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Duplication of the cervical esophagus: An unusual cause of respiratory arrest in a child☆

Mengu Ma Tooh [a], Oumama El Ezzi [a], Mirko Dolci [b], Yann Boegli [b], Christos Ikonomidis [c], Anthony de Buys Roessingh [a,*]

[a] Department of Pediatric Surgery, Centre Hospitalier Universitaire Vaudois, CH-1011 Lausanne, Switzerland
[b] Department of Anaesthesia, Centre Hospitalier Universitaire Vaudois, Lausanne, Switzerland
[c] Department of Otorhinolaryngology, Centre Hospitalier Universitaire Vaudois, Lausanne, Switzerland

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ABSTRACT

Duplications of the cervical esophagus are rare malformations. When the malformation is situated in the thoracic part of the alimentary tract, symptoms can be dyspnea, regurgitation, cough, epigastric discomfort and anorexia. We report the history of a ten-month-old child who presented progressive onset of snoring and stridor, in the course of an upper respiratory tract infection, not responding to antibiotics and drug therapy. The baby developed acute respiratory distress at the hospital in supine position, due to major airway compression, leading to respiratory arrest. Resuscitation was provided and the child underwent an emergency decompression of the cystic mass.

Digestive duplications may occur anywhere from the mouth to the anus. They can be tubular, cystic, diverticular, isolated or multiple. The small intestine is the most common site of duplication and the esophagus the second [1]. Differential diagnoses are bronchogenic cysts, branchial cysts or vascular malformations. A duplication situated in the thoracic part of the alimentary tract is potentially dangerous because it may induce respiratory distress, due to compression and/or tracheomalacia [2,3]. Esophageal duplications may induce respiratory distress in children and therefore should be mentioned in the differential diagnosis of airway obstruction.

1. Case report

A ten-month-old girl, without previous medical history, consulted at the pediatric emergency service for difficulty in breathing and unusual respiratory sounds. Symptoms had started 2 weeks before with snoring and coughing without fever. She was first treated with antibiotics by her pediatrician. Two weeks later, the baby seemed to have more difficulty in breathing. There was no history of epigastric discomfort, cough or regurgitation. Because the symptoms were becoming more and more severe, she was taken to the closest emergency unit by the parents. Physical examination showed that the baby's breathing was noisy with a slight stridor. Her respiratory rate was 44/min. Supine position seemed to increase respiratory distress while prone position did not. During clinical evaluation, the baby had a respiratory arrest; respiratory resuscitation was achieved without intubation. A chest X-ray showed an enlarged mediastinum with important tracheal deviation (Fig. 1). She was then transferred to our hospital for further investigations.

She was apathetic and tachypneic. Oxygen saturation was between 97% and 100% under 50% of oxygen support by mask. As she was definitely better in prone position, with the head turned on the right side, we decided not to move her for the clinical examination and first investigations.

Local status showed a left cervical mass, mobile, painless at palpation and visible in the left cervical region next to the sternocleidomastoid muscle, with no visible cutaneous sinus. It measured 4 × 5 cm. A computed tomography (CT) scan showed a cystic mass full of liquid in the parapharyngeal area, going from the left submandibular region to the upper mediastinum space, situated in

* Corresponding author. Tel.: +41 79 556 37 67; fax: +41 21 314 30 71.
E-mail address: anthony.debuys-roessingh@Chuv.ch (A. de Buys Roessingh).

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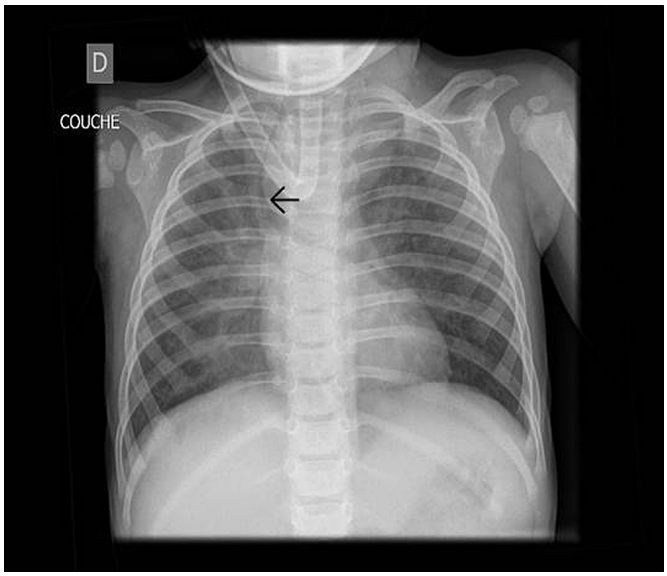

Fig. 1. Chest X-ray, with deviation of trachea (arrow) and widening of the upper mediastinum.

depth and close to the carotid artery and jugular vein with major compression and deviation of the trachea (Figs. 2 and 3).

An emergency drainage of the mass was organized. Anesthesia was induced by inhalation of Sevoflurane®, in prone position. She was positioned on her left side to allow more efficient ventilation. As she was still breathing spontaneously, without airways obstruction, we positioned her after a few minutes in a supine position and proceeded to an endotracheal intubation, without any drugs other than the volatile agent. The puncture of the cyst under ultrasound (US) control obtained a whitish milky fluid. A left dissection of the neck was realized to expose the wall of the cyst which was biopsied. A catheter was left in the cyst. Oxygenation and ventilation were uneventful throughout the procedure. The child was transferred, still intubated, to the intensive care unit.

Pathology confirmed the diagnosis of esophageal duplication and a second surgery was performed 2 weeks later for the complete removal of the malformation through a cervicotomy. At the same time, we performed a laryngoscopy to exclude the presence of a tract originating from the left pyriform fossa seen in pyriform sinus cyst. A

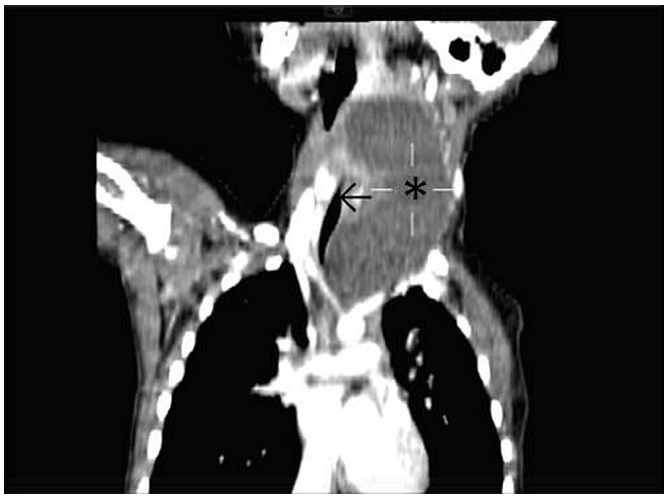

Fig. 2. Initial computed tomography (CT) scan, with a mass (*) creating a major compression of the airway (arrow).

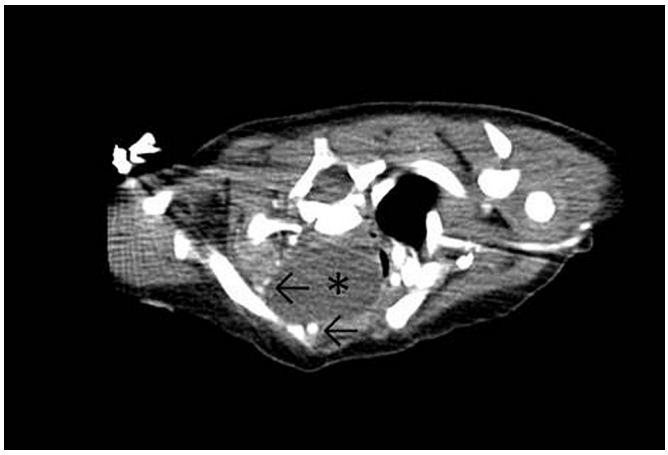

Fig. 3. Computed tomography (CT) scan image, in prone position, showing an ovoid cervical lesion (*) next to the esophagus and the neck vessels displaced laterally (arrows).

fistulous connection between the cyst and the native esophagus was identified, ligated and covered with a flap of fibrous tissue. Macroscopically, the mass was soft, weighed 29 g and measured 3.5 × 3 cm. Microscopic examination showed that the inner wall was lined with stratified squamous epithelium. The outer wall consisted of a thick band of smooth muscle fibers surrounded by skeletal muscle fascicles. The epithelium was strongly positive for various cytokeratins, especially for cytokeratins 19 and 125. These last two are usually seen in the gastro-intestinal epithelium. Neither cartilaginous tissue nor lymphoid tissue were present. The structure of the wall, with its type of epithelium, smooth muscle and skeletal muscle bands, was compatible with esophageal duplication.

A contrast study of the esophagus five days later showed no leakage. A transitory palsy of the left recurrent laryngeal nerve was observed. The child left the hospital 5 days after the operation, by which time she had completely recovered. One month after the operation, the baby ate normally and pre-operative stridor had disappeared.

2. Discussion

This case describes an unusual cause of respiratory arrest in a ten-month-old girl suffering from esophageal duplication. The fact that the child had no history of chronic respiratory symptoms, no digestive tract disturbances and no palpable cervical mass, when she was first admitted, made the evaluation of the risk of respiratory arrest a real challenge.

Duplications of the alimentary tract are rare malformations and the cervical portion of the esophagus is rarely concerned. More than 80% of the cases are diagnosed before the age of two years. Since the first description by Balasius in 1711, only few cases have been reported in the literature [3–5]. Multiple theories have been proposed to account for esophageal duplication origin, but no single theory adequately explains all the known duplications [6]. The most common anatomical presentation is a single cyst, located on the lower part of the esophagus [7,8]. However, Robinson et al. reported a case of multiple cysts in one patient [9]. These malformations can be associated with other malformations such as esophageal atresia, tracheoesophageal fistula, spinal deformities, congenital heart disease, vertebral abnormalities, malrotation of the bowel, diaphragmatic hernia and Meckel's diverticulum [10].

The clinical presentation is usually nonspecific and depends on the anatomic localization, size and nearby organs. An ectopic gastric mucosa is found in approximately two thirds of the duplications,

predisposing the mucosa to ulceration and perforation [11]. In the case of cervical esophageal duplications, the clinical sign is the presence of a cervical mass. Early surgical intervention is required to prevent complications of the duplication [12].

The interest of our case is the presentation of an anterior mediastinal mass, with an initial diagnosis of upper respiratory tract viral infection and a sudden respiratory arrest influenced by the position of the baby. As the acute onset of stridor after an episode of upper respiratory tract viral infection may be very common, the baby was first treated conservatively. In the literature, such cases have either respiratory symptoms or the presence of a cervical mass. Moulton et al. described a case of a 13 month old boy with a history of chronic (10 months) biphasic stridor and dyspnea while feeding [2], and Wootton-Georges et al. described the case of a newborn with a large cervical mass [5], both being duplications of the cervical esophagus. As we did not have a diagnosis for the mass, we therefore planned a surgical drainage of the cyst in order to secure the airway tract, even if this could increase the risk of bleeding, infection and distortion of the anatomy.

Induction of anesthesia in patients with anterior mediastinal mass can lead to cardiorespiratory collapse [13]. Inhalation induction and spontaneous ventilation maintenance are widely recommended [14,15]. The position of the patient can be supine, lateral decubitus, semi-Fowler or prone [13,16]. In case of airway compression during the procedure, the first measure is the repositioning of the patient in the position that caused the least symptoms pre-operatively [15]. As our patient was breathing freely in the prone position, we didn't reposition her for the induction of anesthesia. Depending on the position of the mass, rigid bronchoscopy could be another option. In case of persistent life-threatening cardiovascular issue, the use of cardiopulmonary bypass is often recommended in the literature, but a canulation – even with a dedicated team on stand-by – may require too much time and impede a resuscitation. Immediate sternotomy and elevation of the mass by the surgeon is probably a better option [15].

We describe here a true duplication of the esophagus with a mass that was attached to the native esophagus and an identified fistulous connection to the native esophagus that was ligated. The wall of the cyst lined by a stratified squamous epithelium, encircled by bands of smooth muscle as well as skeletal muscle fibers are all features of the upper part of the esophagus. The embryological etiology of this malformation is not completely understood, but both esophageal duplication and bronchogenic cysts are believed to occur at the time of the division of the embryonic foregut [17]. Histological similarities support their common origin, but a true esophageal duplication contains all the layers of a normal esophagus (mucosa, submucosa and muscularis propria), but no cartilaginous tissue. The presence of cartilaginous tissue or lymphoid tissue in the malformation as well the type of epithelium seen may suggest another malformation such as a bronchogenic or a branchial cyst (third or fourth). The classic location of bowel duplication is on the mesenteric side, sharing a common muscle wall and blood supply with the associated bowel [18].

3. Conclusion

Although they are very rare conditions, esophageal duplications should be mentioned in the differential diagnosis of children with respiratory distress not responding to the usual medical treatment and without defined etiology. Furthermore, according to the clinical presentation of this pathology, the preoperative management can become a challenge for the various medical teams.

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