AMYLOID TUMOR OF THE GASSERIAN GANGLION

REPORT OF CASE

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(Received for publication September 13, 1956)

Tumors of the gasserian ganglion producing trigeminal neuralgia or trigeminal pain are not common and to our knowledge an amyloid tumor of the gasserian ganglion has not been reported previously.

We should like to report a unique case of pain, paresthesias, and loss of sensation in the face produced by an amyloid infiltration of the gasserian ganglion.

REPORT OF CASE

A 42-year-old man was first seen at the Mayo Clinic on March 28, 1956, with the complaint of pain and numbness in the right side of his face. In 1945 he had consulted a dentist after losing a cap from his right upper incisor tooth. The dentist had induced local anesthesia by injection of the right upper alveolar nerve and repaired the tooth. The numbness of the local anesthesia remained. The next day the patient consulted the dentist who was unable to explain the persistent numbness. The patient was treated with injections of thiamine without benefit. Approximately 2 weeks after the onset of the numbness the patient had noted brief, recurrent, lancinating pains in the anesthetic region. These had occurred usually in the evening. In the following years the lancinating pains had decreased in frequency and severity but the patient noted the presence of a persistent, burning, dull pain in the anesthetic region. The part affected by the numbness had slowly increased in size, particularly in the preceding year. The numbness came to extend to below the right eye and over the entire cheek, and the burning pain became severer.

The results of general physical examination were normal. The abnormal findings were limited to the neurologic examination. The right corneal reflex was greatly reduced and the analgesia and anesthesia involved part of the face supplied by the entire 2nd division of the 5th cranial nerve and the superior portion of the 3rd division. The anesthetic region extended to the midline of the bridge of the nose and the upper lip. Weakness or atrophy of the muscles of the face and jaw was not evident.

Routine examinations of the blood and urine gave normal results. The blood serology test for syphilis was nonreactive. Roentgenograms of the skull, including stereoscopic views of the base of the skull, revealed decalcification of the middle fossa on the right side near the petrous tip. Electromyography revealed the motor-unit potentials in the right masseter muscle to be large in amplitude and reduced in number and occasional fibrillation potentials were observed. This was felt to be indicative of neurogenic atrophy.

Because of the history, and neurologic and roentgenographic findings, we made a diagnosis of a tumor involving the right gasserian ganglion. Although the pain that the patient had was of the type usually associated with trigeminal neuralgia, which of course has an unknown etiology, the objective neurologic findings indicated a lesion that was destructive of nerve fibers. Operation for removal of the tumor of the gasserian ganglion was advised. The patient was told prior to operation that he was expected to have more "numbness" rather than less of the right side of the face after operation, for the surgical extirpation of a tumor involving

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* The Mayo Foundation, Rochester, Minnesota, is a part of the Graduate School of the University of Minnesota.
the gasserian ganglion usually necessitates removal of part or all of the gasserian ganglion with division of the posterior sensory root.

On April 2, 1956, with the patient under the effects of intratracheal anesthesia and in the upright position, subtemporal craniectomy was performed on the right side through a small perpendicular incision ½ inch anterior to the tragus; the middle meningeal artery was silver-clipped and divided, and the dura mater was stripped from the 3rd branch of the gasserian ganglion and from the gasserian ganglion as is done when the posterior root of the ganglion is to be exposed for root section for trigeminal neuralgia. Immediately it was noticed that the exposed portions of the 2nd and 3rd branches and the ganglion itself bulged convexly toward the craniectomy, and these structures appeared yellower than normal, not unlike the gross picture of a neurofibroma. The capsule of the ganglion was incised and the tumor, which was

under pressure, began to separate the fibers; it appeared slightly darker than the ganglion but not so dark as a meningioma. The tissue, which had a gross appearance not unlike that of an epidermoid, was gradually removed piecemeal. By means of a small curet it was possible to scoop out the neoplastic tissue not only from the mesial side of the 2nd and 3rd branches and the ganglion but also from the mesial side of the posterior root and over the tip of the petrous bone just beneath the tentorium. In removing the tumor it was necessary to sacrifice the sensory and motor roots of the ganglion. Several pieces of Gelfoam were left extradurally to control some of the oozing about the ganglion and then the wound was closed in layers without drainage.

Grossly the tissue removed was larger in amount than that from the usual "tie" operation. Microscopic verification of such tissue is routine in our laboratory and accordingly the material was sectioned immediately on a fresh-freezing microtome and stained with Terry's polychrome methylene blue. This stain exhibits a beautiful metachromatic effect with amyloid and the diagnosis of amyloidosis was immediately rendered to the surgeon.

Permanent sections made from frozen and paraffin blocks and stained routinely with hematoxylin and eosin, with Congo red, and with methyl violet brought out the characteristics depicted in the accompanying photomicrographs (Figs. 1–4). At least 50 per cent of both the ganglion and its afferent nerve bundles was replaced by a substance having all the

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Fig. 1. Amyloidosis of gasserian ganglion and its afferent nerve trunks. The granular homogeneous areas devoid of nuclei represent the amyloid deposits (hematoxylin and eosin, ×125).
staining characteristics of amyloid. The material was homogeneous and amorphous, and it occurred focally and in irregularly outlined sheets. Small nodular accumulations of it expanded the nerve trunks, and the fibers of the nerve trunks lost their identity as they coursed through the deposits. Ganglion cells were surrounded and occasionally invaded. Others appeared to have degenerated as a result of ischemia from amyloidosis of their nutrient vessels. The nutrient vessels, surprisingly enough, showed irregular involvement with intact
vessels exhibiting wide open lumens and others whose mural coats had been thickened and replaced lying side by side. On the basis of the available material the presenting picture appeared to represent primary parenchymatous involvement rather than secondary encroachment from the adventitia of blood vessels. In this aspect the case is most unusual.

The patient's convalescence was uneventful, and he was allowed to leave the hospital and go directly to his home on the 3rd day after operation. A dry dressing was applied which was to be discarded at home. Postoperative neurologic examination revealed complete anesthesia in the distribution of the right 5th cranial nerve with weakness of the masseter, temporal, and pterygoid muscles on the same side.

The patient was re-examined on June 7, 1956, at which time his condition was satisfactory. He was relieved of the pain that he had had prior to surgical intervention; the neurologic status was the same as it was immediately after operation.

More than 100 years ago Virchow coined the term "amyloid" to describe the starchlike substance present in many of the organs of persons dying from the effects of what previously had been termed "lardaceous disease." The condition was generalized or systemic and although the cause was, and still remains, obscure, it was regarded as being secondary because it almost always occurred as a late complication of chronic, debilitating, inflammatory conditions, such as osteomyelitis, tuberculosis, syphilis and the like. Parenchymatous organs such as the liver, the kidneys and the adrenal glands were the chief targets in this commonly observed form of amyloidosis.

In primary amyloidosis a substance having the same chemical and staining reactions as the material found in lardaceous disease is observed infiltrating mesenchymal tissues such as smooth and striated muscle, cardiac muscle, and muscles of the tongue, the spleen and other organs. Cardiac failure resulting from replacement
of contractile elements by infiltrates of amyloid substance is often a terminal event in patients so afflicted. Antecedent or associated conditions cannot be discovered in contradistinction to those seen in the secondary form—hence the designation "primary amyloidosis." Unlike secondary amyloidosis the primary variety is occasionally limited in distribution rather than diffuse.

Still another type of amyloid degeneration is observed in patients afflicted with multiple myeloma of bone. This type has been separated from the afore-mentioned secondary variety for, although it occurs as a consequence of the myeloma, the distribution of amyloid deposits follows the pattern set in the primary form of the disease. The amyloid material also is found rather frequently in the form of deposits within the myelomatous masses themselves and it may indeed be limited to these sites in cases of amyloidosis with myeloma.

Lastly, amyloid can occur as localized tumorous deposits which are solitary or occasionally multiple and which occur without the systemic effects of the three other forms of the disease. The upper respiratory passages and the urinary bladder account for the great majority of these amyloid tumors, the origin of which is just as obscure as the cause of amyloidosis in general.

The nervous system, both central and peripheral, enjoys a strong relative immunity to amyloid disease which is difficult to understand. Amyloid tumors of the brain have been reported by Fischer and Holfelder. Their patient had had previous therapy for squamous cell carcinoma of the right temporal region and the amyloidosis was considered to be secondary in nature. In the case reported by Saltykow three amyloid nodules were present in a brain which had been sent to him for examination. Whether these nodules represented primary or secondary forms of the disease or whether indeed they were localized amyloidomas we do not know. Bürgi's observed cerebral invasion from an amyloid tumor of the parietal bone. From our experience we would judge that this represented the amyloidosis of myeloma, having observed an entire rib replaced by amyloid substance in a comparable case. Morgenstern's observation of amyloid substance in the blood vessels surrounding a nodular sarcoma which had thickened the parietal bone and invaded the brain may have status comparable to that of Bürgi's case. We have been unable to verify Morgenstern's reference to a case reported by Schwarz of a cerebral amyloidoma in a patient with jacksonian epilepsy.

As regards peripheral nerve involvement in amyloidosis, emphasis is directed to the ischemic degeneration of these nerves as a consequence of occlusion by amyloid material of the blood vessels supplying them. Only rarely does the amyloid substance envelop, infiltrate, and destroy nerve structures. For the occasional exception to this rule the reader is referred to the 1942 review by Kernohan and Woltman.

The present report concerns an unusual instance of trigeminal neuralgia in which the etiologic basis consisted of extensive replacement of the gasserian ganglion and its afferent nerve roots by amyloid substance.

It is most interesting that the results of subsequent studies on this patient were negative for the usual causes of amyloidosis. There was no evidence for myeloma. The existence of primary systemic amyloidosis was ruled out by the absence of a large tongue, a normal electrocardiographic tracing, and other signs of mesenchymal involvement. Paunz' test with Congo red gave negative results. Between the two remaining possibilities, namely, amyloid tumor and a circumscribed form of primary amyloidosis, we cannot choose with discretion. We wish merely to present the unusual features of nerve-tissue involvement by amyloid substance.
SUMMARY

An unusual case is presented in which trigeminal neuralgia was produced by amyloid deposits in the gasserian ganglion and its afferent nerve roots. Involvement of nutrient vessels was not a prominent feature. Evidence is presented to show that the process was not part of a generalized condition, as exemplified by the well-known primary and secondary categories of "lardaceous disease."

REFERENCES