CASE REPORT

A hairy situation

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SUMMARY

A 2-day-old neonate was transferred to a specialist paediatric otolaryngology centre with stridor at rest, feeding difficulties and an apparent mass in the oropharynx. The newborn displayed evidence of respiratory distress, however, she remained self-ventilating. MRI highlighted a fat-containing lesion in the postnasal space with no intracranial extension. A CT scan under general anaesthesia showed no underlying bony abnormality, and hence an examination of the nasopharynx, oropharynx and microlaryngoscopy and bronchoscopy were performed. The macroscopically hairy lesion arising from the superior aspect of the soft palate was resected. Histology displayed a benign growth measuring 28×17×12 mm in keeping with a hairy polyp. This is one of very few cases, to the best of our knowledge, in which a hairy polyp (bigerminial choristomas) has resulted in stridor in the first few days of life. Nasal masses in neonates, although a rare phenomenon, remain clinically important as they are obligate nasal breathers.

BACKGROUND

Congenital oro/nasopharynx masses presenting in the neonatal period resulting in acute respiratory distress and stridor are rare, although a recent review of the literature suggests that the diagnosis is often missed and may be more common than previously anticipated.¹ Their clinical significance, however, is paramount due to their anatomical location. Neonates are predominantly obligate nasal breathers, therefore masses in the postnasal space may present with acute respiratory distress and also cyanosis during feeding, resulting in failure to thrive. Prompt identification and management of this condition can result in the avoidance of significant morbidity and mortality. Hairy polyps, first described in 1918 by Brown-Kelly,² are histological benign lesions derived from two germinal layers, a mesodermal central core (giving rise to cartilage, adipose tissue, muscle) surrounded by ectoderm (giving rise to epithelium, sweat glands, hair follicles and sebaceous glands).

CASE PRESENTATION

A 2-day-old neonate was admitted to a district general hospital after presenting with acute onset stridor, respiratory distress and difficulty feeding. The neonate was promptly transferred to a specialist paediatric otolaryngology service after initial review.

The neonate was born by caesarean section at 39 +2 for a breech presentation; the delivery was otherwise unremarkable (APGAR score 6, 9, 10). The child had a normal cry at birth with no documented

cyanotic episodes during feeding. The mother was otherwise fit and well with no significant medical history. There were no other siblings (gravida 1 parity 1) and no family history of note.

On arrival, the child displayed features of stridor and respiratory distress with signs of increased trouble breathing. Although distressed, she maintained her own airway at all times, with normal oxygen saturations. The child showed no dysmorphic features or cleft lip, and examination of the oropharynx revealed a pedunculated polyp touching the tongue base, which was evident on crying. The remaining ear nose and throat examination was unremarkable.

INVESTIGATIONS

Flexible nasal endoscopy confirmed a bilaterally patent choanae and a pedunculated polyp within the postnasal space, surrounded by secretions.

MRI confirmed a well-defined lobulated 2.3×1.2×8.9 mm mass lying within the postnasal space; the mass showed high T1 and T2 signal, with saturation of the high T1 signal on the fat sequence suggesting a fatty substrate (figure 1). The MRI showed no evidence of intracranial extension, making other differentials less likely.

CT scan was performed under general anaesthesia prior to surgical intervention to ensure there were no concurrent bony abnormalities.

Figure 1 MRI 2.3×1.2×8.9 mm mass within the postnasal space. No evidence of intracranial extension.

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DIFFERENTIAL DIAGNOSIS

The differential diagnoses for a child presenting with stridor are extensive, and thus a thorough history from the parents is paramount followed by a full examination. In the work up of a post-nasal space mass in a neonate, imaging is essential and should include MRI and CT. MRI delineates the soft tissue lesion and excludes the presence of intracranial extension, which can be present in encephaloceles, meningocele and intranasal gliomas. CT assesses for any underlying bony abnormality, which can be of variable significance depending on the age of the child and ossification.

Differential histological diagnoses include teratoma, hamartoma, epidermoid, haemangioma, nasal glioma, neuroblastoma, meningocele and lingual cyst.³

TREATMENT

Following complete radiological assessment, an examination under anaesthesia was performed of the naso/oropharynx with subsequent microlaryngoscopy and bronchoscopy (figure 2). The pedunculated lesion was noted to arise from the superior aspect of the palate on the right side. The base of the pedicle was divided using bipolar cauterisation transorally using a 120° endoscope for visualisation (figure 3). No other abnormalities of the airway were identified. The child was extubated successfully, and was fed via nasogastric tube overnight with close observation in the neonatal intensive care unit. Histology confirmed a polypoid lesion covered by mature skin consisting of epidermis with underlying adnexal structures including pilosebaceous units and eccrine sweat glands, additionally there was subcutaneous mature fibro fatty tissue with scattered islands of mature skeletal muscle and cartilage (figures 4–6).

OUTCOME AND FOLLOW-UP

The child managed full feed 4 days postprocedure and was successfully discharged. Follow-up postoperatively showed no evidence of failure to thrive and she made a good clinical recovery.

The child was subsequently diagnosed with cystic fibrosis. Although an association between inflammatory polyposis and cystic fibrosis is well recognised secondary to hyperviscous secretions and obstruction, no direct correlation between hairy polyps and cystic fibrosis could be delineated in the literature.

DISCUSSION

Arnold's taxonomy subdivides germ lesions of the nasopharynx into four recognised subclassifications. Hairy polyps fall under

Figure 3 Post resection. Hairy polyp measuring 3.3 cm.

the category of dermoids and are bigerminal (compared to teratomas, which are composed of all three germ layers—see below).

1. Dermoids (eg, hairy polyps): Derived from epidermal and mesodermal germ layers. The matrix is composed predominantly of fatty tissue.
2. Teratoids: Composed of poorly differentiated tissue derived from all three germ layers.
3. Teratomas: Trigeminal, but more differentiated; contain histologically recognisable organoid structures.
4. Epignathi: parasitic fetuses derived from all three germ layers and usually incompatible with life.

Hairy polyps are the commonest congenital tumour of the naso-oropharynx, presenting predominantly in the neonatal period and infancy. They display a strong female preponderance and commonly arise from the lateral wall of the nasopharynx or superior aspect of the soft palate,⁴ but have also been described at the Eustachian tube orifice, tongue and tonsil.⁵

Clinical presentation is dependent on the site of manifestation and size. A recent review of 78 patients (64 articles) showed that respiratory distress and obstruction were the most common presenting features, followed by feeding/swallowing difficulty.¹ Other symptoms include vomiting, haemoptysis, coughing, unilateral Eustachian tube dysfunction, snoring and failure to thrive. Interestingly, a review of presenting hairy polyp diameter and clinical manifestation suggests that hairy polyps of less than

Figure 2 Polyp in situ visualised transorally using 120° scope arising from the soft palate.

Figure 4 A polypoid lesion with appearances of normal skin and underlying skeletal muscle and adipose tissue (x20).

Figure 5 Appearances of normal skin and underlying cartilage and adipose tissue (×20).

Figure 6 Appearances of normal skin including hair follicles and eccrine sweat glands (×20).

3 cm in diameter have a higher risk of respiratory distress, as they are more likely to be missed on routine examination.⁶ We present a unique case of a neonate presenting with stridor, as well as the more common features of respiratory distress and feeding difficulty.

Hairy polyps are not known to be associated with any underlying congenital disorder and are commonly found as the sole underlying defect, however, rare associations include hard/soft palate abnormalities and agenesis of the uvula.⁴ In this case, the diagnosis of cystic fibrosis was made at a later stage; we do not believe this to be linked, and have found no supporting literature.

Once appropriate radiological investigation has been performed, as described above, surgical excision with a combined transoral/nasal approach resolves symptomatology. Malignant transformation and recurrence is not a recognised sequelae, and once resected it has a good prognostic outcome.

Learning points

- ▶ A thorough ear, nose and throat examination should be undertaken in neonates presenting with stridor/respiratory distress to assess for rare differential diagnoses.
- ▶ MRI ± CT imaging should be undertaken in lesions arising from the postnasal space to exclude intracranial extension.
- ▶ The 120° endoscope provides helpful visualisation of the postnasal space when definitive treatment is undertaken.

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