Massive macroglossia, amyloidosis and myeloma

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Summary: A 74 year old man with light-chain myeloma developed amyloidosis with macroglossia after 10 years of therapy with alkylating agents. Over a 2-year period his tongue enlarged to persistently protrude from his mouth, inhibit his speech, interfere with normal swallowing and eventually threaten his airway. As a life-saving procedure the tumorous anterior two-thirds of the tongue was resected, with excellent primary healing. Within two weeks the patient's speech became comprehensible and his ability to eat returned to normal. Although rare in amyloidosis, massive macroglossia may occur and surgical correction is easily achieved.

Introduction

Amyloidosis is a term that describes a heterogeneous group of disorders in which various tissues contain the characteristic insoluble fibrillar protein known as amyloid. A number of different forms are recognized and these are described as senile, familial or hereditary systemic, localized, secondary or primary. In all forms the amyloid proteins have a β -pleated sheet conformation that confers upon it certain characteristic features and since several different proteins can be modified to produce this distinctive structure it has been suggested that the term beta-fibrilosis may be more appropriate.¹ In the primary type, amyloid light chain may be κ or λ and the major protein component is the variable portion of the immunoglobulin molecule; the characteristic association of these patients is with multiple myeloma. In our recent experience, which has exceeded 200 consecutive patients with this disease, macroglossia has not been encountered, although it is stated to be the most prominent oral manifestation, occurring in one-fifth of patients, and may interfere with eating, prevent the closing of the mouth and result in constant drooling.² Although amyloidosis has been reported to involve the gastrointestinal tract,^{3–6} the available case reports and photographs do not document a degree of macroglossia approaching that seen in the present patient.

We therefore report enormous enlargement of the tongue due to amyloidosis, which was successfully resected and resulted in both an excellent cosmetic effect and return of the patient's ability to eat and talk.

Case report

A 74 year old man presented in August 1976 with symptoms of spinal cord compression. Collapse of the sixth thoracic vertebral body was found and emergency neurosurgical decompression was undertaken, with good result. The excised tissue was diagnostic of an extradural plasmacytoma. Further investigations showed him to have light-chain myeloma, with extensive plasmacytosis in the bone marrow. He was treated with local radiotherapy, had subsequent complete recovery of function, and was commenced on pulsed courses of melphalan, vincristine, prednisone and procarbazine. Despite long-term therapy with maximum tolerated doses of alkylating agents there was no reduction in the degree of bone marrow involvement or quantity of light-chain excreted in the urine. His only physical limitation was the need to use a walking stick.

In mid-1983 the patient noticed a nodule on his tongue. Two years later he was noted to have general glossal hypertrophy and he experienced difficulties in accommodating his tongue within his mouth, although speech and eating remained normal. Further increase in size resulted in constant protrusion from his mouth, inhibition of speech and interference with normal swallowing and in

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September 1986 he required hospital admission because of infection. A temporary tracheostomy was performed to protect the compromised airway; biopsy confirmed the presence of amyloid infiltration.

By May 1987 the extent of his disfigurement (Figure 1) forced the patient to become a social recluse and he was now unable to take food other than liquids, speech was impossible, and the airway was again threatened. Accordingly, the tongue was resected, almost in its entirety, following preliminary tracheostomy. The procedure was carried out with a cutting diathermy by midline sagittal incision of the mass from the tip to half-way into the posterior third. The front tongue tissue on the right and the left was excised across the base, and this raw area closed by approximation of residual tongue and the mucosal edges in the floor of the mouth. Several carious teeth were removed under the same anaesthetic and the sockets sutured.

Figure 1 Anterior and lateral view of protuberant tongue. The patient was unable to retract the massively enlarged tongue, resulting in inability to eat and airway obstruction, necessitating tracheostomy.

Postoperative recovery was complicated by the development of status epilepticus, which was effectively controlled with standard anticonvulsant therapy. The tracheostomy tube was removed on the fifth postoperative day. With regular mouth care the oral wounds healed well and the gross oedema of the lower lip, which had been present prior to surgery, rapidly and spontaneously disappeared within two weeks. The patient was discharged 3 weeks after his operation and in the ensuing month the initial difficulty with talking was corrected by speech therapy. Histopathology of the tongue confirmed only amyloidosis.

Eight weeks after the operation the patient developed constricting central chest pain and died suddenly before reaching the hospital; autopsy was not obtained.

Comment

Primary amyloidosis developing in the course of multiple myeloma and characterized by the presence of amyloid light-chain in which the major protein component is the variable portion of the immunoglobulin molecule classically involves, among other organs, the tongue.² In addition, the association of Bence-Jones or light-chain myeloma and amyloidosis has been emphasized.⁷ Macroglossia is said to be extremely impressive in patients with myeloma, but was found only in 22% of the Mayo Clinic series,² and was associated with troublesome bleeding due to increased vascularity. It is noteworthy that the degree of macroglossia seen in the present case appears not to have been previously documented^{2,8} and, furthermore, information is limited on management and particularly the feasibility of surgical resection.⁹

The present experience is therefore worthy of three comments. Firstly, it is rare for patients to have a degree of macroglossia that interferes with their life style and complicates speech and ability to eat and maintain nutrition. In these latter circumstances, there is a clearcut indication for surgical removal since chemotherapy is ineffective and tracheostomy provides, at best, a compromise to sustain airway.

Secondly, the reported vascularity and associated bleeding may well have been a deterrent to more frequent use of glossectomy. However, the present case illustrates that this is not an absolute contraindication and has established that surgery for extensive resection of the tongue is feasible and can be safely achieved with minimal bleeding.

Thirdly, and perhaps surprisingly, the postoperative recovery of speech, while not complete and possibly related to the short survival of the patient,

is remarkable. Our patient was able to communicate with his family and medical attendants within 2 weeks of the major portion of the tongue having been removed and virtually as soon as primary surgery healing had occurred. In the ensuing 6 weeks there was a steady further improvement in this function and shortly before the patient died, at a clinic visit, he was able to eat completely normally and his speech was easily understood, greatly improving the quality of his life and returning him to full social activities.

It is concluded that surgical correction of massive enlargement of the tongue as a result of primary

amyloidosis is a feasible and worthwhile procedure. This operation should be considered for all patients where constant protrusion from the mouth or associated infection and bleeding are unsightly symptoms that cause psychological distress and impair eating and speech.

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