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Case Report

Post-Tracheostomy Fistula Formation Between the Trachea and Left Common Carotid Artery in a Pediatric Patient with Trisomy 18

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The long-term survival of individuals with trisomy 18 has improved, and the need for tracheostomy in this population has thus increased. We describe the case of a 3-year-old Japanese boy with trisomy 18 who developed a tracheoarterial fistula involving the left common carotid artery (LCCA). He underwent an emergency tracheostomy for respiratory failure secondary to pneumonia. Massive hemorrhage from the tracheostoma occurred on postoperative day 31, leading to his death. The autopsy revealed that the LCCA originated from the brachiocephalic trunk and coursed medially, forming a fistulous connection with the trachea. This case highlights the clinical importance of preoperative vascular mapping in patients with chromosomal abnormalities.

Key words: tracheoarterial fistula, tracheocarotid fistula, trisomy 18

Trisomy 18 is a chromosomal abnormality associated with a poor prognosis; its 1-year survival rate is ~10%. However, the number of long-term survivors with trisomy 18 has been increasing due to individualized and active medical interventions, and in some individuals with this abnormality, a tracheostomy is performed during the chronic phase [1].

A tracheoarterial fistula (TAF) is a fatal complication associated with tracheostomy, and its onset is often difficult to predict. A common origin of the carotid arteries has been observed in approx. 10-25% of the general population, and this characteristic is more common among individuals with chromosomal abnormalities [2], including up to 52% of those with trisomy 18. Such anatomical variations may increase the risk of an atypical TAF originating from vessels other than the brachiocephalic trunk (BCT).

Although TAFs are rare with a 0.1-1.0% incidence, they are a severe complication that can result in fatal hemorrhage and thus require close attention, particularly in the early post-tracheostomy period [3]. Given the recent increase in tracheostomies performed in children with trisomy 18, a detailed preoperative anatomical evaluation, including the assessment of the patient's cervical vascular anatomy, may contribute to safer surgical planning and improved postoperative outcomes.

We report the case of a child with trisomy 18 who developed a TAF involving the left common carotid artery (LCCA), and we discuss the clinical significance of a preoperative vascular evaluation prior to a tracheostomy in children with chromosomal abnormalities.

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Case Presentation

The patient, a 3-year-old Japanese boy, was born at 34 weeks and 2 days' gestation, with a birth weight of 1,402 g. No prenatal diagnosis was made, and a postnatal chromosomal analysis confirmed trisomy 18 (47, XY, +18). The patient required mechanical ventilation from birth but was weaned on day 3 of life. Thereafter, respiratory support with a high-flow nasal cannula was required due to recurrent apnea. Tetralogy of Fallot was diagnosed and managed without surgical repair. At 2 years and 4 months of age, the patient developed epilepsy, for which antiepileptic drug therapy was initiated. At 2 years and 10 months of age, he was transitioned to home-based care while continuing tube feeding.

At 3 years and 3 months old, the patient was hospitalized at another facility for pneumonia. On hospital day 11, he developed acute respiratory failure. Because of micrognathia associated with trisomy 18, oral intubation was difficult, and an emergency tracheostomy was performed. On the same day, the patient was transferred to our hospital for intensive care. On admission, an endotracheal tube, rather than a tracheostomy cannula, had been inserted through the tracheostoma and was sutured to the skin to maintain the airway (Fig. 1A). Although the tracheostoma appeared somewhat distorted due to traction from the fixation sutures and was considered unstable, adequate ventilation was maintained. Despite the risk of endotracheal granulation caused by the use of an inappropriate tube, the endotracheal tube was left in place during the acute phase, as bedside tube exchange was judged to carry a high risk of loss of the airway access.

On postoperative day (POD) 8, granulation tissue obstructed the patient's tracheal lumen, resulting in a decreased tidal volume. A revision tracheostomy was performed to facilitate the safe exchange of the endotracheal tube for a tracheostomy cannula and create an appropriate tracheostoma. Since the tracheostoma was relatively large, no additional skin incision was required. Granulation tissue was partially excised to clearly expose the tracheal wall, and stay sutures were placed (Fig. 1B). After a tracheostomy cannula was inserted, the bilateral edges of the tracheostoma were partially sutured to reduce the stoma size and address the observed air leakage (Fig. 1C). No arterial pulsation was observed within the operative field. To preserve tracheal wall stability, peritracheal dissection was deliberately kept to a minimum.

The first cannula change was performed on POD 23 using bronchoscopy, which revealed resolution of the granulation tissue in the trachea. On POD 31, immediately after tracheal suction, a massive hemorrhage occurred from the cannula and the patient's mouth and nose, resulting in cardiac arrest due to asphyxiation and hemorrhagic shock. Cardiopulmonary resuscitation was unsuccessful, and the patient died 31min after the onset of bleeding.

An autopsy was performed after obtaining written

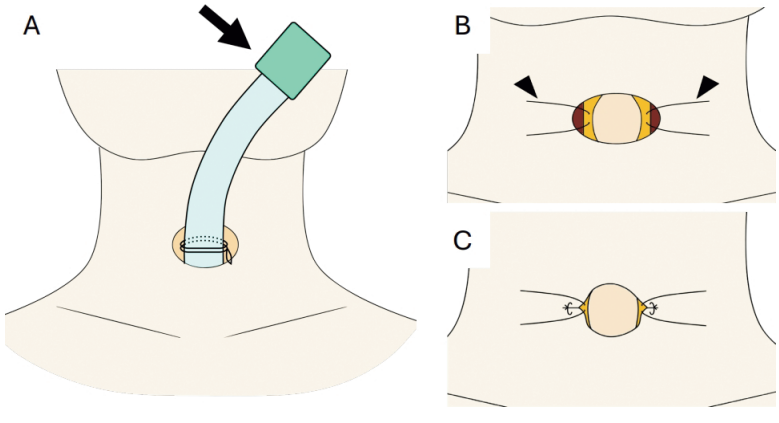

Fig. 1 **A**, Upon the patient's transfer to our hospital, an endotracheal tube (*arrow*) was inserted through the tracheostoma and secured with sutures. The tracheostoma was large, and air leakage was observed; **B**, During the revision procedure, the tracheal wall was exposed and stay sutures (*arrowheads*) were placed; **C**, The bilateral edges of the tracheostoma were partially sutured to reduce its size (the tracheostomy cannula is omitted in the figure).

informed consent from the patient's parents. The LCCA was found to originate from the BCT rather than directly from the aorta, representing a branching pattern known as a common origin of the carotid arteries. The course of the LCCA was more medial than is considered typical. The LCCA was located adjacent to the left side of the tracheostoma (Fig. 2), and there were two fistulous connections between the LCCA and the trachea (Fig. 3). In contrast, no fistula or granulation tissue was observed at the cannula tip, which is a typical site for a tracheoinnominate artery fistula (TIF; Fig. 4). Granulation tissue with neutrophilic infiltration and gram-positive cocci were observed adjacent to the LCCA, accompanied by inflammatory destruction and

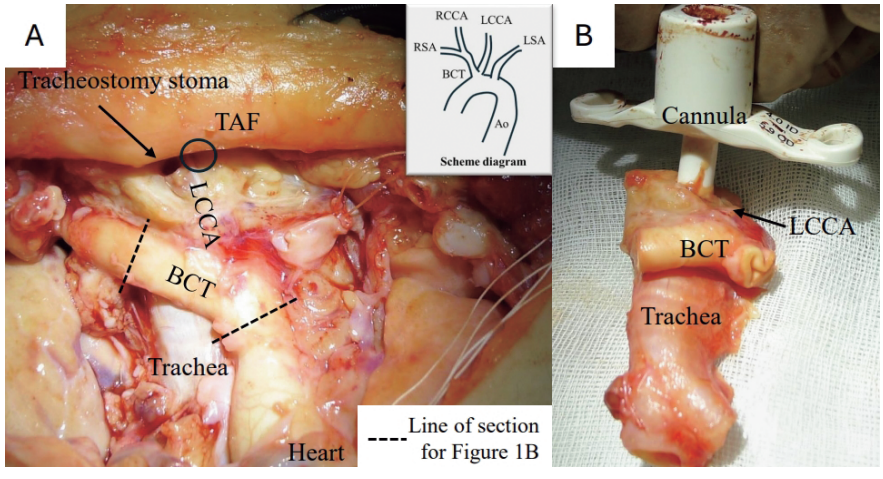

Fig. 2 Autopsy view of the patient's cervical region. **A**, The tracheoarterial fistula (TAF) was located superior to the brachiocephalic trunk (BCT) on the left side of the trachea. The left common carotid artery (LCCA) originated from the BCT; its course was more medial than is typical, forming a fistula with the trachea. The diagram shows the courses of the right subclavian artery (RSA), right common carotid artery (RCCA), left subclavian artery (LSA), and LCCA branching from the aorta (Ao) in the present case; **B**, The relative positions of the tracheal cannula, trachea, BCT, and LCCA.

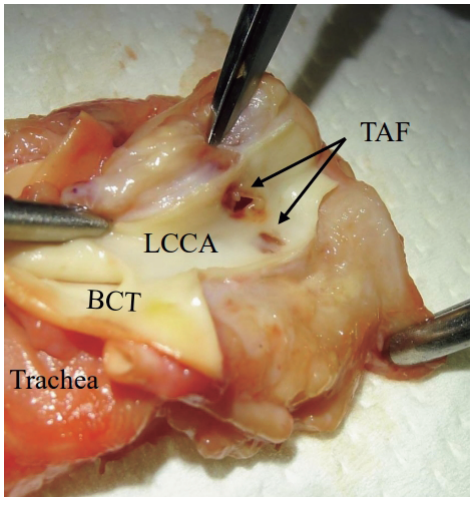

Fig. 3 Luminal view of the LCCA showing two fistulous openings to the trachea.

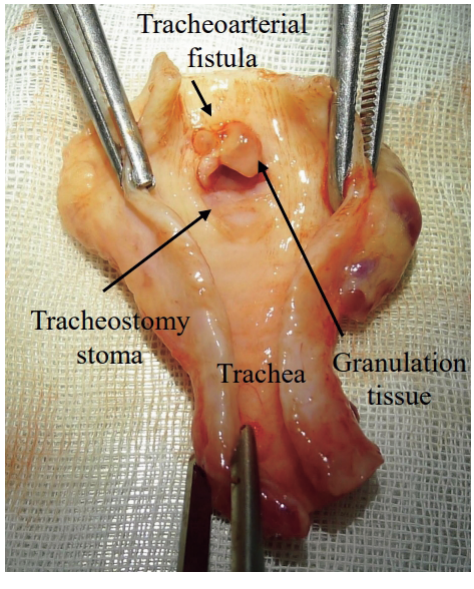

Fig. 4 Posterior luminal view of the trachea showing no fistula formation around the cannula tip, which is a typical site of fistula formation in tracheoinnominate artery fistula cases.

a loss of elastic fibers in the arterial wall. Based on these findings, the cause of the patient's death was deemed a fatal hemorrhage due to a trachea-LCCA fistula.

Discussion

Trisomy 18 is a chromosomal aneuploidy syndrome caused by the presence of a full or partial third copy of chromosome 18 [4]. Children with trisomy 18 have a poor prognosis, and aggressive treatment has traditionally been avoided. However, accumulating evidence has led to a shift in treatment strategies from conventional palliative care to individualized aggressive management, including surgical intervention [5]. Consequently, the number of long-term survivors with trisomy 18 has increased, which highlights the importance of comprehensive management during the chronic phase. In this context, central apnea often persists from the neonatal period and becomes a major management issue during the chronic phase, thereby increasing the need for a tracheostomy [1].

Among tracheostomy-related complications, TAF is rare with an incidence of 0.1-1.0%, but it is widely recognized as a fatal complication [6]. A TAF most commonly develops because of pressure necrosis between the trachea, BCT, and sternum. Reported risk factors for TAF include low-level tracheostomy, high cuff pressure, infection, malnutrition, steroid use, and excessive head movement, with multiple factors often contributing to its development [6]. In high-risk patients, prophylactic resection of the BCT has been reported as a preventive strategy [7]. Granulation tissue within the trachea is also clinically significant as it can amplify local infection and mechanical irritation.

In our patient's case, inflammatory injury to the arterial wall caused by local infection associated with granulation tissue is thought to have induced the degradation of elastic fibers, causing the vessel wall to be unable to withstand the intraluminal pressure and leading to the subsequent arterial rupture.

Anatomically, a TAF most commonly occurs as a TIF, involving the BCT. However, several cases of atypical fistulas between the trachea and other vessels, referred to as tracheocarotid fistulas (TCFs), have been reported [8-17] (Table 1). Among the eleven previously reported cases of TCF, ten originated from the right common carotid artery; the present case is the second report of a TCF involving the LCCA. Notably, at least three of the reported TCF cases were associated with a common origin of the carotid arteries. This type of variation could be a causative anatomical abnormality of atypical TAF.

A common origin of the carotid arteries — including variants in which the LCCA arises from the BCT or forms a bicarotid trunk with an anomalous right or left subclavian artery — was observed in approx. 10-25% of the general population in prior investigations, but it occurs at a significantly higher frequency among individuals with chromosomal abnormalities, including trisomy 18 (52%), trisomy 13 (64%), and trisomy 21 (42%) [2]. Given this background, clinicians should recognize that patients with chromosomal abnormalities may develop a TAF involving a vessel other than the BCT.

The accurate identification of fistula-forming vessels is critical because it directly influences the treatment strategy. Although prophylactic or emergency ligation is technically feasible in TIF cases, the interruption of carotid artery blood flow in the TCF carries an immediate risk of cerebral ischemia, necessitating surgical repair or endovascular treatment. A delayed or incorrect diagnosis may result in inappropriate management. In the reported TCF cases with survival, a definitive diagnosis was achieved by conducting a detailed vascular evaluation pre- or post-operatively, underscoring that the requirement of a precise understanding of vascular anatomy for achieving an accurate diagnosis and the optimal treatment selection.

An emergency tracheostomy was performed for our patient because of rapidly progressive respiratory failure, which made it difficult to evaluate the course of the carotid artery and anatomical variations preoperatively. If the course of the carotid artery had been recognized preoperatively, preventive measures might have been implemented, such as an adjustment of the length or insertion angle of the tracheostomy cannula or avoiding the compression of adjacent vascular structures. In addition, in certain high-risk patients, preventive surgical strategies — such as the interposition of soft tissue (*e.g.*, the sternohyoid muscle) between the trachea and adjacent vessels or a prophylactic repositioning of the common carotid artery—may also be considered to reduce the risk of a TAF. To prevent the fatal outcome of the present case, preoperative vascular mapping, including contrast-enhanced computed tomography (CECT), may be valuable for assessing the risk of the

Table 1 Previously reported cases of post-tracheostomy tracheocarotid fistula

Reports (year)	Age/Sex	Interval	Causative artery	COCA	Presumed mechanism	Treatment	Outcome
Dahill and Dempsey (2021) [8]	25/M	9 weeks	RCCA	N/A	Pressure necrosis	Saphenous vein graft	Survival
Hsu *et al.* (2016) [9]	60/M	9 months	RCCA	NO	N/A	Endovascular stent grafting	Survival
Discalzi *et al.* (2020) [10]	27/M	N/A	RCCA	YES	Pressure by cuff	Endovascular stent grafting	Survival
Hamano *et al.* (2008) [11]	14/M	11 years	RCCA	N/A	Granulation-associated infection	–	Death
Hamano *et al.* (2008) [11]	20/M	8 months	RCCA	N/A	Granulation-associated infection	–	Death
Schenken and Brown (1954) [12]	8/M	18 days	LCCA	N/A	Pressure necrosis and infection	–	Death
Scalise *et al.* (2005) [13]	48/F	142 days	RCCA	N/A	N/A	–	Death
Shylendran *et al.* (2014) [14]	64/F	2 weeks	RCCA	YES	Pressure by cuff	–	Death
Tungekar and Path (1999) [15]	62/F	3 years	RCCA	N/A	Infection	–	Death
Billy *et al.* (1994) [16]	39/M	2 years	RCCA	N/A	N/A	RCCA segmental resection	Survival
Steitz *et al.* (2015) [17]	25/F	8 months	RCCA	N/A	Infections in carotid stents	A right carotid to subclavian end-to-side anastomosis	Survival
Present Case	3/M	31 days	LCCA	YES	Granulation-associated infection	–	Death

M, male; F, Female; "Interval," date of tracheostomy and onset of the fistula bleeding; COCA, Common origin of the carotid arteries; RCCA, Right common carotid artery; LCCA, Left common carotid artery; N/A, Not available.

development of an atypical TAF in patients with chromosomal abnormalities. The optimal timing of CECT in patients with trisomy 18 has not been established. Our findings suggest that a vascular evaluation should be considered under stable clinical conditions, such as prior to discharge from the neonatal intensive care unit or before an elective tracheostomy.

Conclusion

We have described the case of a child with trisomy 18 who developed a TAF involving the LCCA. The autopsy revealed that the carotid arteries had a common origin, with the course of the LCCA being close to the tracheostoma. When planning a tracheostomy in a pediatric patient with a chromosomal abnormality, it is crucial to consider the possibility of anatomical variations in the origin of the carotid artery and to perform a preoperative evaluation of the carotid artery course. The present case highlights the importance of such an evaluation when implementing preventive strategies against TAF.

References

1. Fick TA and Sexson-Tejtel SK: Trisomy 18 trends over the last 20 years. J Pediatr (2021) 239: 206–211.
2. Wells TR, Landing BH and Shankle WR: Syndromal associations of common origin of the carotid arteries. Pediatr Pathol (1993) 13: 203–212.
3. Grant CA, Dempsey G, Harrison J and Jones T: Tracheo-innominate artery fistula after percutaneous tracheostomy: three case reports and a clinical review. Br J Anaesth (2006) 96: 127–131.
4. Cereda A and Carey JC: The trisomy 18 syndrome. Orphanet J Rare Dis (2012) 7: 81.
5. Kosho T, Nakamura T, Kawame H, Baba A, Tamura M and Fukushima Y: Neonatal management of trisomy 18: clinical details of 24 patients receiving intensive treatment. Am J Med Genet A (2006) 140: 934–944.
6. Chauhan JC, Hertzog JH, Viteri S and Slamon NB: Tracheoinnominate artery fistula formation in a child with long-term tracheostomy dependence. J Pediatr Intensive Care (2019) 8: 96–99.
7. Iodice F, Brancaccio G, Lauri A and Di Donato R: Preventive

ligation of the innominate artery in patients with neuromuscular disorders. Eur J Cardiothorac Surg (2007) 31: 747–749.
8. Dahill KE and Dempsey G: Tracheocarotid artery fistula in a patient who had tracheostomy successfully treated with a saphenous vein graft. BMJ Case Rep (2021) 14: e237854.
9. Hsu YC, Huang YC and Hsu CW: Tracheo-carotid artery fistula: an unusual cause of tracheostomy bleeding. QJM (2016) 109: 209–210.
10. Discalzi A, Fronda M, Gibello L and Ruffino MA: Endovascular treatment of tracheocarotid fistula. J Vasc Interv Radiol (2020) 31: 1083.
11. Hamano K, Kumada S, Hayashi M, Uchiyama A, Kurihara E, Tamagawa K, Enomoto S and Chou H: Hemorrhage due to tracheoarterial fistula with severe motor and intellectual disability. Pediatr Int (2008) 50: 337–340.
12. Schenken JR and Brown JM: Tracheocarotid fistula with fatal hemorrhage following tracheotomy for poliomyelitis; A case report. J Pediatr (1954) 45: 94–97.
13. Scalise P, Prunk SR, Healy D and Votto J: The incidence of tracheoarterial fistula in patients with chronic tracheostomy tubes: A retrospective study of 544 patients in long-term care facility. Chest (2005) 128: 3906–3909.
14. Shylendran S, Baliyan V, Yadav AK and Gamanagatti S: Post tracheostomy carotid-tracheal fistula. Indian J Otolaryngol Head Neck Surg (2014) 68: 97–99.
15. MF Tungekar and FRC Path: Tracheocarotid artery fistula infected with methicillin–resistant staphylococcus aureus. J Laryngol Otol (1999) 113: 689–691.
16. Billy ML, Snow NJ and Haug RH: Tracheocarotid fistula with life–threatening hemorrhage: report of case. J Oral Maxillofac Surg (1994) 52: 1331–1334.
17. Steitz JT, Cappello ZJ, Katrib Z and Tennant PA: Surgical Management of Recurrent Tracheocarotid Fistula following Endovascular Stent Placement. Case Rep Otolaryngol (2015) 2015: 547248.