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Review

Congenital Epulis: Diagnosis and Management

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Introduction

Congenital epulis was first described in 1871 by Neumann and over 200 cases of this rare lesion have since been reported, with an estimated incidence of 0.0006% per live birth. It has also been described as congenital gingival granular cell tumour (CGCT) of the newborn, congenital granular cell lesion, congenital myoblastoma (historically), and Neumann's tumour. There appears to be a female predilection, with an estimated ratio of 9-10:1 female to male predominance. The most common site of presentation is the anterior part of the maxillary alveolar ridge, usually in the region of the lateral incisors or canines. However, multiple lesions may occur simultaneously in 10% of cases. Congenital Epulis appears clinically as a smooth-surfaced, sessile or pedunculated mass with typical oral mucosa colour. Tumour size may vary, ranging from a few millimetres to several centimetres in diameter and can cause respiratory and feeding difficulties. In recent years, prenatal imaging of such oral lesions has facilitated diagnosis and operative planning. We report a rare case of a large congenital epulis with striking images and highlight the use of prenatal imaging in aiding diagnosis and surgical management.

Case Presentation

A 33-year-old Para 0 presented for routine obstetric foetal anatomy scan at approximately 19 weeks gestation, to a local district general hospital. During this scan a large mass was detected appearing to involve foetal cheek or mouth.

Magnetic Resonance Imaging (MRI) of foetal brain was arranged which revealed a prominent exophytic mass arising from the right maxillary region extending from the oral cavity through the mouth *(Figure 1)*. Its maximum diameters measured 40 x 42 x 37mm (lr x cc x ap) on imaging. Importantly, the visualised upper airway appeared patent. Liquor volume was also satisfactory and this indicated that the mass was not affecting foetal swallow. There was no evidence of facial clefting.

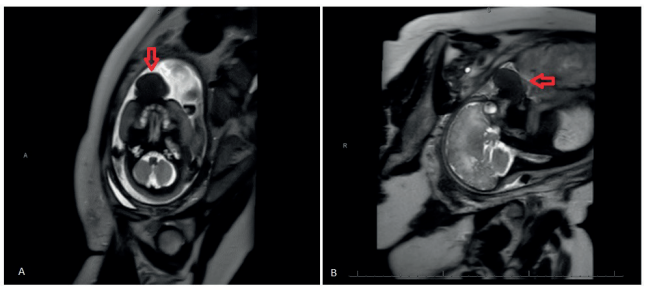

Figure 1 *T2-weighted MRI showing a mass (red arrows) arising from right maxillary region* ***(A)*** *and extending through the mouth* ***(B)****.*

Due to maternal medical history and MRI findings, her case was discussed at the Foetal Medicine Multidisciplinary Team meeting, where a decision was made to proceed with planned caesarean section under general anaesthetic, which took place at 39+3 weeks gestation. This would be coordinated with a multidisciplinary team present including neonatal, paediatric anaesthetics and paediatric Ear, Nose and Throat (ENT) for an ex-utero intrapartum treatment (EXIT) procedure if required. This takes advantage of uteroplacental blood flow and maternal-foetal gas exchange while the foetus' airway is secured with endotracheal intubation. Tracheostomy set would be on stand-by if airway could not be secured.

At delivery, infant B vocalised immediately suggesting airway was patent. A large mass was apparent, obscuring both nostrils, right cheek, right eye and most of mouth. At initial inspection, poor respiratory effort and heart rate <60 were noted. A nasopharyngeal airway (NPA) was inserted and 5 inflation breaths were given via this with initial chest lift and improvement in heart rate. The mass occluded the NPA unless retracted and intubation was planned for ongoing resuscitation. Intubation attempts proved difficult due to the large size of mass making insertion of laryngoscope difficult. The mass was vascular and friable, which meant that bleeding was a significant problem and this further obscured visualisation of vocal cords during intubation. Endotracheal tube placement was eventually established at 39 minutes of life. Airway was maintained in between intubation attempts by gentle manipulation of the mass allowing infant B to cry and breathe.

The following day, surgical excision of this large exophytic mass was performed by ENT. Intraoperatively, a large pedunculated mass was seen to arise from the right maxillary gingival surface. This was excised at the base using bipolar diathermy dissection with minimal blood loss and haemostasis was achieved with no immediate complications *(Figure 2)*. Infant B was extubated in theatre and oral feeding was established quickly in NICU. Infant B was

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then discharged to the post-natal ward on day 3 of life with discharge home on day 4.

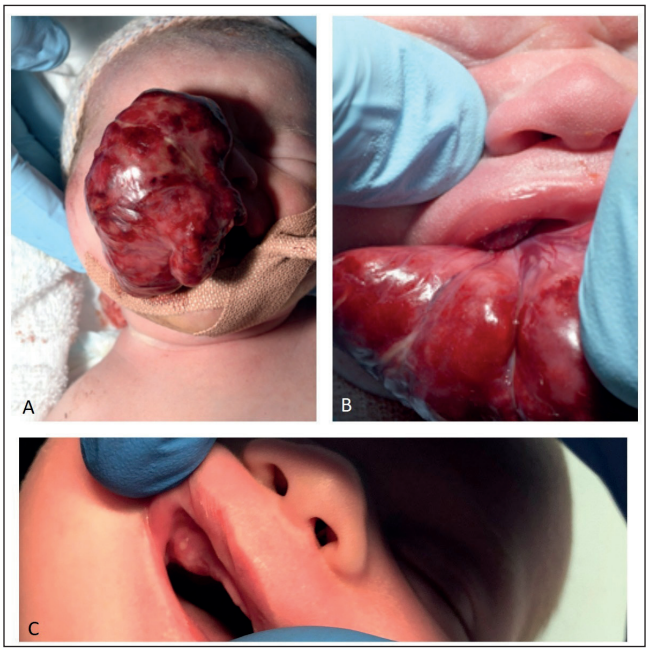

Figure 2 (A) *A large vascular mass obscuring infant's face.* ***(B)*** *Mass originated from the right maxillary gingival mucosal surface.* ***(C)*** *Post-operative image showing successful surgical excision with bipolar diathermy.*

The specimen, measuring 59 x 46 x 23mm with a weight of 36g, was sent to histopathology for analysis with the help of various immunohistochemical stains. There was diffuse fine granular cytoplasm staining with CD68, which is a marker of inflammation associated with macrophages or histiocytes[1]. Staining with S100 occasionally highlighted a specific type of histiocyte known as Langerhans' cells. CD11c, a marker for dendritic cells, was also positive. Estrogen receptor (ER) and progesterone receptor (PR) staining were negative. There were no nuclear pleomorphism or evidence of malignancy. The features therefore confirmed the clinical diagnosis of a congenital epulis.

Infant B was later followed up in the ENT outpatient clinic. On examination of the gingival mucosal surface, the excision site healed well with no evidence of recurrence. There were no issues with feeding and infant B continued to thrive. No further ENT follow-up was required and infant B was discharged.

Discussion

Congenital epulis, also known as congenital granular cell tumour or congenital granular cell myoblastoma is an extremely rare condition in the new-born with a predilection for females[2]. Although the aetiology of this oral cavity tumour is unclear, some theories suggest that it may be hormone-related[3]. A reactive theory has also been proposed in which the tumour arises from gingival stromal cells such as histiocytes, which is now considered an overarching term to describe cells of dendritic cell or macrophage lineage[4]. Immunohistochemical staining in our case supports the latter theory.

The majority of cases are recognised at birth and can cause concern for feeding or airway compromise. Prenatal diagnosis remains a challenge due to the absence of specific signs and the tendency for tumour development beyond the 22nd week of gestation[5]. In our case, the tumour was diagnosed on prenatal ultrasound at 19 weeks and subsequent magnetic resonance imaging (MRI), which proved beneficial for safe delivery planning. Although an ex-utero intrapartum treatment (EXIT) procedure was not performed in this case, it remains a viable option in cases in which significant airway compromise is a potential concern at delivery. The EXIT procedure takes advantage of uteroplacental blood flow and maternal-foetal gas exchange while the foetus' airway is secured safely with endotracheal intubation and has shown to improve outcomes in cases of airway obstruction[6]. Members of the EXIT team should include an experienced paediatric anaesthetist, ENT consultant and neonatologist. The ENT surgeon in this case ensured tracheostomy kit was available at delivery for emergency tracheostomy if endotracheal intubation could not be established.

Classically, a congenital epulis comprises a single firm tumour with a regular surface and can be sessile or pedunculated, but in some cases multiple tumours may occur. It most frequently occurs at the maxillary location opposite the future canines or incisors, but the mandibular region can also be involved. A range of tumour sizes has been recorded from a few millimetres to around 10 centimetres at its widest diameter[7], therefore this case was relatively large.

Although benign, immediate surgical excision is recommended for larger lesions as there is a significant risk of airway compromise. Other clinical manifestations may include dyspnoea and difficulty feeding[8]. Prenatal MRI is therefore recommended in determining the characteristics of the mass to aid surgical planning[9]. Surgical excision can be performed either under local or general anaesthesia and this depends on size and location of the tumour. The literature suggests a preference for excision under general anaesthesia when tumour is large in size, such as in this case[10].

Conclusion

Large congenital epulis, although benign, requires surgical removal due to risks to airway and feeding. Prenatal MRI can be a useful imaging modality and a multidisciplinary approach to management should be adopted. Follow-up is essential to recognise recurrence after surgical removal.

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