Glossopharyngeal neuralgia

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Various factors have been considered in the etiology and pathogenesis of glossopharyngeal neuralgia. Vascular compression of the involved cranial nerves has been demonstrated in sporadic cases. In this series of six patients, it was noted with the aid of the operating microscope that the ninth and tenth cranial nerves were compressed by a tortuous vertebral artery or posterior inferior cerebellar artery at the nerve root entry zone in five cases. In selected patients, microvascular decompression without section of the nerves may result in a cure.

KEY WORDS - glossopharyngeal neuralgia - vascular compression - microvascular decompression

Glossopharyngeal neuralgia is uncommon in comparison to trigeminal neuralgia. The reported relative frequency of glossopharyngeal neuralgia to trigeminal neuralgia ranges from 1% to 0.75%. Currently, the usual method of treatment of glossopharyngeal neuralgia is surgical, although some have reported satisfactory pain relief with medical treatment. The surgical techniques vary from an extracranial section of the ninth cranial nerve to an intracranial section of the same nerve plus the upper rootlets of the vagus. Others have advocated selective bulbar tractotomy as the procedure of choice, particularly if combined neuralgias are evident.

In the present communication we analyze our experience with glossopharyngeal neuralgia in six patients and describe yet another mode of treatment, namely, microvascular decompression of the ninth and tenth cranial nerves where vascular compression appears to be the causative factor for irritation of the nerves.

Summary of Cases

Clinical Material

The present series consists of six patients with glossopharyngeal neuralgia treated at the Presbyterian-University Hospital from 1971 to 1975. There were three men and three women, ranging in age from 36 to 62 years. The duration of symptoms varied from 6 months to 7 years, and the left side was affected more commonly (five of six patients). One of these patients (Case 6), the youngest in our series, had combined atypical trigeminal neuralgia and glossopharyngeal neuralgia. Another patient (Case 1) had intracranial section of the ninth nerve alone in 1968 followed by a pain-free period of about 2 years; the pain had recurred predominantly in the ear.

The onset of pain was sudden, transitory, and paroxysmal (except in Case 6). The pain was described variably as excruciating, electric shock-like, lancinating, jabbing, cutting, and burning. In Case 6 the pain was constant,
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aching, and burning in nature involving the left orbit, face, oropharynx, and ear, with radiation of pain toward the neck and shoulder; ptosis was noted occasionally during the attacks of severe pain. The onset of pain in this patient followed trauma to the face. None of our patients had syncopal episodes, asystole, or seizures. Five patients had a trial of carbamazepine (Tegretol) and/or phenytoin sodium (Dilantin) for a variable length of time. Only one patient obtained significant relief from the pain for 1½ years with carbamazepine.

In all cases, the skull x-ray films were normal and the styloid processes were not elongated. Five patients underwent vertebral and carotid angiography or vertebral angiography alone. In two patients tortuous ectatic arteries (vertebral and posterior inferior cerebellar) were seen on the affected side. A summary of the clinical material is presented in Table 1.

Operative Treatment

All patients were operated on in the sitting position by a retromastoid craniectomy approach. In Case 1 the upper rootlets of the vagus were sectioned. The patient had recurrent glossopharyngeal neuralgia after previous section of the ninth cranial nerve. Microvascular decompression was performed in four patients. In one of these the ninth and upper rootlets of the tenth cranial nerve were also sectioned. In all of these patients either the vertebral artery or the posterior inferior cerebellar artery (PICA) was seen to be cross-compressing the ninth and tenth cranial nerves at the nerve root entry zone. With the aid of the operating microscope under ×16 to ×25 magnification, the vessels were mobilized away from the nerve and a polyvinyl chloride sponge* was interposed between the nerve and the vessel. In the remaining patient a tortuous vertebral artery was seen cross-compressing the rootlets of the ninth and tenth cranial nerves, and the vessel was not disturbed for fear of causing infarction of the brain stem. In Case 6, in addition to vascular decompression of the ninth and tenth, the fifth cranial nerve was also decompressed from a loop of the superior cerebellar artery and the anterior limb of the petrosal vein for treatment of coexisting atypical trigeminal neuralgia.

Operative Results

Three patients had division of rootlets alone (two), or in conjunction with vascular decompression (one). They have been followed for 2 to 4½ years with good results. The other three patients had microvascular decompression alone. Of these, one remains pain-free 6 months after surgery and one, followed for 1 year, continues to have some pain. The remaining patient treated by microvascular decompression alone died in the postoperative period. This patient did well in the immediate postoperative period but developed severe hypertension in the recovery room and soon lapsed into coma. Re-exploration of the operative site revealed a contused cerebellum and a small intracerebellar clot. Despite removal of the clot and partial resection of the right cerebellar lobe the patient died. Autopsy revealed a deep intracerebral hemorrhage with intraventricular extension on the right side. There was uncal herniation on the right with secondary brain-stem hemorrhages.

Discussion

The diagnosis of glossopharyngeal neuralgia is made by the history of severe paroxysmal pain of sudden onset in the oropharynx, tonsillar fossa, base of the tongue, ear, or in both locations, often precipitated by such activities as swallowing, chewing, or coughing. Occasionally, the neuralgic attack may be associated with syncope, cardiac arrest, or seizures. Trauma, local infection, elongated styloid process, ossified stylohyoid ligament, vascular abnormalities, and intra- or extracranial tumors have all been considered as etiological factors in glossopharyngeal neuralgia.

*Ivalon sponge manufactured by Unipoint Laboratories, High Point, North Carolina.
<table>
<thead>
<tr>
<th>Case No.</th>
<th>Age (yrs)</th>
<th>Side</th>
<th>Duration (yrs)</th>
<th>Chief Site of Pain</th>
<th>Radiation</th>
<th>Associated Symptoms</th>
<th>Examination†</th>
<th>Angiography</th>
<th>Surgery</th>
<th>Findings</th>
<th>Complications</th>
<th>Follow-up Period</th>
<th>Results</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>62, M</td>
<td>lt</td>
<td>5</td>
<td>ear &amp; pharynx</td>
<td>front of ear &amp; face</td>
<td>—</td>
<td>—</td>
<td>—</td>
<td>division of upper rootlets of X</td>
<td>root entry zone not visualized due to local scarring</td>
<td>—</td>
<td>g; mild palatal weakness</td>
<td>4½ yrs</td>
</tr>
<tr>
<td>2</td>
<td>55, F</td>
<td>rt</td>
<td>4</td>
<td>pharynx &amp; tonsil</td>
<td>deep in ear</td>
<td>—</td>
<td>hypalgesia 1 cm diameter anterior to tragus</td>
<td>normal</td>
<td>microvascular decompression of IX &amp; X</td>
<td>vascular compression by PICA at root entry zone</td>
<td>—</td>
<td>hypertension postop; intracranial hem with uncal herniation &amp; 2nd brain-stem hem</td>
<td>—</td>
</tr>
<tr>
<td>3</td>
<td>59, M</td>
<td>lt</td>
<td>½</td>
<td>deep in ear</td>
<td>pharynx &amp; left side of face</td>
<td>hypalgesia V2, decreased hearing in left ear</td>
<td>normal</td>
<td>tortuous vertebral artery &amp; PICA</td>
<td>microvascular decompression &amp; section of IX &amp; upper rootlets of X</td>
<td>ecstatic vertebral artery compressing root entry zone; demyelination of nerves on histology</td>
<td>transient hypertension for few days</td>
<td>2½ yrs</td>
<td>good</td>
</tr>
<tr>
<td>4</td>
<td>57, F</td>
<td>lt</td>
<td>2</td>
<td>oropharynx</td>
<td>external ear</td>
<td>—</td>
<td>weak gag reflex (lt)</td>
<td>tortuous vertebral artery &amp; PICA</td>
<td>section of IX &amp; upper rootlets of X</td>
<td>nerve compression at root entry zone by vertebral artery, vessel not mobilized</td>
<td>transient dysphagia; gag</td>
<td>2 yrs</td>
<td>good</td>
</tr>
<tr>
<td>5</td>
<td>42, M</td>
<td>lt</td>
<td>7</td>
<td>tonsillar pillars</td>
<td>ear &amp; angle of jaw</td>
<td>hypertension</td>
<td>normal</td>
<td>normal</td>
<td>microvascular decompression of IX &amp; X</td>
<td>PICA compressing the nerves at root entry zone</td>
<td>—</td>
<td>1 yr</td>
<td>still has some pain</td>
</tr>
<tr>
<td>6</td>
<td>36, F</td>
<td>lt</td>
<td>2½</td>
<td>oropharynx, ear, face</td>
<td>neck &amp; shoulder</td>
<td>ptosis during episodes of pain</td>
<td>normal</td>
<td>microvascular decompression of IX, X &amp; V</td>
<td>loop of PICA compressing the IX &amp; X at root entry zone; SCA compressing V</td>
<td>—</td>
<td>6 mos</td>
<td>good</td>
<td></td>
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*PICA = posterior inferior cerebellar artery; SCA = superior cerebellar artery; hem = hemorrhage. None of these patients had a specific trigger point.†CSF normal in all patients but Case 1, who was not tested.
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spasm is well known. Jannetta, et al. have presented evidence in this regard in trigeminal neuralgia and hemifacial spasm. In 1889, Pope reported loss of taste at the back of the tongue in a patient found to have a thrombosed vertebral artery pressing on the glossopharyngeal nerve at autopsy. Lillie and Craig described a patient with severe neuralgic pain in the ear associated with increasing deafness. At operation, either the anterior inferior cerebellar artery or one of its branches was seen to be looping over the eighth and ninth cranial nerve. The loop of the vessel was mobilized from the nerves after sectioning the ninth cranial nerve. The patient was cured. Brihaye, et al. described the postmortem findings of a 77-year-old woman with glossopharyngeal neuralgia. An enlarged atheromatous vertebral artery was seen compressing the lower cranial nerves. The compressed nerve roots showed partial demyelination. Kempe and Smith found a large persistent hypoglossal artery at angiography and operation that was angulating the lower cranial nerve in a 42-year-old woman with glossopharyngeal neuralgia.

In five of our six cases either the vertebral artery or PICA was seen to cross-compress the rootlets of the ninth and tenth cranial nerves. This cross-compression was strikingly seen at the root entry zone of the nerves. The root entry zone was not visualized in the sixth patient (Case 1) who had local scarring from a prior ninth nerve section. In the patient with associated atypical trigeminal neuralgia, the fifth cranial nerve was also compressed by the superior cerebellar artery and the anterior limb of the petrosal vein. It may be noted that the preoperative angiogram showed elongated, tortuous vertebral and PICA arteries in only two of these patients.

Although there are cases where section of the ninth nerve alone has relieved the pain, the subsequent recurrence of pain in some cases and the common association of vagal neuralgia have led several authors to include the upper rootlets of the vagus routinely. This had to be done in one of our patients with recurrent neuralgia who had previous section of the ninth nerve. It has also been suggested that the upper rootlets of the vagus be included in only selected cases. Occasional side effects have been observed following section of the rootlets of the vagus. Also, transient elevation of blood pressure following section of the ninth nerve has been reported. This was postulated as the cause of the intracerebral hematoma in the postoperative period in Case 2. The postoperative sensory or motor deficit following section of the nerves has been variable. The operating microscope has allowed us to recognize vascular compressions of the nerves that otherwise might be missed when seen with the naked eye. The mobilization of the vessels in contact with the nerves will often be sufficient for symptomatic relief. However, if this cannot be done with safety, the standard sectioning of the nerves may be carried out as was done in one of our cases. Nerve section may be necessary when the vessel cannot be satisfactorily kept away from the nerves without causing medullary compression. Although we have used a small piece of polyvinyl chloride sponge (Ivalon) or silicone sponge for such a purpose, we feel that the application of a minute amount of tissue adhesive into a smaller prosthesis may be more desirable. The small number of cases in this series precludes any definite conclusion, but considering our satisfactory experience with the microvascular decompression of the trigeminal nerve, we are inclined to believe that a favorable outcome can be expected.

References

9. Ekbom KA, Westerberg CE: Carbamazepine
in glossopharyngeal neuralgia. Arch Neurol 14:595-596, 1966
27. Weisenburg TH: Cerebello-pontile tumor diagnosed for six years as tic douloureux. The symptoms of irritation of ninth and tenth cranial nerves. JAMA 54:1600-1604, 1910

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