—Report on Experiments and Clinical Cases—

Fatal Tracheo-innominate Artery Fistula after Tracheostomy in a Patient with Pelizaeus-Merzbacher Disease

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Abstract

Tracheo-innominate artery fistula (TIF) is a serious, life-threatening complication following tracheostomy. We report a fatal TIF in a 15-year-old girl with Pelizaeus-Merzbacher disease. She received a tracheostomy for prolonged translaryngeal intubation due to acute respiratory failure without a trial of noninvasive ventilatory support before intubation. Severe hemorrhage from the TIF occurred 6 months after tracheostomy; immediate resuscitation failed. Antemortem fiberoptic bronchoscopy showed tracheal stenosis accompanied by granulation tissue, and postmortem examination revealed TIF with ulcerative granulation. Preventive intervention is required to avoid catastrophic TIF due to its high mortality rate. Moreover, to avoid prolonged translaryngeal intubation leading to tracheostomy, noninvasive ventilatory support before translaryngeal intubation, if applicable, is beneficial.
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Key words: tracheo-innominate artery fistula, tracheostomy, complication, Pelizaeus-Merzbacher disease, noninvasive ventilation

Introduction

Tracheostomy is a common surgical procedure used to maintain long-term ventilation. The American Thoracic Society published a statement for the care of children with chronic tracheostomy in 2000[1] stating that proper tracheostomy management should consist of an appropriately sized tracheostomy tube, gentle maneuvering during tracheostomy care, and close monitoring of the tracheal lumen with flexible bronchoscopy; however, various complications have been reported in the early and late phases[2]. Tracheo-innominate artery fistula (TIF) is a fatal complication following tracheostomy in which a fistula communicates between the anterior tracheal wall and the posterior wall of the innominate artery. Close juxtaposition of the innominate artery and trachea in the upper-anterior mediastinum lays the foundation for future TIF[3]. Airway management without prolonged translaryngeal intubation is required to avoid

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tracheostomy and its complications, i.e. noninvasive ventilation (NIV) support is often helpful for patients with acute hypoxemic respiratory failure[4].

In this article, we describe a fatal TIF in a 15-year-old girl who died of massive hemorrhage 6 months after tracheostomy; TIF was confirmed with postmortem examination. The tracheostomy had been performed to replace prolonged translaryngeal intubation; however, she had not received NIV support before translaryngeal intubation when acute respiratory failure developed. Congenital leukodystrophy of the central nervous system[5], also called Pelizaeus-Merzbacher disease (PMD), had been diagnosed when the patient was younger.

Case Report

A 15-year-old girl was referred to our department because of prolonged fever and cough despite having been treated with oral medications and intravenous antibiotics at the referring outpatient clinic. She had tachypnea, and the SpO_2 was 90% breathing room air. She also had had a postprandial cough for the past few days. Chest X-ray examination **(Fig. 1)** revealed scoliosis and perihilar infiltration, indicating aspiration pneumonia. Because of nystagmus and delayed cognitive developmental, PMD had been diagnosed when the patient was an infant, although she did not take any medications and had not been hospitalized; furthermore, she had never had feeding issues with her daily peroral thick gruel meal.

She was admitted to our department on the first day of her visit and given supplemental oxygen, intravenous antibiotics, and fluid hydration. Peroral feeding was suspended during the first few days of admission. In the convalescent phase, she resumed eating thick gruel; however, feeding was halted because of choking episodes. Transnasal or transcutaneous gastric tube feeding was recommended to avoid anticipated recurrent episodes of aspiration, but her family withheld consent. During the course of swallowing rehabilitation, she had repeated episodes of choking that resulted in aspiration pneumonia, severe respiratory distress, and hypoxemia, for which an endotracheal tube was placed. An extubation attempt failed because of severe dyspnea and stridor despite NIV support.

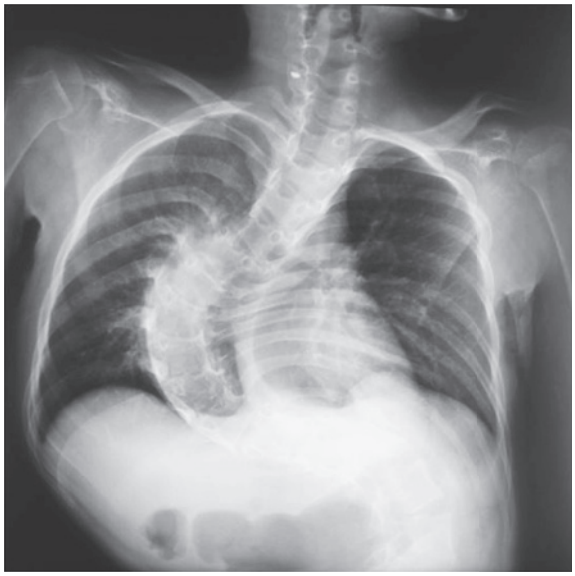

Fig. 1 Chest X-ray examination on the patient's first visit to our hospital revealed scoliosis and perihilar infiltration.

Following the failure of the second extubation attempt because of severe laryngeal stridor, tracheostomy was performed. Preoperative computed tomography (CT) revealed lordosis and juxtaposition of the trachea and innominate artery behind the sternum. One week after tracheostomy, the tracheal tube was exchanged without respiratory distress. However, stridor and spasmodic respiratory distress occurred in the third week after tracheostomy. Examination with a flexible bronchoscope revealed swelling of the tracheal epithelium at the mid-tracheal level; in addition, the tracheal lumen at that level collapsed during inspiration **(Fig. 2)**. A smaller tracheal tube was inserted emergently beyond the swollen portion, and systemic steroids were administered. Thereafter, the respiratory symptoms resolved, and the swollen tracheal epithelium diminished within a few weeks. Despite the smaller tracheal tube and minimal cuff pressure, similar episodes recurred occasionally; minimal bleeding from the tracheal lumen sometimes followed these episodes.

Tracheal reassessment with contrast-enhanced CT 2 months after tracheostomy revealed that the

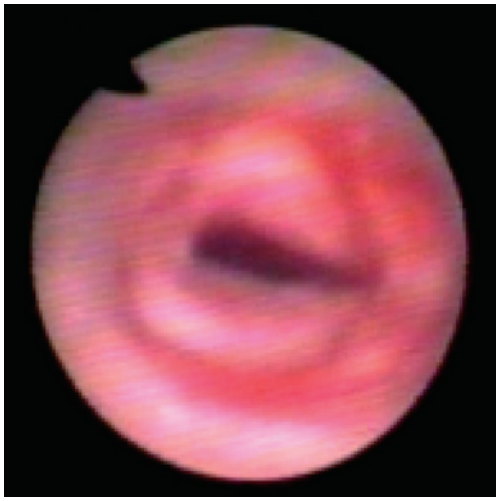

Fig. 2 Flexible bronchoscopic examination revealed tracheal membrane swelling at the mid-trachea.

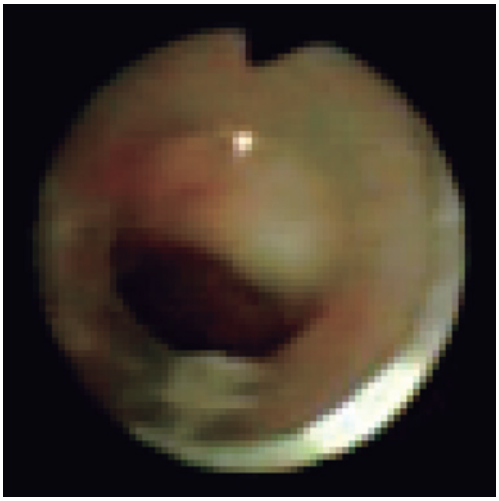

Fig. 4 Flexible trans-tracheal tube bronchoscopic examination revealed tracheal stenosis accompanied by granulation tissue on the tracheal lumen.

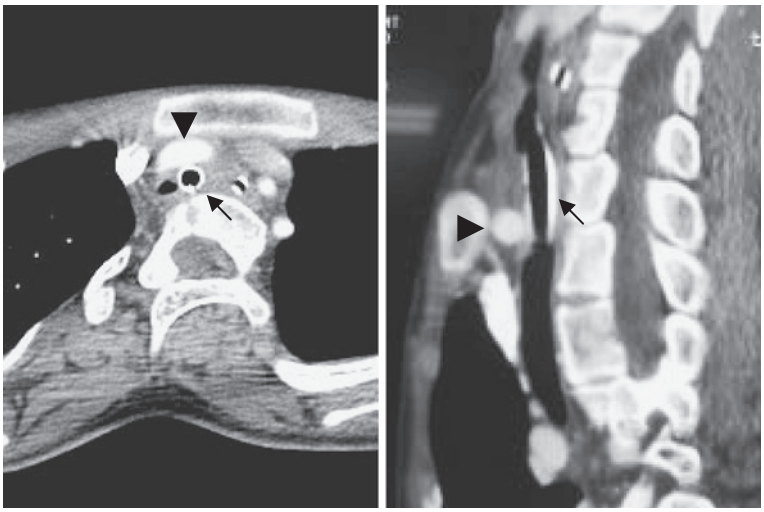

Fig. 3 Contrast-enhanced computed tomography scan from the neck to the chest indicated lordosis and juxtaposition of the innominate artery; the trachea is pinched between the top of the sternum and the spinal column (**arrow**: trachea with tracheal tube, **arrowhead**: innominate artery).

cervical trachea was filled with the tracheostomy tube and that the tip of the tube was set behind the innominate artery **(Fig. 3)**. Plans to address these findings with a tracheal stent or innominate artery occlusion were suspended because conservative treatment with a smaller tracheal tube had temporarily relieved the symptoms. The tracheal stenosis had occurred through obstruction by granulation tissues **(Fig. 4)**. Methicillin-resistant *Staphylococcus aureus* (MRSA) was detected in the sputum during treatment. Sudden massive hemorrhage from the trachea occurred 6 months after tracheostomy; the patient died despite an immediate attempt of cardiopulmonary resuscitation.

Autopsy was performed with the family's consent. The trachea was incised from the posterior side; thereafter, ulcerative granulation was evidenced on the anterior wall of the mid-trachea **(Fig. 5a)**.

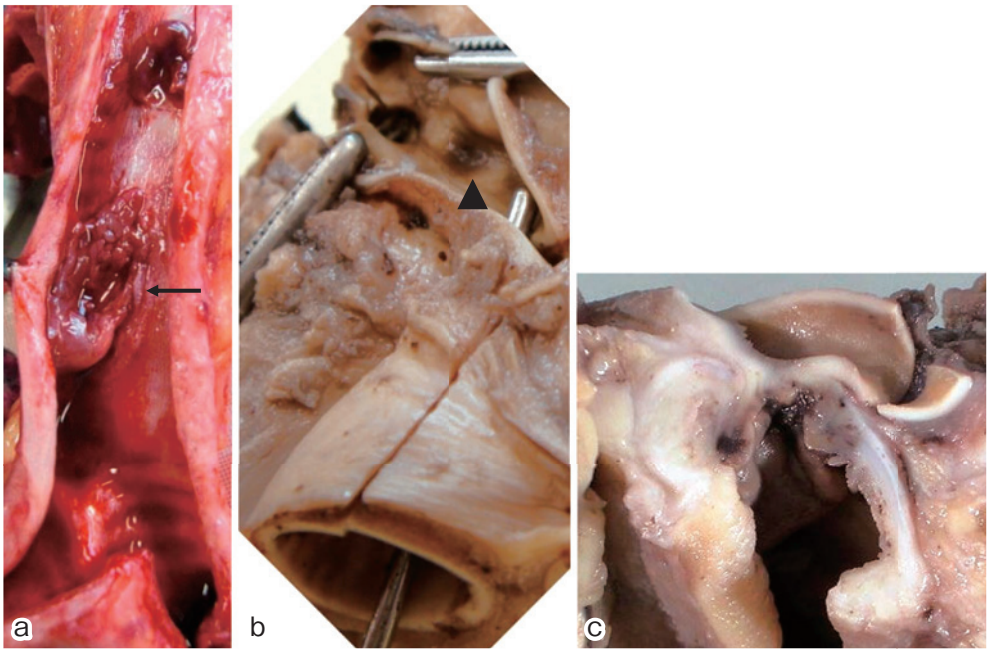

Fig. 5 (a) Ulcerative granulation was identified on the anterior wall of the mid-trachea (**arrow**).
(b) **Arrowhead** indicates a small pit at the posterior wall of the innominate artery.
(c) An incision through the level of ulcerative granulation and the small pit failed to reveal tracheal cartilage.

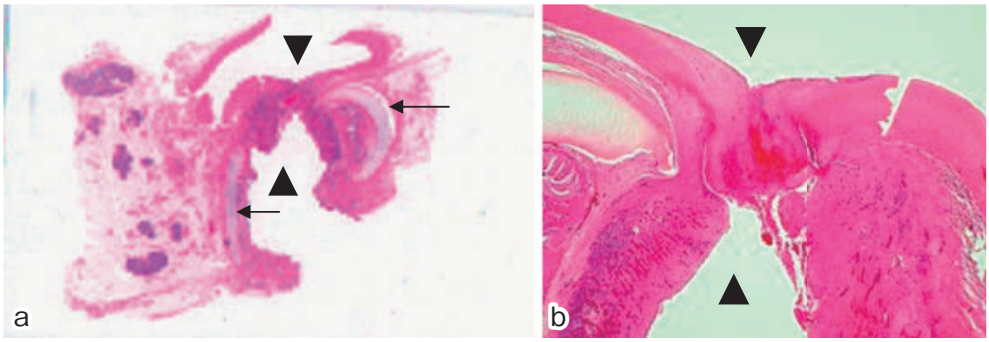

Fig. 6 Histopathological examination revealed the defect in tracheal cartilage and granulation tissue accompanied by inflammatory cells and fibroblasts alternating with tracheal cartilage (**arrow**: tracheal cartilage, **downward arrowhead** [▼]: innominate artery, **upward arrowhead** [▲]: tracheal lumen). (a) Low-power field; (b) high-power field

Furthermore, a small pit at the posterior wall of the innominate artery was identified adjacent to the ulcerative granulation tissues on the anterior wall of the trachea **(Fig. 5b)**. An incision through the ulcerative granulation and the small pit failed to reveal tracheal cartilage **(Fig. 5c)**. Histopathological examination revealed a defect in the tracheal cartilage that was occupied with granulation tissue accompanied by inflammatory cells and fibroblasts **(Fig. 6a, b)**. These pathological findings confirmed the diagnosis of TIF. No obvious ulcers or granulation were observed in the right or left main bronchus. The vesicles in the lung parenchyma were filled with clotted blood.

Discussion

Tracheostomy is a life-saving procedure; its most common indication is anticipated prolonged ventilatory support, especially for patients receiving

mechanical ventilation via translaryngeal intubation[6]. The reported rate of complications following tracheostomy ranges from 4%[7] to 65%[8], with an average risk of approximately 15%[7]. The reported mortality rate of patients undergoing tracheostomy ranges from 0.5% to 1.6%, most often caused by hemorrhage or tube displacement[7]. Complications of tracheostomy are divided into 3 groups according to the time of occurrence: immediate (0–24 hours following tracheostomy), intermediate (1–7 days), and late (>7 days). TIF and tracheal stenosis are usually late complications.

Some degree of tracheal stenosis develops after tracheostomy in 40% to 60% of patients[9]; however, no symptoms occur until the lumen area has decreased by 50% to 75%[8]. Tracheal stenosis typically results from bacterial infection or chondritis of the anterior and lateral tracheal walls with growing granulation tissues in the tracheal lumen. The first symptom in the present case suggested tracheal stenosis. Using properly sized tracheostomy tubes, inflating cuffs only when indicated, and maintaining intracuff pressure at less than 15 to 20 mm Hg may decrease the incidence of tracheal stenosis[8]. In the present case, tracheal stenosis occurred after only 3 weeks of routine management, thereafter the cuff pressure having been maintained at less than 10 mm Hg.

Scahlaepfer reported initial review of 115 cases with fatal hemorrhage after tracheostomy, and concluded that those hemorrhage were caused by TIF[10]. Incidence of TIF is reported to be as high as 1%[3]; the average is estimated to be 0.3%[11]. The mortality rate of severe hemorrhage from TIF without immediate resuscitation or surgical management is estimated to be 100%. Wood et al.[12] have reported that only 24 of 175 patients survived, even with rapid surgical treatment.

The mechanism of injury in TIF is necrosis following erosion of the anterior tracheal wall and cartilage adjacent to the posterior wall of the innominate artery[11]. Three mechanisms induce pressure necrosis with the tracheostomy tube[3]. The first and most common mechanism involves fistula formation caused by pressure necrosis from an overinflated high-pressure (> 20 mm Hg) tracheostomy cuff. The second mechanism is the tip of the cannula impinging against the anterior tracheal wall. The third mechanism is abutment of the elbow of the cannula against the innominate artery. As mentioned above, the cuff pressure was maintained at less than 10 mm Hg in the present case.

The innominate artery usually crosses the anterior trachea at the level of the 9th tracheal ring behind the manubrium but can cross anywhere from the 6th to the 13th tracheal rings[2,10]. A high-lying innominate artery, which can reasonably be expected to cross the trachea above the 4th tracheal ring or sternal notch, is a risk factor for fistula formation, particularly in thin and young patients[13].

Neuromuscular diseases that involve encephalitis are the most common diagnoses underlying TIF[14]. Patients with neuromuscular diseases often have spinal deformities and abnormal posture, such as scoliosis, lordosis, and neck extension, as observed in our case; these result in the innominate artery crossing the trachea at higher levels; however, TIF associated with PMD has not, to our knowledge, been previously reported.

A high-lying innominate artery, infection, weakened local tissue, steroid therapy, and malnourishment also contribute to TIF[15]. In the present case, MRSA colonization and the need for long-term intubation, even with low cuff pressure across the stenosis, might contribute to a chondritis and the progression of TIF.

The current management guidelines for TIF call for prompt diagnosis, rapid hemostasis, and definitive operative repair of the TIF or catheter occlusion of the innominate artery. Up to 50% of patients with TIF have had previous sentinel hemorrhages, as did our patient[14].

Furthermore, to avoid long-term intubation and tracheostomy, a practical palliative approach, such as NIV support, is essential before translaryngeal intubation. NIV support for children is reported to be useful[4,16]. The benefits of NIV support are its easy application and rapid assessment of its effects (i.e., within several hours)[16]. However, some patients cannot tolerate nasal or full-face masks; failure to keep the mouth shut would lead to air leaks, poor

efficiency, and poor efficacy. Although we did not try NIV support before translaryngeal intubation in the present case, we have changed our airway management policy to give priority to NIV support, especially in cases of acute respiratory failure.

PMD is an X-linked recessive leukodystrophy caused by a mutation of the proteolipid protein gene[9]. Leukodystrophies affect the growth of the myelin sheath that covers nerve fibers in the central nervous system. The extent of axonal injury increases with age and may account for the progression of neurologic signs, including poor swallowing function, in patients with PMD.

In conclusion, complications can occur in the early and remote phases after tracheostomy; TIF is potentially fatal and can develop despite prudent airway management following tracheostomy with a low-pressure tracheostomy cuff. Therefore, close observation and preventive measures are required to manage and avert TIF. Furthermore, NIV support should always be attempted before translaryngeal intubation.

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