THE ETIOPATHOGENESIS OF IDIOPATHIC TRIGEMINAL NEURALGIA*

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The pathogenesis of essential trigeminal neuralgia still remains obscure. One can easily conceive that an irritative or compressive lesion on a peripheral nerve can cause many painful phenomena, however, it is difficult to explain facial neuralgia in the absence of obvious lesions of the roots or branches of the trigeminus, or moreover, to explain the manifestations so characteristic of Trousseau's malady: the triggering of an attack by excitation of the «dolorogenic zone», ferocious appearance, rapid development of the attack, and a refractory period at the end.

We attempt to answer in this paper certain pertinent questions.

1. Where is the causal lesion situated?

There is no point of nerve trajectory or possible site of causative lesion which has not been considered: terminal branches, Gasserian ganglion, root, or nucleus.

The hypothesis of an irritative nuclear or paranuclear lesion as the cause of trigeminal neuralgia does not hold because of the absence of a sensory deficiency or a partially neuralgic condition, nor does a protuberant lesion ever result in a typical neuralgia.

The theory of neurodocitis held by J. A. Sicard and supported by many authorities presupposes an involvement of the trunk or peripheral branches. Pain results from a congestive condition caused by constriction of the nerve branches in bony canals already narrowed by osteophytes, a thickening of the periosteum, or by a fibrous change of connective tissue filling up the orifices (supra-orbital, infra-orbital, or mandibular).

The hypothesis of an irritation of the sensitive root of the nerve was first suggested by Dandy. «Of those causes of neuralgia of the trigeminus,» he said, «it is necessary to consider the sensory root.» We also prefer this latest theory of the origin of essential trigeminal

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neuralgia for it is supported by strong evidence, which we will discuss, and which relates directly to the mechanism of the trigeminal crisis and its causes.

2. What is the mechanism of the trigeminal crisis?

It is interesting to note the similarities existing between crises of facial neuralgia and facial spasm. Facial neuralgia involves a mixed nerve, mainly sensory; facial spasm involves a mixed nerve which is essentially motor. Both are followed by spontaneous paroxysms similar to the effect of electrical stimulation. Both begin in a nerve branch and have a tendency to spread to all branches of the fifth nerve. Both, moreover, can exist in the same person.[1]

Kugelberg and Lindblom, in 1955, studied in detail the refractory period following a trigeminal crisis in patients suffering from trigeminal neuralgia.[2] They provoked the crisis with intense surface stimulations of varying amplitudes. The refractory period lasted from 20 seconds to three minutes, depending on the length or intensity of the preceding crisis; the painful trigger is perhaps explained by a simple interaction or short circuit of the sensory fibers and the pain fibers. Like other authors, they place the responsible lesions in the brain stem.

W. J. Gardner, in 1962, suggested that paroxysmal crises, physiopathologically unstable and reversible states, resulted from a transaxonal short circuit caused by the rapid chemical action in the nerve fibers.[3] A light and chronic compression causes atrophy of the myelin sheath and permits the current of one fiber to pass through a neighboring fiber. Trigeminal neuralgia is the result of a reflex of a tactile stimulus of the efferent parasymapathetic arch which is excited by the short circuit, at the point of compression. The dimensions of the pain fibers are similar to those of the parasympathetic fibers (Figure 1).

The other mixed cranial nerves (facial, glossopharyngeal and vagus) could be the site of the same phenomenon because the afferent fibers are both present at the point of contact. Gardner's theory presupposes intervening parasympathetic efferent fibers, which are present in the facial, glossopharyngeal and vagus nerves, but, as far as is now known, not present in the trigeminal nerve. Thus Gardner's theory is poorly adapted as an explanation of trigeminal neuralgia, although it is remarkably suitable as an explanation of neuralgia of the glossopharyngeal nerve and all excitation of the saliva-

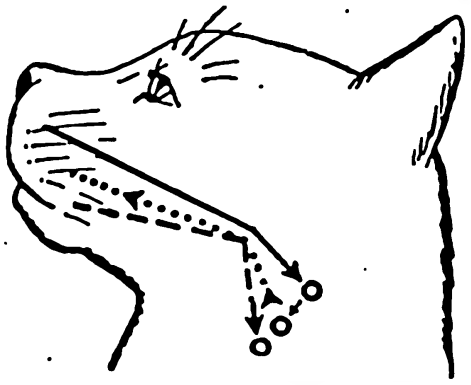

Figure 1. The tactile influx (solid line) passes through the main sensory nucleus where it triggers a parasympathetic discharge (dotted line). The blockage of this efferent route at the point of compression excites the pain fibers (dashes). (After W. J. Gardner)

ry reflexes (gustatory, olfactory and psychic) which, returning via the secretory fibers, are able in effect to trigger the pain.

It is also possible to question whether or not a simple short circuit between sensory fibers and pain fibers would actually cause pain, and whether an ordinary tactile stimulus can be re-routed from its normal course and cause a sensation of pain (Figure 2).

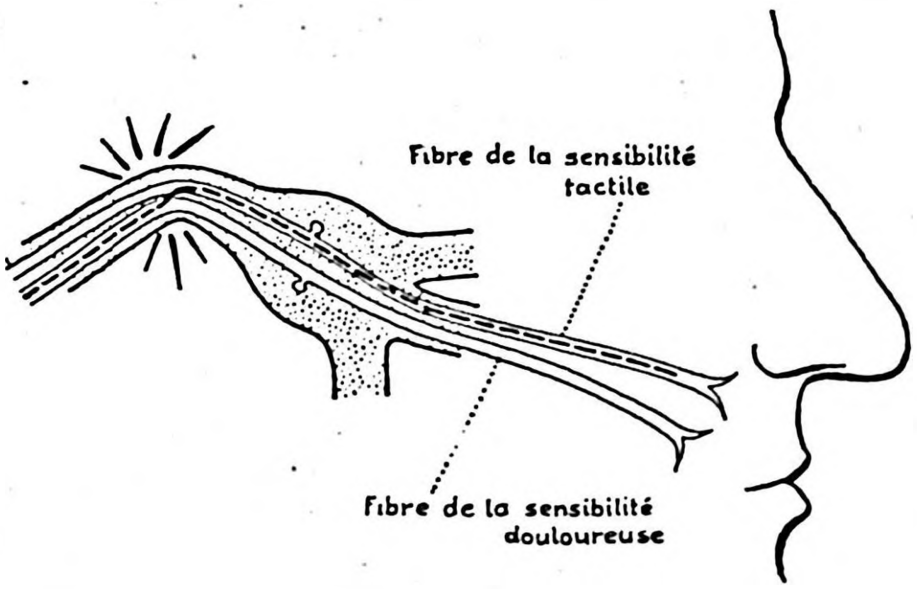

Figure 2. Theory of the transaxonal short circuit.

In support of the hypothesis of the transaxonal short circuit comes evidence of a therapeutic type: first, the trigeminal crises (the same as clonic facial attacks) can be immediately cured by the simple manipulation of the roots of the nerves (Gardner and Sava). The operation of Taarnhoj, a simple opening of the dura in the

middle of the root at the point where it crosses the os petrosa, is followed by a lasting cure in a certain percentage of cases. Second, the relief obtained by an infiltration or sectioning of the peripheral branch, inexplicable in the case of an irritative lesion situated at the root beyond the infiltration, is understandable on the assumption that the interruption of the sensory fibers prevents their short circuit with pain fibers.

It still remains to be explained why trigeminal neuralgia is most often situated, at least initially, in the area of the middle branch; that is, in the superior maxillary nerve. Sicard, in his studies published in 1931 and 1932,[10] had studied the place of the radicles in the trigeminal root, and had shown that, despite possible variations, fibers destined to the superior maxillary nerve are in the center, while those of the two other terminal branches, opthalmic and mandibular, are on the outside, or twist around. It is possible that the fibers in the middle of the nerve are more susceptible to compression than the others, and are thus more frequently the site of a short circuit capable of causing a trigeminal crisis.

3. What are the causes of trigeminal neuralgia?

The compression which triggers the short circuit should occur on the sensory root, either in the posterior fossa or the upper edge of the tympanic nerve.

According to Dandy, 60 percent of idiopathic facial neuralgias are secondary to a lesion situated in the posterior fossa, a concept which supported his preference for operating by the occipital route rather than the temporal route. The majority of neurosurgeons have not accepted his concept however.

The neoplasms capable of triggering trigeminal neuralgia are those which evolve very slowly, and lead to a chronic compression of the nerve and a persistent tactile sensitivity. These neoplasms are, in particular, cholesteatomas of the ponto-cerebellar angle, vascular malformations of the basal trunk or the cerebellar arteries, etc.

The neoplasms which evolve more rapidly, the neurinomas for example, seem on the contrary to change tactile sensitivity earlier, and bring painful crises less frequently.

In a great number of cases, even the most minute investigations did not reveal neoplasms in the posterior fossa. It is difficult to discover the causes of non-tumoral compression, but particularly

suspect are those which would become more evident with age, in as much as idiopathic trigeminal neuralgia occurs chiefly in aged patients. We have classified the causes which seem to us capable of provoking a trigeminal crisis into three groups.

1. Osseous causes : The root of the nerve rests directly on the top of the os petrosa or the petrous apex, much like the strings of an instrument across a bridge (Figure 3). Any super-elevation of the summit of the petrous bone exaggerates the angle of the nerve root.

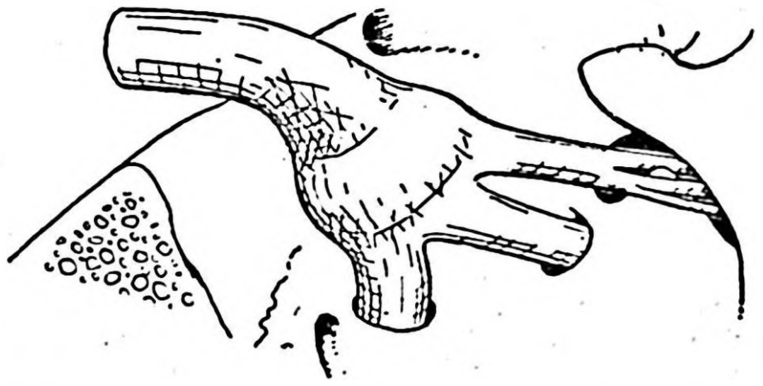

Figure 3. Trigeminal root and top of the petrous bridge.

Classically, the os petrosa constitutes a «buttress» leaning on the body of the sphenoid. In fact, however, even with its summit, the bone is less firmly attached to the base of the cranium than has been thought. In front, the petro-sphenoidal suture is fixed poorly to the apex; behind, the petro-occipital suture is more solid, but is creased by the sinusal groove and the fixation is poor. A lateral compression of the skull displaces the point of the os petrosa by one to three mm. The same compression of the cranium causes the folding of the dura mater in the latero-basilar area (Figure 4 and 5).[9] It is easily conceivable that any modification of the base of the skull, basal invagination in particular, could displace the bridge of the os petrosa.

In a certain number of patients with Paget's cranium, trigeminal neuralgia was also observed. In five cases of Paget's disorder complicated by basilar invagination, we noted especially three with facial neuralgia. The super-elevation of the petrous apex was visible by radiography.[8] In numerous observations of Paget's cranium, one finds marked diminution of the corneal reflexes, facial paresthesis, and sometimes, facial neuralgia.

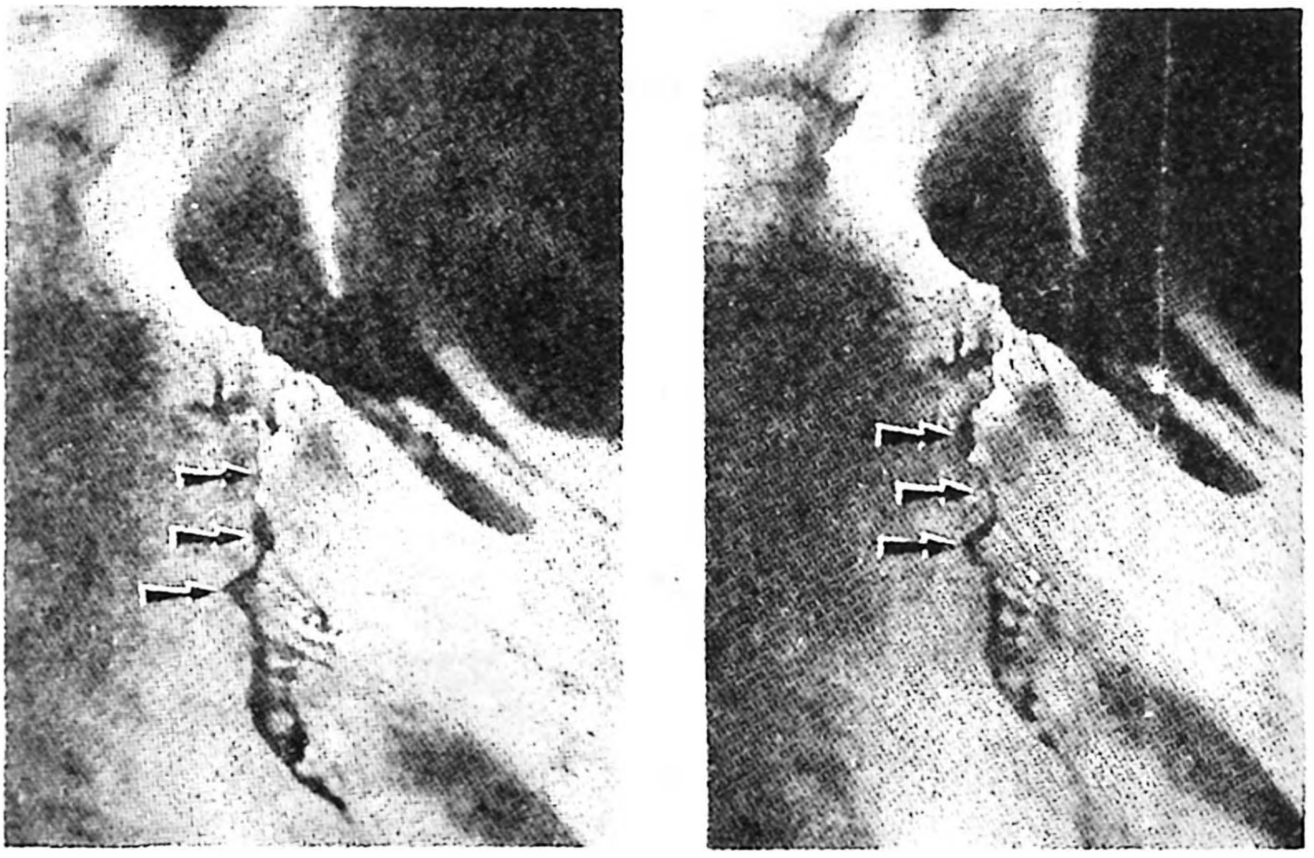

Figure 4. Skull. At left, without transverse compression; at right, compressed.

It is possible that senile osteoporosis of the petrous bridge is the result of traction of the muscles of the neck on the projection of the sterno-cleidal mastoideum. The motion of these neck muscles (trapezium and splenius) could make the petrous pyramid «see-saw», so much so that its base would decrease and its summit rise. The stronger traction to the right (in right-handed people) explains perhaps why the localization is more often on the right than on the left in facial neuralgia. Gardner et al, after a study of cranial radiography made on 130 patients with facial neuralgia and 200 normal subjects, stated that in a large percentage of the patients with facial neuralgia, the right petrous summit was higher than the left.

In 1963, Muller and Levy compared the function of the odontoid epiphysis with reference to the bimastoid line and line of MacGregor in 56 patients with neuralgic attacks of the fifth cranial nerve with 112 controls, and found that the odontoid epiphysis is 5.3 to 0.9 mm below the bimastoid line in female patients with neuralgia of the fifth cranial nerve, and between 3.0 to 0.6 mm in the controls. No important difference was noted among the males. Neuralgia of the fifth cranial nerve is also rare in males.

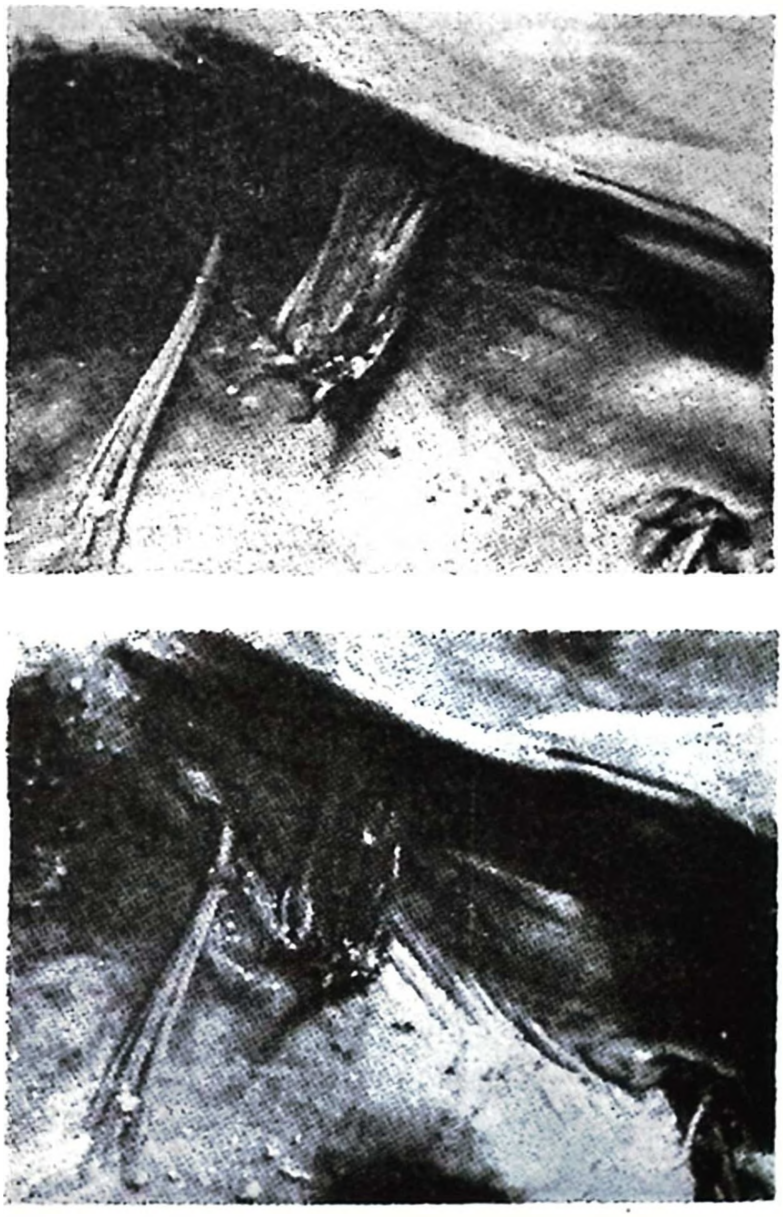

Figure 5. Cranium. Above, without compression; below, the oblique folds of the dura mater proceed from the posterior jagged hole toward the subdural entry of the sixth nerve. The fibrous material develops inside the nerve.

2. Meningeal causes : In a previous article we reported a study of 58 skulls and 16 dissections and noted the osteo-dural orifices in which the root of the fifth cranial nerve passes over the point of the petrous.

In the majority of cases, the trigeminus is large in the orifice at the entrance to Meckel's cavity. One out of three of our patients had an interior calcification, as seen in the dura mater, just below the root of the fifth nerve, one or two mm below the superior petrous sinus. It developed either from a lenticular formation situated on the roof of the orifice of the cavity, or from a bony lamina bridging the posterior clinoid and the petrous in the petrous clinoidal

ligament (Figure 6). The calcification of this ligament is visible in radiography.

The surface of the bone may also be studded with bony spines, or there may be a small osteophytic point on the exterior side of the trigeminal incision on the higher edge of the os petrosa (Figure 7). In one instance, we found a bony spiculum on the edge of the anterior

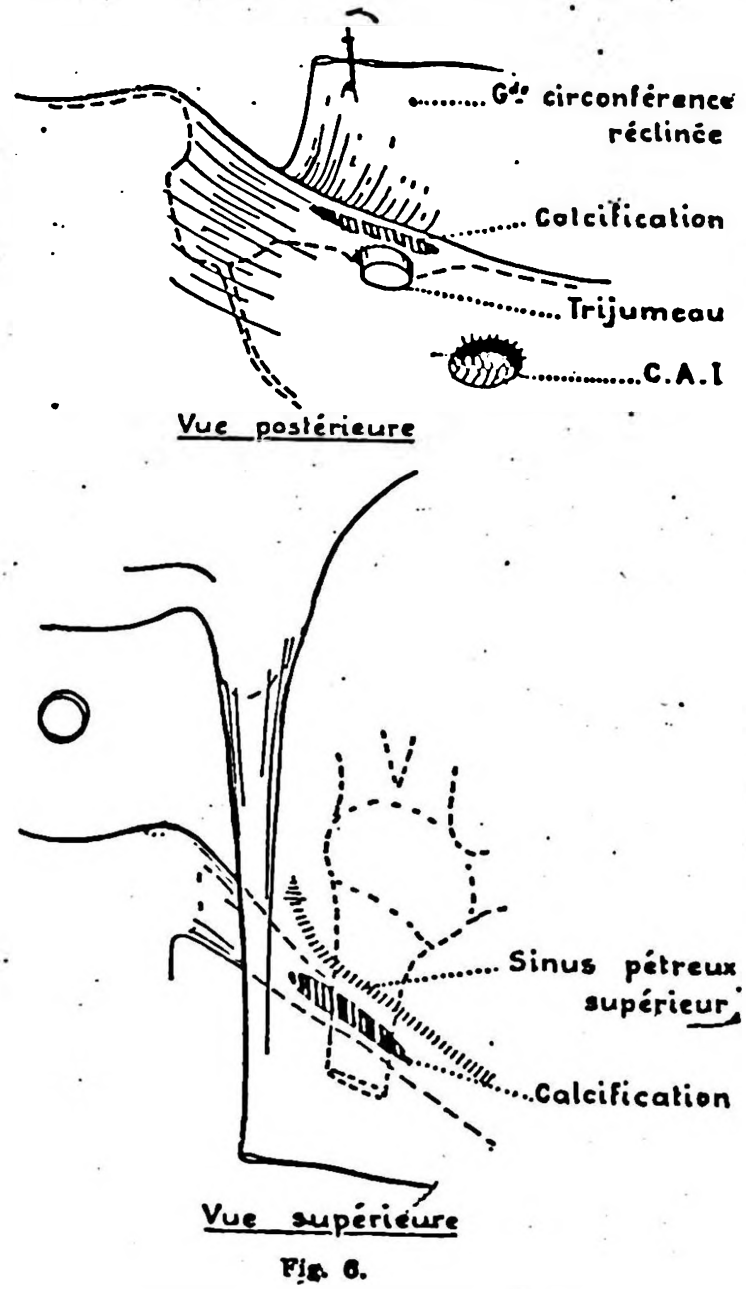

Fig. 6.

Figure 6. Intradural calcification.

orifice which left its imprint on the lower face of the Gasserian ganglion (Figure 8). Woringer and Kominoth have made an analagous preoperative verification. These intradural bony calcifications are perhaps capable of irritating the sensitive trigeminal root and of

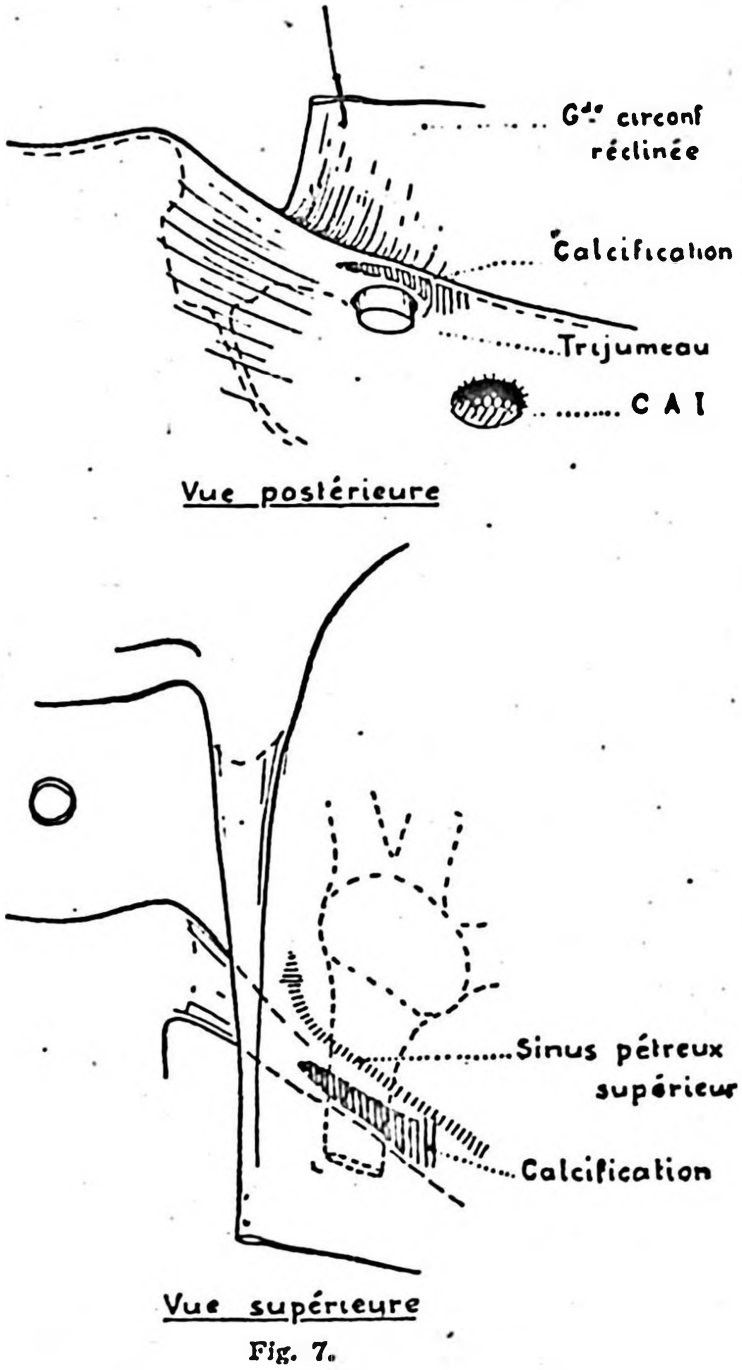

Figure 7. Osteophytic point on the higher edge of the petrous bridge.

provoking painful crises. This theory is supported by the fact that, in a certain number of cases, opening of the osteodural orifice of the trigeminus brings about relief from trigeminal neuralgia.

3. Vascular causes : Aside from the major vascular anomalies, such as aneurism or arteriovenous angioma, it is possible that, in aged patients, the vessels of the posterior fossa may take an unusual course and come into contact with the trigeminal root. The basilar artery, in particular, rarely rests on the median line and

may come in contact with the nerves of the pontocerebellar angle (Figure 9). The same pathology may also explain the origin of facial spasms,. vertiginal crises (Meniere's Syndrome), or simple acouphonia. Some authors, discussing the origin of essential facial neuralgia, confused the internal carotid contact at its exit from the

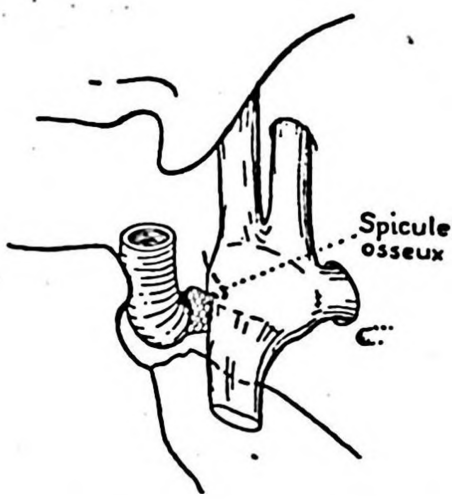

Figure 8. Bony spicule on the jagged anterior edge of the hole.

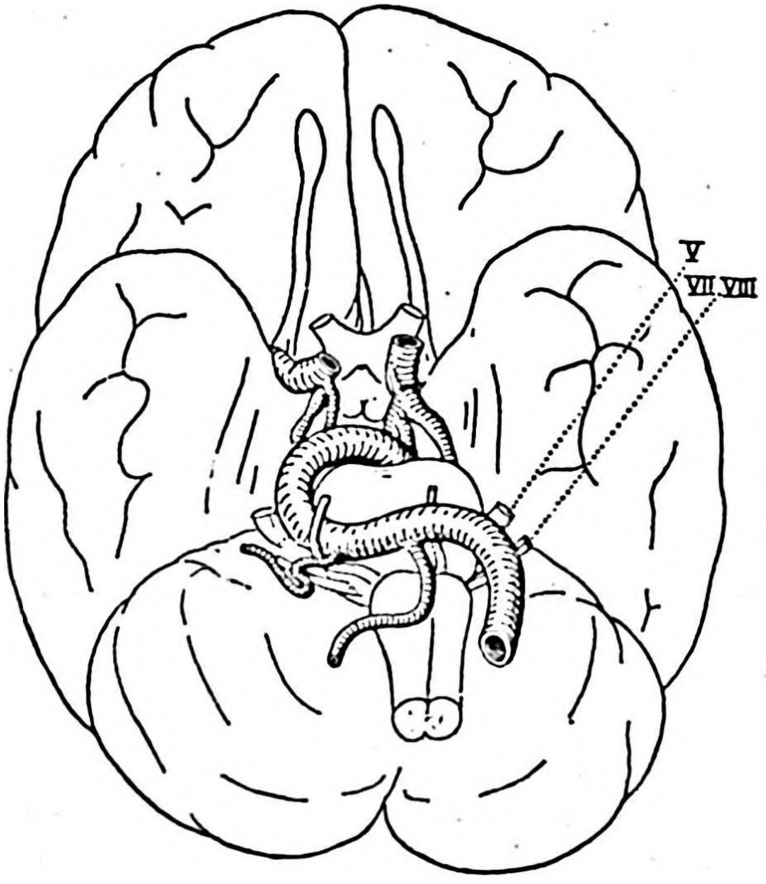

Figure 9. Compression of the fifth and eighth nerves by the arteriosclerotic and tortuous basilary trunk. (After W. E. Dandy)

carotidian canal with the Gasserian ganglion, a contact which is blamed on age because of the sinuosity of the artery and the osseous rarefaction.

In a previously published work,[6] we studied the relation between the superior cerebellar artery and the trigeminal root. At the point where the artery goes from the cerebellar trunk to the superior surface of the cerebellum, it forms one or more ansas in the ponto-cerebellar angle, and rests on the sensitive trigeminal root. It often happens that an arterial kink presses down on the root, or even insinuates itself between the radicles. We also noted that the contact between the artery and nerve occurs more often on the right than on the left; of 25 examined specimens, contact was on the right in 14 cases, on the left in four.

It is certain that in aged patients, arterial lesions involve a lengthening and hardening of the arteries. A continuous vascular contact, increased by the beating of the pulse, could cause irritation of the nerve root and thus account for the greater frequency of trigeminal neuralgia in aged people.

In conclusion, there are three possible conditions which can cause irritation or chronic compression of the sensitive trigeminal root: super-elevation of the petrous apex, calcification of the roof of the orifice of Meckel's cavity, and pressure from a tortuous and hardened artery. These conditions occur in the posterior fossa or on the higher petrous surface, and are often unrecognized because there are no clinically obvious signs. In view of these possibilities, we have developed a slight modification of the technique of retrogasserian neurotomy. We force the sectioned root into the posterior fossa, breaking its contact with Meckel's cavity and eliminating the persistent irritation.

The causes of irritation discussed here are quite ordinary, but we do not recommend their unqualified acceptance in all cases of essential facial neuralgia. The pathogenesis of individual cases must of course be verified by minute examination and operative confirmation.

Summary

The theory of a transaxonal short circuit between sensory fibers and pain fibers of the trigeminal root as an explanation of essential facial neuralgia is satisfactory in that it elucidates the character of the crises and clarifies their pathogenesis.

The etiology of these crises is often obscure and one should look for possible sources of irritation at the root of the nerve: osseous causes, super-elevation of the petrous apex, meningeal causes, and contact with a sclerotic or tortuous artery of the posterior fossa. This concept permits us to look at the pathogenesis and etiology of idiopathic facial neuralgia in a new light and to modify the technique of retrogasserian neurotomy which it suggests.

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