CASE REPORT

Surgical management of macroglossia secondary to amyloidosis

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SUMMARY

The authors report a case of amyloidosis-induced macroglossia treated with surgical reduction of the tongue using a keyhole to inverted T method with particular emphasis on the postoperative sequelae. Significant tongue swelling persisted for longer than anticipated requiring tracheostomy to remain in situ for 14 days.

BACKGROUND

Amyloidosis is defined as a group of complex diseases leading to extracellular tissue deposition of misfolded proteins that can be local or systemic. There are many subtypes, which are classified by the type of protein that makes up the majority of the deposits.¹ The most common; immunoglobulin light chain amyloidosis (AL), is caused by the production of misfolded light chains by clonal plasma cells.² Macroglossia is a feature in 20–26% of patients with primary systemic AL amyloidosis.³ Treatment for this is still widely contested with no formal guidelines currently in place. Surgical management of macroglossia has been reported in the literature but the associated complications and morbidity to the patient can prove problematic.⁴

We present a case of primary systemic AL amyloidosis presenting as macroglossia that underwent surgical reduction of the tongue. We discuss the specific challenges encountered in the postoperative phase.

CASE PRESENTATION

A 64-year-old woman presenting with a progressively enlarging tongue with protrusion of her teeth leading to an anterior open bite (figure 1) was referred to the oral and maxillofacial surgery department. In addition, her macroglossia was causing speech impediment, dysphagia and obstructive sleep apnoea requiring nasal continuous positive airway pressure. Medical history also included hypertension and hypothyroidism.

INVESTIGATIONS

MRI was performed to rule out pathological changes, followed by a biopsy of the tongue. Congo red and monospecific antibody staining confirmed amyloid deposits with a diagnosis of AL amyloidosis (κ subtype) (figures 2 and 3). A referral to haematology was made and the patient had extensive investigations, which confirmed that no further amyloid deposits were present in any organs, visceral tissues or bone marrow. These

Figure 1 Preoperative photograph showing severe macroglossia causing splaying of the dentition.

included cardiac, liver and renal function tests, bone marrow biopsy as well as a serum amyloid P component (SAP) scintigraphy scan, which identifies systemic amyloid deposits using a gamma ray scanner.

DIFFERENTIAL DIAGNOSIS

Macroglossia can be secondary to many acquired or genetic diseases.⁵ These can manifest as discrete lesions or a generalised enlargement. In this case, biopsy confirmed the diagnosis of AL amyloidosis (κ subtype).

TREATMENT

Due to the significant morbidity caused by the macroglossia, it was decided that surgical intervention was appropriate. The patient was consented

Figure 2 Intermediate power view of corium seen under normal halogen bulb illumination depicting Congo red positivity of these deposits, both interstitially and occupying tunica media of a medium calibre arteriole (bottom right corner).

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Figure 3 Replicate photomicrograph of corium stained by Congo red, viewed in high intensity cross-polarised light, demonstrating anomalous red/green colouration (often erroneously referred to as 'apple green birefringence') of the aforementioned congophilic deposits, in the same interstitial and perivascular location.

for a tongue reduction with the possibility of also requiring an immediate or delayed tracheostomy. After fiberoptic nasal intubation, the tongue was injected with a Lignocaine based anaesthetic. The tongue was marked up (figure 4) and a keyhole to inverted T incision was made using a bipolar cutting and sealing device (figure 5). This anterior wedge excision together with central debulking allowed for the reduction in width and length of the tongue and multilayered closure was completed with resorbable sutures (figure 6).

The patient was kept intubated for protection of the airway. Two postoperative doses of intravenous co-amoxiclav 1.2 g and

Figure 5 Intraoperative photograph showing initial incisions.

dexamethasone 8 mg were administered. This was later followed up with a course of co-amoxiclav suspension via nasogastric tube. Forty-eight hours postoperatively, it was decided that a tracheostomy was required for the patient to maintain a secure airway due to significant tongue swelling. Nutrition was provided via nasogastric feed. On postoperative day 14, the tracheostomy tube was removed and oral intake began on day 15. With the support of speech and language therapists, and

Figure 4 Intraoperative photograph with markings for keyhole to inverted T incisions.

Figure 6 Intraoperative photograph showing closure of the surgical site with resorbable sutures.

Figure 7 Postoperative photograph showing marked reduction in the width of the tongue and healing of the surgical site, however, some swelling still remains.

dieticians, the patient made a full recovery and was discharged on day 18.

OUTCOME AND FOLLOW-UP

Although the patient has had a notable reduction in the size of the tongue, some swelling did persist 2 months postoperatively (figure 7). She reported improvement in speech and was able to tolerate a soft diet, however, solid food still remained a challenge. Granted the surgery improved the patient's symptoms, amyloid regression from the tongue can be slow so the aim of this treatment was stabilisation. To reduce the supply of amyloid-forming proteins the patient may be a candidate for chemotherapy or stem cell transplant. This could decrease the risk of progression of amyloid to other tissues or organs.⁶ Therapeutic treatment for AL amyloidosis is based on antimyeloma therapy and must be tailored to each individual patient considering their age, comorbidities, extent of organ involvement and, of course, the patient's wishes. Typical regimens usually include chemotherapy agents combined with steroids, for example, bortezomib or thalidomide and dexamethasone. Monitoring of the patient's response to treatment is advised after each chemotherapy cycle by measuring free light chains or M-protein levels, with a view to switch treatment if proving ineffective. High-dose melphalan and autologous stem cell transplantation can be used in selected patients with relapsed/refractory disease or with early relapse of plasma cell dyscrasia after chemotherapy. Further SAP scintigraphy scans monitor systemic amyloid response and organ function tests should be carried out where indicated.⁷

There will be ongoing follow-up for this patient, to monitor the tongue for localised recurrence.

DISCUSSION

Tongue reduction in amyloidosis has been previously documented, but we raise the issue of significant postoperative swelling and need for tracheostomy. We originally consented the patient for tongue reduction and return to the intensive treatment unit

intubated for 24 h with the potential to perform a delayed tracheostomy. As predicted, the procedure was necessary, however, there was clearly significant swelling, which persisted far beyond the expected duration.

There are no specific guidelines to aid clinicians with the management of macroglossia secondary to amyloidosis. Multidisciplinary discussions between head and neck surgeons, haematologists and allied health professionals, are key to deciding a patient-specific treatment plan. In this case, the patient had no other systemic amyloid deposits, thus improving her long-term prognosis. In addition, she was experiencing significant morbidity due to her macroglossia, hence tongue reduction was indicated.

Many forms of tongue reduction have been described in the literature, but the keyhole to inverted T partial glossectomy allows for midline debulking as well as anterior wedge excision in order to achieve successful shortening and narrowing of the tongue while maintaining the neurovascular bundles.^{5 8}

Learning points

- ▶ Oral manifestations of systemic disease cannot be overlooked and therefore amyloidosis should be included in a differential diagnosis when macroglossia is noted.
- ▶ Surgical tongue reduction in amyloidosis can lead to significant oral swelling and, therefore, elective tracheostomy should always be considered preoperatively.
- ▶ The inclusion of allied health professionals such as speech and language therapists, dieticians and physiotherapists is crucial to a patient's successful recovery and these specialists should be involved in cases such as this from the treatment planning stage.

Competing interests None declared.

Patient consent Obtained.

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