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Case Report
Craniofacial Anomalies

Severe macroglossia after posterior fossa and craniofacial surgery in children

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Abstract. Massive swelling of the tongue can occur after posterior fossa and craniofacial surgery. Several hypotheses have been proposed to explain the occurrence of such severe postoperative macroglossia, but this phenomenon is still poorly understood. Severe postoperative macroglossia can be a life-threatening condition due to upper airway obstruction. Three cases of severe postoperative macroglossia that occurred after cervical spine, craniofacial, and posterior fossa surgical procedures are reported here. These cases required specialized maxillofacial management and a prolonged stay in the intensive care unit. Causal factors involved in this condition are reported, in order to highlight appropriate prevention and treatment options adapted to the management of paediatric patients. An overview of the current literature on severe postoperative macroglossia in paediatric populations is also provided.

Key words: postoperative macroglossia; tongue swelling; paediatric surgery; craniofacial surgery.

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Severe postoperative macroglossia (SPOM) is a massive swelling of the tongue occurring within hours following posterior fossa[1] or craniofacial surgery[2,3]. SPOM is mostly reported in adults[4]. Only a few cases have been described in children after posterior fossa and craniofacial procedures, and several of these have involved subjects with underlying congenital airway anomalies[5–9].

SPOM is responsible for longer hospital stays and increased morbidity and mortality. The management of this condition is challenging because of the potential occurrence of life-threatening airway obstruction. Some causal factors have been described, such as an inadequate surgical position during a long surgical procedure[10] and perioperative biting of the tongue by the patient[11,12]. However, the origins of SPOM are not clearly established, and guidelines for its prevention, early diagnosis, and treatment are yet to be defined. The causal factors involved in SPOM are reported herein, as well as proposed prevention and treatment options, based on three case reports and an overview of the international literature.

Materials and methods

SPOM was defined as acute postoperative macroglossia occurring within hours or days after surgery in patients without preoperative macroglossia[1–3], and which required specialized maxillofacial management and a

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prolonged (>24 h) stay in an intensive care unit. The records of the maxillofacial and plastic surgery unit of Hôpital Necker Enfants-Malades were screened, starting in the year 2000, and two cases matching this definition were identified. A third case managed at another centre was added to this series.

Case reports

Case 1: macroglossia after cervical spine surgery in mucopolysaccharidosis

A 15-year-old boy with progressive cervical spine compression due to mucopolysaccharidosis type I (Hurler syndrome) underwent a C1 laminectomy. The procedure was performed under general anaesthesia, in the prone position, after an uneventful awake nasotracheal intubation with a fibre-optic scope, and lasted for 6 h. No macroglossia had been noted in the preoperative airway assessment. No pharyngeal pack was used.

The immediate postoperative period was marked by a rapidly progressive macroglossia with impossible extubation. A dental splint was initially used to prevent a tongue biting trauma, as the tongue was rapidly externalized from the mouth (Fig. 1A, B). Specific airway issues were encountered due to the subglottic and tracheal stenosis secondary to mucopolysaccharidosis (Fig. 1C). After the placement of a bite block on day 2 postoperative, partial regression of the macroglossia was observed before stabilization of the tongue swelling (Fig. 1D). This situation lasted for 2 months and the patient could not be extubated during this period. Medical treatment with corticosteroids had no effect on the tongue oedema. Furthermore, long-term ventilator support decreased the tolerance to spontaneous ventilation because of a severe weakening of the respiratory muscles[13]. There was a high risk of death with the two alternatives to extubation – a tracheal stent and a tracheotomy – especially if these were combined[14]. After a multidisciplinary discussion, it was decided to perform a tracheotomy under extracorporeal circulation. The tongue oedema decreased within the first 24 h following the procedure. Unfortunately the patient died of tracheal obstruction after 15 days, due to tracheal infiltration, tracheobronchomalacia, and inflammatory granuloma.

Case 2: macroglossia after fronto-facial monobloc advancement

A 10-year-old girl with an unusual form of syndromic facio-craniosynostosis, including Chiari malformation, severe scoliosis, and facial anomalies, underwent Le Fort III and bipartition osteotomies with the placement of an external distraction device (RED frame; KLS Martin, Mühlheim, Germany) for the management of severe obstructive sleep apnoea (Fig. 2A). No macroglossia had been noted in the preoperative airway assessment. This procedure was performed under general anaesthesia, with uneventful oral intubation, in a supine position, and lasted for 5 h. The patient was transferred intubated to a postoperative intensive care unit.

On day 3, the patient developed tongue oedema. On day 10, the patient presented severe macroglossia. Acute management included the promotion of drainage with manual manoeuvres and tongue suspension to the distraction device (Fig. 2B, C). Corticosteroids were used to control the oedema. Local complications (biting of the tongue) were prevented using a dental wedge and a protective splint. Despite this protocol, the macroglossia persisted for 2 weeks after surgery. The tongue oedema was attributed to two main factors potentially causing tongue compression: (1) posterior tilt of the maxilla secondary to the Le Fort III osteotomy, and (2) prolonged oral intubation. On day 23, it was decided to diminish the pressure exerted on the tongue and to increase the volume of the oral cavity by performing (1) a tracheotomy after external distraction device removal (Fig. 2D), and (2) bilateral mandibular distraction using internal devices (KLS Martin) (Fig. 2E). A major decrease in tongue volume was noted immediately after mandibular distraction (Fig. 2F). The tracheotomy tube was removed on day 120 postoperative. The rest of this patient's follow-up was uneventful.

Case 3: macroglossia after posterior fossa tumour resection

A 20-month-old boy underwent a craniotomy with resection of a right cerebellomedullary angle tumour for an atypical teratoid rhabdoid tumour of the posterior fossa with spinal metastases. The procedure was performed under general anaesthesia with uneventful endotracheal intubation, in a prone position with head in flexion, and lasted for 5 h 30 min. No macroglossia had been noted in the preoperative airway assessment. During surgery, systolic blood pressure was maintained at >70 mmHg and 500 ml of diverse fluids were infused. The patient underwent magnetic resonance imaging (MRI) of the brain postoperatively before transfer to a postoperative intensive care unit (Fig. 3A, B).

The patient was extubated on the first postoperative day and significant macroglossia was observed. The patient had no signs of respiratory distress or any associated rash or lip swelling. Conservative management without reintubation was performed and corticosteroids were administered. The patient was discharged to the ward in a stable condition on day 3 after surgery. Over the next 2 days, patient assessments showed no tongue movement, no improvement in the size of the tongue, and no issues for airway management. A reinterpretation of the initial MRI showed that there was an immediate change in the anterior tongue signal without evidence of abscess, haematoma, or other cystic abnormality. On day 5 after surgery, an examination performed with a flexible laryngoscope by a surgeon from the otorhinolaryngology department identified a remaining swollen tongue protruding 2 cm past the teeth, with no evidence of pharyngeal or laryngeal swelling.

On day 11 postoperative, significant macroglossia was present but not causing distress. An MRI was performed and showed a change in the tongue signal (Fig. 3C, D). Chemotherapy for the brain tumour could be administered in the following days. The macroglossia had resolved completely at 5 weeks post-craniotomy.

Discussion

SPOM is a rare complication occurring after neurosurgical (including spine) and craniofacial procedures[21]. Seventeen cases of SPOM occurring after a limited set of surgical procedures have been reported in the literature (Table 1). Among these cases, seven occurred in children[5–9,15]. Six of the 16 cases had congenital airway anomalies[6–8,15,16]. The procedures involved were diverse, but nevertheless had two points in common: all procedures involved oral intubation and lasted for more than 2 h[17]. In most cases, the postoperative macroglossia occurred immediately or progressed rapidly during the first 48 h after surgery. The macroglossia persisted for at least 3–5 days after surgery in all cases. The authors reporting these cases often incriminated diverse sources of mechanical compression during surgery[18] and proposed a combination of risk factors, such as the duration of the procedure (more than 1.5 h), the pressure applied to the tongue secondary to the use of retractors or other oral devices[16], use of a throat

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