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CASE REPORT

Acute upper airway obstruction by a hairy polyp in an infant

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ABSTRACT

A respiratory distress in the newborn is a severe, life-threatening emergency. Hairy polyp (HP) is an uncommon developmental lesion. It develops because of anomalies in 2 germinal layers. The aim of this paper is the presentation of a newborn girl with an acute respiratory distress in the course of a HP located in the left soft palatal wall. The lesion was successfully removed. HP of the oropharyngeal region must be included as a differential diagnosis of acute respiratory distress in infants. A transoral surgical approach allows successful and minimally invasive excision even in a newborn. Diagnostic imaging is essential to outline the HP site of origin, which can simplify surgical treatment.

KEY WORDS:
hairy polyp, respiratory distress, newborn.

INTRODUCTION

Respiratory distress in a newborn is a severe, life-threatening emergency. It has a high mortality rate and remains the leading cause of hospitalization in neonatal intensive care units (NICUs). The inherent cause of upper airway obturation in a newborn varies; it could be due to head and neck malformations (teratomas, airway haemangiomas, lymphatic malformations or papillomas, thyroid goitres, neuroblastomas, or neural tube defects) or congenital high airway obstruction syndrome [1, 2].

Tumours occurring in neonates are rare, with an estimated incidence rate of approximately 1 in every 12,500-27,500 live births [3]. Hairy polyps (HP) are an uncommon developmental lesion. They develop because of anomalies in 2 germinal layers (the mesoderm-fibroadipose tissue, cartilage, and muscle fibres; and the ectoderm-epithelium, hair follicles, and sebaceous and sweat glands) during embryogenesis at varying levels of differentiation. HP presents as a pedunculated tumour mainly comprised of fatty tissue covered by skin. In most cases, HP is seen within the first month after birth, and depending on its location and size it can cause stridor, tachypnoea, cyanosis, desaturation, respiratory obstruction, and dysphagia. It can potentially cause fatal respiratory failure.

A case of acute respiratory distress in the course of an HP located in the left soft palatal wall in a newborn girl is presented.

CASE REPORT

The patient, a girl, was born at 39 weeks of gestation by caesarean delivery. The indication for the planned caesarean section was breech presentation. The neonate weighed 2.95 kg (10^{th}–25^{th} percentile) and was 52 cm long (75^{th}–95^{th} percentile); she had Apgar scores of 7, 8, 8, and 9 at 1, 3, 5, and 10 minutes after birth, respectively. She was the second child of a 30-year-old woman and her 40-year-old husband. Two earlier spontaneous miscarriages were reported. In the first trimester of the pregnancy, the expectant mother had experienced light bleeding, vaginal and urinary tract infections, and anaemia. In the

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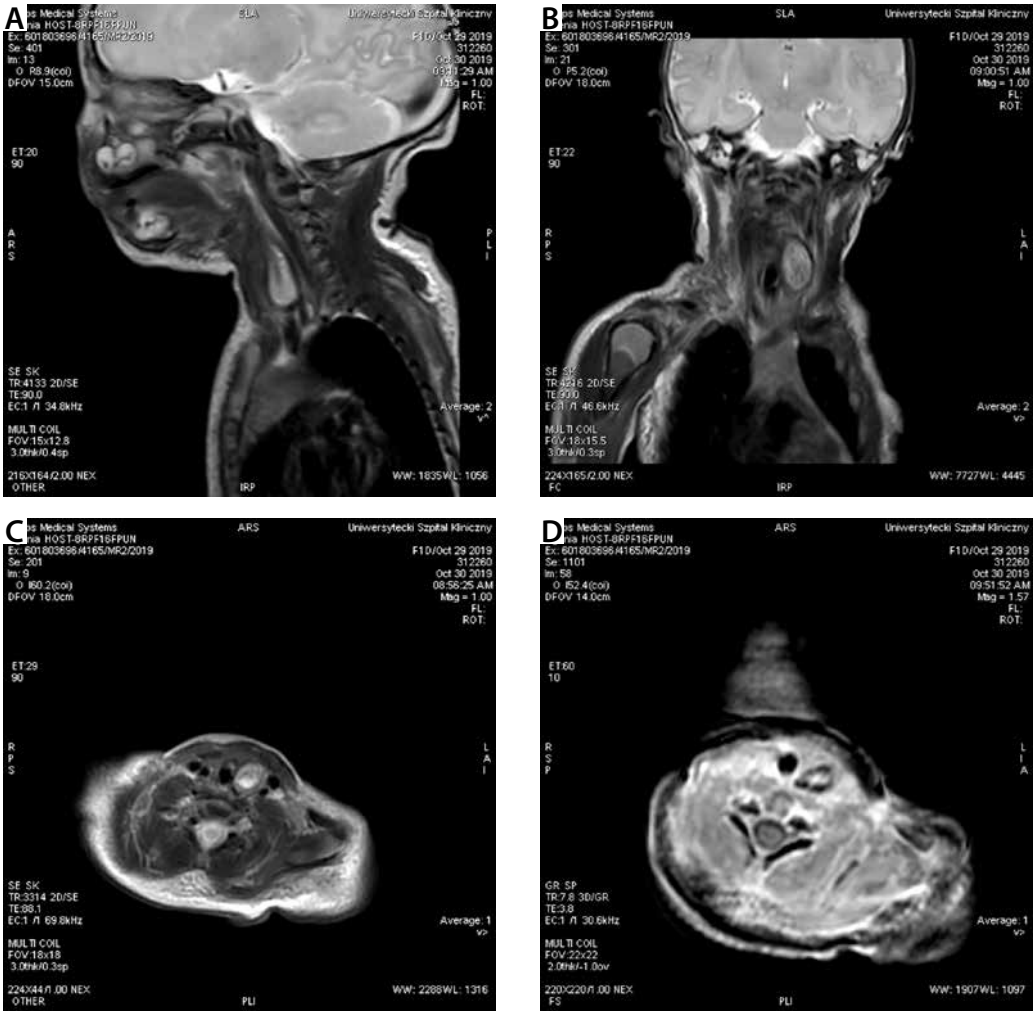

FIGURE 1. Preoperative magnetic resonance imaging in T1- and T2-weighed imaging with Fat Sat option in sagittal (**A, B**) and axial (**C, D**) planes, showing the presence of a well-defined, pediculated soft-tissue, iso-intense lesion located within the left side of the oropharynx

13th week of gestation she was admitted to the Obstetrics and Gynaecology Clinic with a threatened miscarriage. During pregnancy she was treated with progesterone, amoxicillin, amoxicillin + clavulanic acid, and ferrous sulphate + ascorbic acid. The child's mother smoked 2 cigarettes daily throughout the pregnancy. Her GBS status was positive. The infant was born with decreased breathing effort, a cough, and large amounts of foetal secretions in the respiratory tract. In the 20th minute of life, the girl presented signs of acute respiratory obstruction. Physical examination revealed a pedunculated, fleshy mass arising from the oropharynx and obturating the upper aero-digestive tract. The infant was intubated with a 3.5 endotracheal tube and transferred to the NICU. Magnetic resonance imaging was performed to delineate the anatomic abnormality. It revealed the presence of a well-defined, pediculated, soft-tissue, iso-intense lesion located within the left side of the oropharynx (Figure 1). On the 5th day of life, under general anaesthesia, a well-circumscribed, firm mass was completely removed from the left soft palatal wall through a transoral route by cold dissection and bipolar electrocautery (Figure 2). The patient was returned to the NICU and was extubated successfully on the second day after the operation. No further pulmonary complications were noted. Subsequent examinations did not identify any recurrent growth of the mass, and findings on the otolaryngological exami-

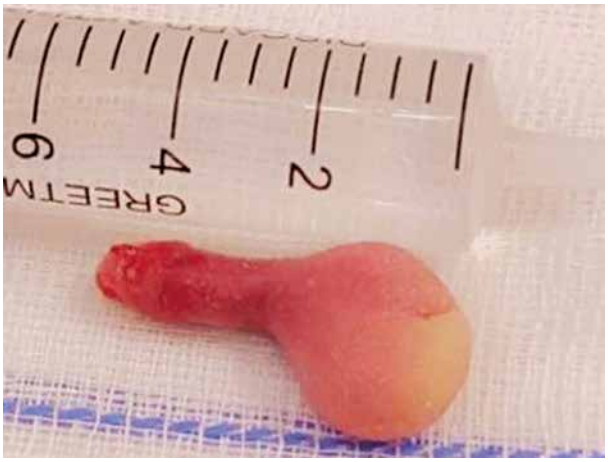

FIGURE 2. Photo of the excised specimen, measuring approximately 4 cm in length

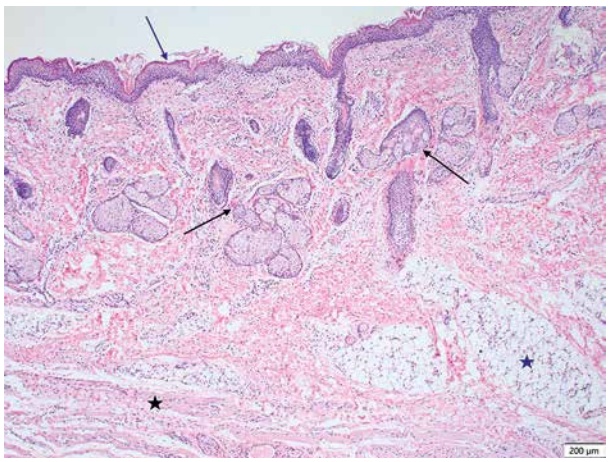

FIGURE 3. Microscopic view of the hairy polyp. The polyp is covered by skin-like keratinized squamous epithelium (blue arrow) with pilosebaceous units (black arrows). The core of the tumour is composed of fibroadipose tissue (blue asterisk) and skeletal muscle bundles (black asterisk). HE, 40×

nation were completely normal. Histological examination revealed a polypoid tumour covered by a skin-like surface composed of keratinized squamous epithelium and pilosebaceous units. Fibro-adipose tissue and skeletal muscle bundles were identified in the core of the polyp. No necrosis or atypia were found. These histological features were consistent with a diagnosis of HP (Figure 3). At the 6-month follow-up, the girl showed no signs of lesion recurrence.

DISCUSSION

This report describes a newborn with acute respiratory failure secondary to an upper-airway obstruction. Acute airway distress in infants is a common cause of intensive care unit hospitalization [4] that may subsequently result in cardiac arrest. Clinical symptoms that are considered indicators of acute respiratory distress are irregular breathing, inspiratory breathing, tachypnoea, bradypnoea, or apnoea, nasal flaring, grunting, retractions (subcostal, intercostal, supracostal, or jugular), cyanosis, inspiratory stridor, wheezing, and hypoxia [5]. Stridor is created by a turbulent air flow through the narrowed airway lumen and indicates a compromised airway that demands prompt and proper evaluation of its aetiopathology. Based on the location of the stenosis, it can be inspiratory (above the vocal cords), expiratory (lower trachea), or biphasic (at the glottis or subglottis). Stridor may subsequently lead to dyspnoea, which is a life-threatening condition. The most common cause of laryngeal stridor appearing immediately after birth or in the first weeks of life is laryngomalacia. The differential diagnosis should include airway masses – e.g. vascular malformations, papillomas, or cysts – congenital anomalies of large vessels (vascular rings), and abnormalities of the central and peripheral nervous system. Nasopharyngeal obstruction should also be considered, including nasal or airway secretions, congestion, choanal atresia or stenosis, enlarged or redundant upper airway tissue or tongue, and nasopharyngeal tumours [6].

Prenatal diagnosis of an upper-airway obstruction allows strategies to be planned for management of the newborn's airway during delivery. The ex-utero intrapartum treatment (EXIT) [7, 8] enables assessment of the obturation and airway control on placental support. During caesarean section, the newborn emerges from the uterus to chest level while foeto-maternal circulation is supported through the umbilical cord, providing time to restore airway control either by bronchoscopy, intubation, tracheostomy, or tumour excision. EXIT is considered to be an optimal and safe treatment option, especially in foetuses with concurrent polyhydramnios, ascites, hydrops, enlarged lungs, flattened or inverted diaphragm, intrinsic obstruction of the upper airway, or solid neck mass and potential congenital airway obstruction [9, 10]. Foetal airway obstruction proceedings should include a detailed evaluation of prenatal imagery and the involvement of an experienced interdisciplinary team. EXIT is recommended in cases of endophytic lesions or a primary mass in the neck area. Caesarean delivery seems to be sufficient in the absence of respiratory distress, as in cases of exophytic tumours that grow out through the foetal mouth [11]. In our case, EXIT was not planned because there was no suspicion of foetal airway anomalies.

Different terms, often used interchangeably, have been used to define HP, including teratoma, dermoid, nasooropharyngeal choristoma, and hamartoma [12-19]. A choristoma is a developmental anomaly resulting in the development of a tumour-like tissue mass of normal cells in an ectopic location. It may present mixed tissue types, including bone, cartilage, glial tissue, gastric mucosa, and tumour-like masses of the sebaceous glands. A hamartoma is a non-neoplastic, unifocal or multifocal developmental malformation that consists of various cytologically normal, mature cells and tissues that are typical of the anatomic location, showing a disorganized architectural pattern with a dominant single component. A dermoid cyst is the most common form of teratoma. It is a tumour composed of all 3 germinal layers, with a predominance of ectodermal elements. Histologically, it is composed of stratified keratinized epithelium with cutaneous structures, like hair and sebaceous and sudoriparous glands. Some authors categorize HPs as teratomas. Head and neck teratomas are very rare and mostly benign tumours, found predominantly among the paediatric population. The oropharyngeal cavity is one of the rarest sites for congenital teratomas (2% of all teratomas) [20]. In our case, teratoma was ruled out in the main differential diagnosis, due to an absence of endodermal components. However, HP is a malformation of bigerminal origin with both ectodermal and mesodermal elements. The bigerminal origin of HP remains in contrast to teratomas, which are

composed of cell types representing all 3 blastodermic germ layers, indicating the totipotent nature of their cells. In contrast to teratomas, HPs do not show malignant potential.

HPs mostly appear in female neonates and with a left-sided predilection (6.5 times more commonly involved). Dutta *et al*. stated that the left-sided preponderance is a result of the cooperation of Hox, which regulates the epithelial–mesenchymal interaction, and sonic hedgehog (*SHH*) gene products, which influence left-right asymmetry during morphogenesis [14].

Clinical differential diagnoses of the lesions include oropharyngeal teratoma, cystic hygroma, lymphangioma, duplications, and neuroblastoma. The pathogenesis of HP is unclear. It is thought to be a developmental anomaly because several congenital conditions – such as cleft palate, low-set ears, osteopetrosis, hypospadias, left carotid artery atresia, agenesis of the external auricle, and bifurcation of the tongue – are known to be related with this type of lesion [21]. Some authors have stated that HP may be thought of as displaced tissue from the first pharyngeal pouch or second branchial arch [15, 22], considering its connection with various branchial arch malformations: absence of the uvula, external auricle, ankyloglossia, and facial hemihypertrophy [23].

The most common location of HP is the naso-oropharynx: 60% of such lesions develop in the nasopharynx; most of them derive from the lateral nasopharyngeal wall or the eustachian tube, while 25% are located on the tonsil or tonsillar pillars. The hard palate, middle ear, tongue, and soft palate are considered rare locations.

As in the case reported here, conservative complete surgical excision is curative, and because the lesion may be only suspected upon clinical examination, histology and immunohistochemistry are mandatory for a definitive diagnosis.

CONCLUSIONS

HP in the oropharyngeal region must be included as a differential diagnosis of acute respiratory distress in infants. A transoral surgical approach allows for a successful and minimally invasive excision, even in a newborn. Diagnostic imaging is essential to outline the HP site of origin, which can simplify surgical treatment.

DISCLOSURE

The authors declare no conflict of interest.

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