Amyloidomas in the cerebellopontine angle and jugular foramen

Case report

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S O-CALLED "tumor forming" masses of amyloid, or amyloidomas, have been found in bone, brain, breast, eyelids, Gasserian ganglia, larynx, lungs, lymph nodes, orbit, pituitary gland, skin, stomach, and urinary tract. In this report we present a patient with large amyloidomas in the left cerebellopontine angle and jugular foramen.

Case Report

This 47-year-old man was admitted on November 21, 1977, because of a gradual deterioration of hearing in the left ear for 15 years, hoarseness for 5 years, and left hemifacial spasm for 3 years.

First Admission. Neurological examination showed hypesthesia of the left side of the face, left facial paresis associated with hemispasm, left-sided deafness, hoarseness, dysphagia, nystagmus, and left cerebellar ataxia. A plain skull x-ray film in the half-axial projection demonstrated two linear longitudinal calcifications, 12 mm and 4 mm in length, on the left and right sides, respectively, of the upper posterior fossa. Skull tomography showed no widening of the left internal auditory canal but an erosion of the inferolateral part of the left posterior fossa. Computerized tomography (CT) revealed a large, well circumscribed, patchy, high-density mass in the left cerebellopontine angle, with several calcifications at its upper edge (Fig. 1). The mass was slightly enhanced by contrast medium. Vertebral angiography demonstrated a marked upward stretching of the left anterior inferior cerebellar artery, which encircled an avascular mass lesion in the left cerebellopontine angle (Fig. 2).

First Operation. A left suboccipital craniectomy revealed a whitish, encapsulated, well circumscribed, hard tumor in the left cerebellopontine angle. The tumor was about 6.0 cm in diameter, extending from the left trigeminal nerve root downward to the foramen magnum and possibly into the left jugular foramen and compressing the pons and medulla oblongata. The contents of the capsule were removed, then the tumor capsule was dissected from the trigeminal nerve, pons, medulla oblongata, facial nerve, and anterior inferior cerebellar artery, and removed. The postoperative course was uneventful, except for persistence of left-sided deafness and hoarseness. A postoperative CT scan showed marked dilatation of the fourth ventricle, but no mass in the left cerebellopontine angle (Fig. 3).

Second Admission. The patient did well until about 1 month prior to the second admission, when he noticed increased hoarseness, left shoulder weakness, and left glossal atrophy. On the second admission on January 6, 1984, CT scanning at a gantry angle of +30° to the canthomeatal line revealed a large, patchy, high-density mass in the left eroded jugular foramen (Fig. 4).
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There was no mass in the left cerebellopontine angle. The lesion was associated with two areas of calcification in its anterior part. The left jugular foramen was enlarged and the left hypoglossal canal was destroyed by erosion due to tumor growth. The mass enhanced slightly on administration of contrast medium.

A complete blood count was normal. Erythrocyte sedimentation rate and serum protein electrophoresis did not show any abnormalities. Quantitative immunoglobulin (Ig) testing revealed normal levels of IgG, IgA, and IgM. Serum immunoelectrophoresis was normal. Protein was not detected in the urine. Blood urea nitrogen and creatinine levels, values for uric acid and calcium, and liver function tests were normal. There were no abnormalities in subpopulations of lymphocytes. Cytological and chemical analysis of the cerebrospinal fluid was normal. There was no evidence of chronic inflammatory or infectious diseases. Review of the patient's medical and family history for amyloidosis was negative.

Second Operation. At surgery, the jugular foramen was found to be enlarged and filled with a pinkish-white encapsulated tumor. The tumor was separate from the thrombosed jugular bulb and was dissected from the surrounding bone. The anteromedial and pos-

Fig. 1. Computerized tomography scans on first admission showing a large mass with patchy high-density areas in the left cerebellopontine angle (upper), which is slightly enhanced with contrast medium (lower).

Fig. 2. Left vertebral angiography on first admission demonstrating a marked upward stretching of the left anterior inferior cerebellar artery, which curves around an avascular mass lesion in the left cerebellopontine angle.

Fig. 3. Computerized tomography scan after the first operation exhibiting no mass lesion in the left cerebellopontine angle and marked enlargement of the fourth ventricle.

Fig. 4. Computerized tomography scans at a gantry angle of +30° to the canthomeatal line on second admission. A mass with patchy high-density areas can be seen in the eroded left jugular foramen (left), which is slightly enhanced with contrast medium (right).
teromedial parts of the tumor were attached to a fine membranous tissue and the dura of the posterior fossa, respectively. The tumor was totally delivered out of the jugular foramen, and the surgical defect was closed with a free abdominal fat graft. Postoperatively, left-sided deafness, hoarseness, and left glossal and shoulder weakness persisted.

Pathological Examination. The tumors in the cerebellopontine angle and jugular foramen showed similar histological features. They were mainly composed of large acellular deposits of eosinophilic material arranged in fibrillary or honeycomb fashion (Fig. 5 left). Lymphocytes and occasionally plasma cells infiltrated around some acellular deposits (Fig. 5 right), and degenerated nerve fibers and periosteum were found at the periphery of some acellular deposits. There was no clear evidence of close association between the acellular deposits and the blood vessels. The acellular deposits were positive for amyloid staining by Congo red (Fig. 6 left), and demonstrated greenish-yellow birefringence under polarized light (Fig. 6 right). The permanganate method revealed that the acellular deposits were resistant to potassium permanganate.

Discussion

Excessive accumulation of amyloid is composed of protein fibrils measuring 7.5 to 10 nm in width, and the term “β-fibrilloses” is suggested to encompass all forms of amyloidosis. This would be consistent with the fibrillar pleated-sheet structure of the deposits, which produces the characteristic birefringence after Congo red staining.

Although amyloidosis may develop in the absence of any other recognizable disease process, its appearance is often preceded by long-standing inflammatory disorders, such as tuberculosis, chronic osteomyelitis, and rheumatoid arthritis. These observations give rise to the concept of “primary” and “secondary or reactive” amyloidosis. Immunoglobulin light chain-derived proteins, AL, constitute the major fibrillar component of amyloid associated with primary amyloidosis, multiple myelomas, and Waldenström’s macroglobulinemia. An entirely different amino acid sequence pattern is exhibited by the amyloid protein, AA, which constitutes the major component of amyloid associated with prolonged inflammatory disease and in amyloid associated with familial Mediterranean fever. In addition, the amyloid-fibril proteins in familial Portuguese polyneuropathy, medullary carcinoma of the thyroid, and senile cardiomyopathy have been designated “AFp,” “AEi,” and “ASCi,” respectively.

The amyloid protein of immunoglobulin origin, AL, appears to be resistant to potassium permanganate, as does amyloid of endocrine origin, and the application of the permanganate technique proves useful in distin-
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Fig. 6. Photomicrographs of the tumor section. Left: Congo red preparation demonstrating a reddish staining of the acellular deposits. Right: When the same section is viewed under polarized light, a greenish-yellow birefringence is apparent. × 160.

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References


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