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Amyloidoma of the Gasserian Ganglion

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Summary: We report an unusual case of an amyloidoma of the gasserian ganglion associated with trigeminal neuralgia. MR imaging showed a mass in Meckel's cave, which was isointense with surrounding tissue on T1-weighted images and hypointense on T2-weighted images. After contrast administration, the mass enhanced homogeneously, and thin cuts through the region showed involvement in the expected location of the gasserian ganglion and, more laterally and inferiorly, in the proximal part of V₁. A review of the literature revealed that only one similar case has been reported since the advent of modern neuroimaging. Amyloidoma, although rare, may be considered as a rare differential diagnosis of a mass in this region.

Amyloid deposits presenting as mass lesions in the CNS are exceedingly rare. In healthy individuals, the amyloid protein does not accumulate in any organ; however, in certain pathologic conditions it may accumulate in the extracellular space and present as amyloidosis (1). If the deposit appears as a mass lesion, it is often referred to as amyloidoma. To our knowledge, only four other cases of an amyloidoma of the gasserian ganglion have appeared in the literature (2–5), and we are aware of only one other case that has been reported since the advent of modern neuroimaging (5).

Case Report

A 46-year-old woman noticed an intermittent tingling sensation in her right cheek in October 1994. A year later the symptoms had worsened to involve the right side of the nose and roof of the mouth. She also became hypersensitive to cold liquid in her mouth, and started having intermittent pain in the right cheek, upper lip, and mental region. The pain became unbearable despite medical treatment, and the patient's physician sought a neurosurgical opinion.

Upon presentation at our clinic, she reported no previous headaches, neurologic deficits, or any associated symptoms. In fact, her medical history was completely unremarkable except for some depression stemming from the long duration of her symptoms. General and neurologic examinations disclosed no abnormalities. Specifically, there was no sensory loss in the trigeminal distribution, and the corneal reflex was intact.

MR imaging of the brain showed an extraaxial mass filling and enlarging Meckel's cave on the right side, isointense with surrounding tissue on T1-weighted images (Fig 1A) and hypointense on T2-weighted (Fig 1B) and fluid-attenuated inver-

sion recovery images. After administration of contrast material, the mass enhanced brightly and homogeneously (Fig 1C–E). Thin cuts through the area of interest showed the mass to extend inferior and lateral to the gasserian ganglion, involving the proximal parts of V₁ and V₂. The extracranial distribution of the nerve appeared normal. These findings were thought to be most consistent with a trigeminal schwannoma.

A right frontotemporal craniotomy was performed with an extradural skull base approach to expose the cavernous sinus. The gasserian ganglion was noted to be pushed laterally by an underlying mass. When the undersurface of the ganglion was explored, a large 1.5-cm tumor was uncovered. The epicenter of the lesion was clearly distinguishable and was well separated from the gasserian ganglion, but on the periphery of the lesion, the amyloidoma was intermingled with fibers of V₁, V₂, and V₃, and the surgical plane was lost. The tumor was resected, sparing the motor branch of V₃, and carefully dissected off the intracavernous portion of the internal carotid artery. Intraoperative biopsy specimens revealed an unidentifiable "tumor" with inflammatory cells. Final pathologic analysis disclosed extensive amyloid deposition with focal chronic inflammation. A Congo red stain confirmed the presence of amyloid.

The patient recovered from surgery uneventfully, with complete resolution of the preoperative pain. Because the tumor was resected along with the gasserian ganglion, the patient experienced complete anesthesia of V₁ and V₂, and a small area of decreased sensation in the mental region. The eye was well protected and without irritation. A follow-up MR study 1 month after surgery showed no evidence of residual mass. There was no evidence of systemic amyloidosis or multiple myeloma.

Discussion

Amyloidomas may occur in a variety of locations, including the kidney, liver, heart, skin, lymph nodes, skull, gastrointestinal tracts, endocrine tissue, and urinary tracts (6–8). Isolated cases have even been described in the lung, eye, breast, soft tissues, skull base, and nasopharynx (9–11). In the CNS, amyloid deposits are commonly found within cortical senile plaques and in blood vessel walls, known as amyloid or congophilic angiopathy. Amyloid deposits have also been described along peripheral nerves, in autonomic ganglia, and in neurofibrillary tangles in association with dementia (12). Rarely, focal accumulation of amyloid protein has been known to appear clinically as a mass lesion in the brain (13–16), and rare cases have been reported of tumorlike amyloidosis in the orbit (17), sella turcica (18), and spinal canal (19).

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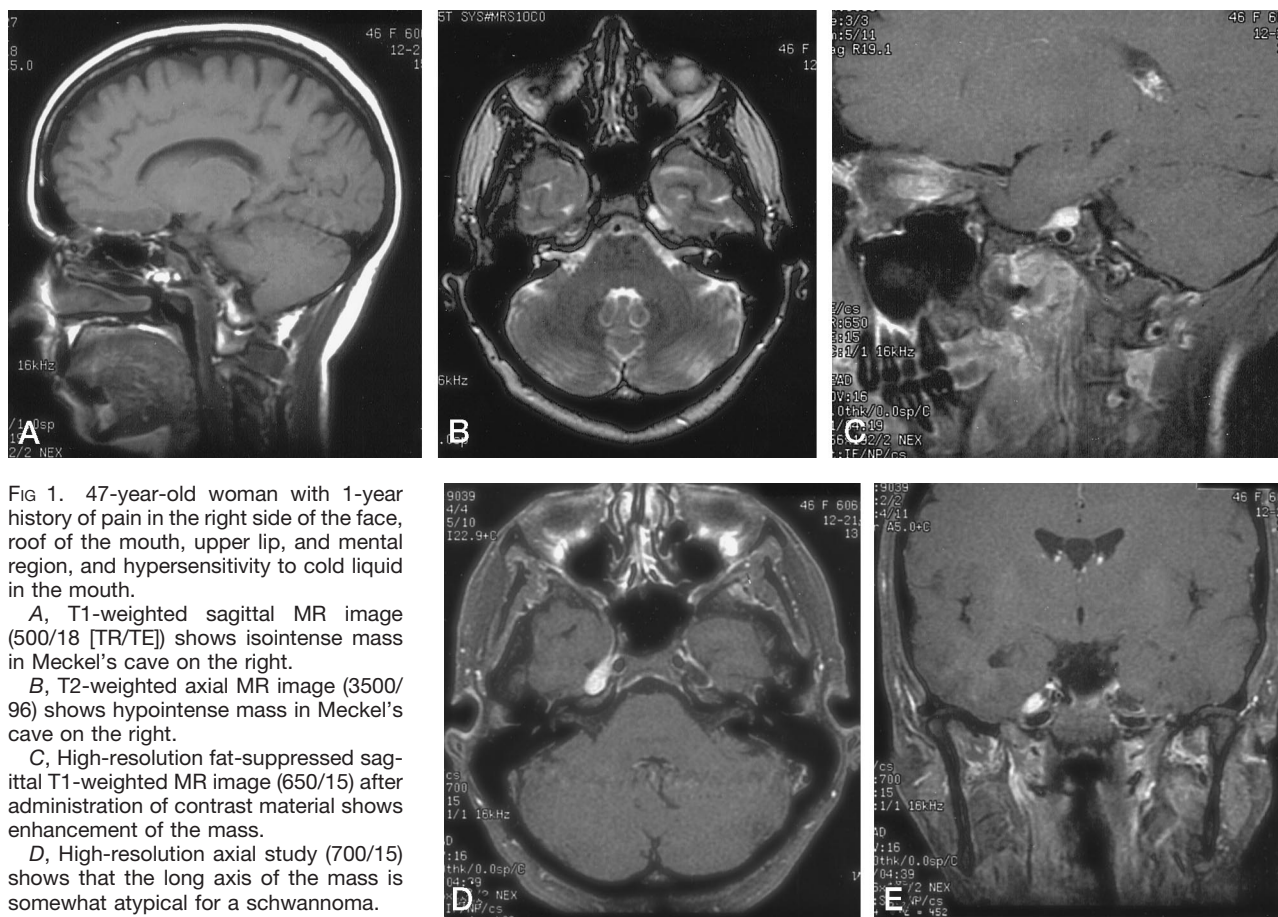


FIG 1. 47-year-old woman with 1-year history of pain in the right side of the face, roof of the mouth, upper lip, and mental region, and hypersensitivity to cold liquid in the mouth.

A, T1-weighted sagittal MR image (500/18 [TR/TE]) shows isointense mass in Meckel's cave on the right.

B, T2-weighted axial MR image (3500/96) shows hypointense mass in Meckel's cave on the right.

C, High-resolution fat-suppressed sagittal T1-weighted MR image (650/15) after administration of contrast material shows enhancement of the mass.

D, High-resolution axial study (700/15) shows that the long axis of the mass is somewhat atypical for a schwannoma.

E, On the coronal study (700/15), the mass is indistinguishable from a schwannoma.

Amyloidosis may be a primary disease with an occasional familial tendency or a secondary disease that occurs in association with an underlying chronic inflammation, infection, or neoplastic process. Examples of such conditions include, but are not limited to, multiple myeloma, osteomyelitis, and Sjögren syndrome. Rather lengthy analyses of various biochemical types and hereditary amyloid syndromes have been described, but a clinical classification is most useful (8, 20):

- Primary (AL type) amyloidosis with no coexistent disease (occasionally associated with plasma cell dyscrasia or multiple myeloma).
- Secondary (AA type) amyloidosis associated with chronic infectious disease, such as tuberculosis.
- Familial (AF type) amyloidosis.
- Focal (tumorlike) amyloidoma.
- Amyloidosis associated with aging or chronic hemodialysis.

When the disease is suspected, diagnosis depends on histologic demonstration of amyloid in tissue with appropriate stains, and subsequent differentiation between AA and AL types can be done with permanganate followed by Congo red staining. All tissue is examined for green birefringence with the polarizing

microscope. In the permanganate-sensitive AA type, the green birefringence is preserved, whereas it is abolished in the permanganate-resistant AL and hereditary types. The secondary (AA) type requires further medical workup to exclude underlying disease, such as osteomyelitis, tuberculosis, and chronic inflammatory disease (eg, rheumatoid arthritis). In the primary (AA) type, serum electrophoresis may be necessary to rule out multiple myeloma. The reasons for a solitary deposit of this protein are unknown, although it has been postulated (20) that the amyloid in cerebral amyloidoma forms with a mechanism similar to the systemic variety, with microglial cells secreting the protein instead of macrophages. One mechanism under examination includes a multifactorial process combining an inflammatory stimulus with a genetic predisposition. The protein subunit of the amyloid fibrils has been shown to be derived from a λ -immunoglobulin light chain (21), and is usually surrounded by plasma cells that are presumably responsible for focal secretion.

Little is known about the radiologic appearance of amyloidoma of the CNS. In descriptions of the CT and MR findings of brain parenchymal lesions (13, 16, 20), the CT appearance varied from hypodense to

patchy increased density, enhancing with contrast administration; on MR images, the lesions varied from hypointense on T1-weighted sequences to hyperintense on T2-weighted sequences, with mixed signal intensity, which was believed to be due to nonuniform deposition of protein. In all these studies, the authors reported varying degrees of enhancement after contrast administration, and associated dense calcifications were also noted. In the only other case of gasserian ganglion amyloidoma reported in recent years, MR imaging revealed only a mild uniform enhancement after contrast administration along the length of the trigeminal nerve (5). In our patient, the lesion appeared to be a definite solitary mass, and it would have been impossible to anticipate the diagnosis prospectively. However, retrospectively, the lack of high signal on T2-weighted images, the orientation of the long axis of the mass, and the epicenter of the mass appearing lateral and inferior to the expected location of the trigeminal nerve (the ganglion itself was not seen clearly) should have made the diagnosis of schwannoma less likely.

The general appearance of the lesion should probably prompt the differential diagnosis of trigeminal schwannoma or meningioma followed by a host of rare conditions. An extensive retrospective review of 76 cases with trigeminal neuropathy found six (8%) with masses harbored in the gasserian ganglion, including three metastases and three schwannomas (22).

Conclusion

Amyloidoma, although rare, should be included in the differential diagnosis of mass lesions affecting the gasserian ganglion and presenting with trigeminal neuralgia.

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