td>
<td>V2, 3</td>
<td>no</td>
<td>ab. AICA from Mc</td>
<td>yes</td>
<td>42</td>
<td>facial hyposthesia</td>
<td>42</td>
<td>0</td>
</tr>
<tr>
<td>2</td>
<td>77, F</td>
<td>lt</td>
<td>V2</td>
<td>20</td>
<td>yes ab. SCA from df</td>
<td>yes</td>
<td>37</td>
<td></td>
<td>37</td>
<td>0</td>
</tr>
<tr>
<td>3</td>
<td>53, F</td>
<td>lt</td>
<td>V2</td>
<td>4</td>
<td>no ab. PICA from Mc (ICA origin), AICA, &amp; SCA</td>
<td>yes</td>
<td>31</td>
<td></td>
<td>31</td>
<td>0</td>
</tr>
<tr>
<td>4</td>
<td>54, F</td>
<td>rt</td>
<td>V2, 3</td>
<td>5</td>
<td>yes ab. AICA from Mc &amp; SCA</td>
<td>yes</td>
<td>30</td>
<td></td>
<td>30</td>
<td>0</td>
</tr>
<tr>
<td>5</td>
<td>74, F</td>
<td>rt</td>
<td>V2, 3</td>
<td>6</td>
<td>no PTA from Mc, AICA, &amp; SCA</td>
<td>no</td>
<td>29</td>
<td></td>
<td>29</td>
<td>0</td>
</tr>
<tr>
<td>6</td>
<td>56, F</td>
<td>lt</td>
<td>V2</td>
<td>6</td>
<td>no ab. AICA from Mc (ICA origin)</td>
<td>no</td>
<td>21</td>
<td></td>
<td>21</td>
<td>0</td>
</tr>
<tr>
<td>7</td>
<td>69, F</td>
<td>lt</td>
<td>V1, 2</td>
<td>1</td>
<td>no ab. AICA from df (ICA origin)</td>
<td>no</td>
<td>19</td>
<td></td>
<td>19</td>
<td>0</td>
</tr>
<tr>
<td>8</td>
<td>67, F</td>
<td>lt</td>
<td>V2, 3</td>
<td>3</td>
<td>no ab. AICA from Mc (ICA origin)</td>
<td>no</td>
<td>12</td>
<td></td>
<td>12</td>
<td>0</td>
</tr>
</tbody>
</table>

* PTA = primitive trigeminal artery; ab. = aberrant; AICA = anterior inferior cerebellar artery; Mc = Meckel’s cave; SCA = superior cerebellar artery; df = dural foramen at the dorsum sellae; PICA = posterior inferior cerebellar artery; ICA = internal carotid artery.
† V1, 2, 3: first, second, and third divisions of the trigeminal nerve.
‡ The sensory and motor roots of the trigeminal nerve.

was interposed between the sensory and motor roots of the trigeminal nerve, the roots were separated from each other and the artery was moved toward the peripheral part of the motor root. Because arterial contact with the trigeminal nerve within Meckel’s cave is not considered to be a cause of tic douloureux, the vessel in this location was simply separated from the nerve using pieces of Surgicel.

**Operative Findings**

**Primitive Trigeminal Artery.** Surgical exploration of Case 5 revealed an anomalous artery emerging into the posterior fossa from Meckel’s cave, compressing the distal portion of the trigeminal root and joining the basilar artery. At the nerve entry zone, the anterior inferior cerebellar artery (AICA) and the superior cerebellar artery (SCA) arising from the basilar artery were pinching the trigeminal nerve. Postoperative angiography revealed that the abnormal artery was a Salzmann type 3 PTA (Fig. 1A).

**Primitive Trigeminal Artery Variant.** In five patients (Cases 1, 4, 6, 7, and 8), aberrant arteries emerged from Meckel’s cave or from an isolated dural foramen at the dorsum sellae, ran the intracranial course of an AICA, and directly compressed the root entry zone of the fifth cranial nerve (Figs. 1B, 2, and 3). In two cases, these arteries passed between the sensory and motor roots of the trigeminal nerve (Fig. 2). One patient (Case 2) was found to have an abnormal SCA which entered the posterior fossa from a dural foramen at the dorsum sellae and intruded between the trigeminal nerve and the pons. Another patient (Case 3) showed complicated nerve compression by the SCA, the AICA, and an abnormal artery emerging from Meckel’s cave. This abnormal artery was interposed between the sensory and motor roots of the trigeminal nerve and coursed caudally. Postoperative angiography demonstrated an aberrant posterior inferior cerebellar artery (PICA) arising from the ICA (Fig. 1C). The intraoperative findings in these cases are illustrated in Fig. 4.
Tic douloureux and primitive trigeminal artery

Operative Results

During the average follow-up period of 28 months (range 12 to 42 months) after MVD, all eight patients had excellent pain relief without recurrence. The facial hypesthesias which had been observed preoperatively in two patients remained unchanged. Another patient, who had an anomalous artery situated between the sensory and motor nerve roots, experienced moderate facial hypesthesia postoperatively. No other complications were observed. Postoperative angiograms were obtained in five cases, three of which demonstrated an AICA arising from the ICA. In one patient the PICA and in another a PTA originated from the ICA. Angiography was not performed in three patients; in those cases the anomalous arteries were identified solely by the operative findings.

Discussion

According to the medical literature, the clinical significance of a PTA lies in its occasional association with aneurysms and its potential for causing tic douloureux. Although the possibility of nerve compression was suggested by Sunderland in 1948 and has been mentioned by other authors, only three cases of tic douloureux caused by PTA have been reported. Kempe and Smith described the first case, a left-sided tic douloureux with compression by a PTA. This patient had an area of hypesthesia corresponding to the second division of the left trigeminal nerve. Exploration of the posterior fossa revealed that the PTA was elevating the fifth nerve and was in intimate contact with the root. The nerve root of the affected division was dissected.
Jackson and Garza-Mercado\textsuperscript{10} described a woman who experienced episodic left tic douloureux. Percutaneous carotid angiography showed a left PTA, but surgery was not performed. The third case was reported by Piatt and Wilkins;\textsuperscript{18} their patient was included in a series of 104 patients undergoing MVD for tic douloureux and the offending vessel was a PTA. That case was not described in detail. Madonick and Ruskin\textsuperscript{15} reported a patient with recurrent oculomotor paresis and a PTA who also had episodes of facial pain resembling those of tic douloureux, although this symptom was not singled out for comment.

Among 1257 patients with tic douloureux who underwent MVD on our service, only one had a PTA. Posterior fossa exploration in that case revealed that the trigeminal neuralgia was caused by a combination of the PTA, SCA, and AICA. Complete pain relief was achieved by decompression of all of the offending vessels. The fact that PTA's rarely cause tic douloureux is probably explained by their intracranial course; since these vessels run medially and join the basilar artery after entering the posterior fossa, they are less likely to compress the root entry zone and thus do not often cause tic douloureux.\textsuperscript{5,11,12} Those that do cause tic douloureux may be extremely long and tortuous.

On the other hand, in our series of 1257 patients, we encountered seven in whom the nerve root entry zone was directly compressed by an anomalous cerebellar artery that did not arise from the basilar artery but originated from the ICA. These anomalous cerebellar arteries were first reported by Teal, \textit{et al.},\textsuperscript{23,24} in 1972. To our knowledge, there have been 46 cases reported since then; 13 patients had an aberrant SCA arising from the ICA, 28 had an aberrant AICA, and five had an aberrant PICA.\textsuperscript{1,2,7-9,16,23-25} Haughton, \textit{et al.},\textsuperscript{7} postulated that this anomalous artery is a variant of a PTA resulting from incomplete fusion of the longitudinal neural arteries. Therefore, these vessels have been designated "primitive trigeminal artery variants." These anomalous vessels do not join the basilar artery as a PTA does. After they enter the posterior fossa from Meckel's cave or from an isolated dural foramen,\textsuperscript{7} they course dorsally and run near the root entry zone of the trigeminal nerve.

Among the 46 reported cases of PTA variant, one patient with an aberrant AICA was described as having episodes of trigeminal neuralgia, but the authors did not comment on its etiology.\textsuperscript{8} Although some authors have pointed out the clinical significance of PTA variants in cases with carotid artery ligation,\textsuperscript{9,20,23,24} none has mentioned their possible role as the cause of tic douloureux. Ito, \textit{et al.},\textsuperscript{9} described autopsy findings of an anomalous AICA arising from the cavernous ICA and running dorsally between the fifth and eighth cranial nerves, in close proximity to the root entry zone of

\textbf{FIG. 4.} Artist's summary of the intraoperative findings of the trigeminal nerve-vascular complex in eight cases. TN = trigeminal nerve; df = dural foramen; Mc = Meckel's cave; ab = aberrant artery; s = superior cerebellar artery; a = anterior inferior cerebellar artery.
Tic douloureux and primitive trigeminal artery

the fifth cranial nerve. We found seven among our series of 1257 patients in whom a PTA variant was responsible for the nerve compression. Therefore, PTA variants must be seriously considered as a possible cause of tic douloureux.

The clinical features of trigeminal neuralgia associated with compression by a PTA or PTA variant were indistinguishable from those caused by the usual SCA or AICA compression. The average patients' age at onset did not differ from that of typical cases. Whereas the patients reported by Kempe and Smith and Jackson and Garza-Mercado had facial hypesthesias on the affected side, our cases presented with classical tic douloureux without neurological deficits (except for two patients who had previously received percutaneous gasserian ganglion block). The distribution of pain most commonly corresponded to the second and third divisions; however, no special correlation was noted between the type of aberrant artery and the pain distribution. The CT scans were normal in all cases. The only diagnostic procedure that demonstrated the anomaly was carotid angiography.

The results of MVD surgery were excellent in all except one patient who suffered persistent facial hypesthesia postoperatively. When the aberrant artery interposes between the sensory and motor roots of the trigeminal nerve, excessive manipulation of the nerve should be avoided to prevent possible postoperative facial hypesthesia. Mobilization and transposition of a PTA or PTA variant should be performed without kinking the vessel or stretching the perforators, because any circulatory disturbance of these arteries may result in serious complications.

Percutaneous gasserian ablative procedures (gasserian ganglion alcohol block, radiofrequency gangliolysis, or retrogasserian glycerol injection) may result in severe hemorrhagic complications when anomalous arteries present within Meckel's cave. Sunderland stated that these arteries run very close to the gasserian ganglion and the trigeminal nerve root. Despite their low incidence (0.1% and 0.6% in our series), a PTA or PTA variant should be considered as a possible etiology for tic douloureux.