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Case Report

Successful Endovascular Management of a Tracheo-Innominate Fistula in a Patient with Cantú Syndrome

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Abstract

Tracheo-innominate fistula is a rare complication in patients with tracheostomy, which is most commonly corrected surgically with a high mortality rate. We report the first case of a patient diagnosed with Cantú Syndrome who presented a tracheo-innominate fistula that was managed through the placement of an endovascular covered stent with good long-term results.

Keywords: *Cantu Syndrome; Hypertrichotic Osteochondrodysplasia; Tracheo-Innominate Artery Fistula; Endovascular Management*

Abbreviation

TIF: Tracheo-Innominate Artery Fistula

Introduction

Tracheo-innominate artery fistula represents a rare complication observed in patients with tracheostomy, with a variable incidence of 0.1 - 1% coupled with a high mortality rate in those without adequate intervention [1]. Hypertrichotic osteochondrodysplasia, an autosomal dominant disorder with a prevalence of < 1/1,000,000 is characterized by short stature, coarse facies, hypoplasia of the middle third of the face, upwardly inclined palpebral fissures, blepharophimosis, abundant eyebrows and eyelashes, brachymetacarpalia, brachymetatarsia and brachyphalangia [2]. There have been 150 confirmed cases of this entity reported worldwide. Herein, we report the first case of tracheo-innominate artery fistula of a patient with this distinct pathology that has been reported without any relationship with susceptibility to the formation of fistulas or important alterations in the vasculature at the cervical level, nor has any relationship been reported with the pathogenic variants characteristic of this disease such as ABCC9 or KCNJ8 [2,3].

The clinical manifestation of the tracheo-innominate artery fistula typically presents initially with an auto-limited hemorrhagic episode followed by substantial hemorrhage in majority of the cases approximately 14 days after tracheostomy. The subsequent hemorrhage can be managed via tracheostomy balloon inflation, stabilization of blood loss and subsequent surgical management; interventional management with endovascular stenting has gained relevance in recent years.

In this article, we present a case report of a patient diagnosed with Cantu syndrome tracheostomized that exhibited characteristic bleeding associated with a tracheo-innominate artery fistula, which underwent successful management with an endovascular stent in the innominate artery effectively resolving the hemorrhage.

Case Presentation

A 2-year-old male patient diagnosed with Cantu syndrome via genetic analysis has a medical history of severe pulmonary arterial hypertension at birth that required aggressive management (atrioseptostomy, sildenafil, bosentan, and iloprost); once resolved, subsequent closure of patent ductus arteriosus and interatrial communication was performed in separate occasions. Additionally, the patient currently has a tracheostomy that was performed two years ago with the last change of the cannula being a month ago and gastrostomy.

Two years after the placement of the tracheostomy, the patient presents to the emergency department with hemoptysis and active bleeding via the tracheostomy, associated with choanal bleeding. Although the patient was hemodynamically stable, hospital resuscitation protocol was activated and nebulized racemic adrenaline was administered resulting in reduced bleeding. Patient was discharged from the hospital with tranexamic acid and close monitoring. Four days later, the patient returns to the emergency room due to abundant tracheal secretions and intermittent and self-limited hemoptysis via the tracheostomy. A fluid resuscitation protocol was activated, and the patient was admitted to hospitalization for bronchoscopy study.

Bronchoscopy was performed reporting narrowing of more than 50% of the tracheal lumen, accompanied by discharge of fresh blood from tracheostomy cannula that led to the formation of a clot that completely obstructed the inferior respiratory tract. During aspiration of the clot, the patient started to desaturate and presented hemodynamic instability requiring aminergic support to halt the bleeding. Anterior nasal packing was performed and the patient was transferred to intensive therapy. An otorhinolaryngologist was brought to the case who related this episode to choanal bleeding. An angio-computed tomography of the supra-aortic vessels was performed identifying partial filling defect from the origin of brachiocephalic trunk to the origin of right common carotid artery, as well as loss of interface between adjacent tissues and tracheostomy cannula tip, without evidence suggestive of fistulous trajectories. Given the clinical suspicion of tracheo-innominate artery fistula, an emergency cardiac catheterization was performed for further evaluation.

A percutaneous procedure was undertaken via right femoral artery access using a 4 Fr introducer sheath and via left femoral venous access with 5 Fr introducer sheath. Arteriography via arterial access performed at the level of the brachiocephalic trunk and aortic arch revealed a unobstructed left aortic arch with expected origin and trajectory of the supra-aortic vessels, close proximity of the brachiocephalic trunk with tracheostomy cannula, without evidence of contrast leakage (Figure 1). Selective cannulation of emerging arterial branch from the brachiocephalic trunk demonstrated arterial circulation consistent with that of the thyroid gland. The Teflon-coated arterial exchange guide via arterial access was advanced to the right subclavian artery and through which a premade covered stent Gore Viabahn VBX 10 x 20 mm was deployed. Controlled insufflation was performed at 13 atm with complete stent expansion and slight anterograde displacement, devoid of deformation or rupture. The correct placement of the stent was confirmed by the antegrade flow and spatial relation with the tracheal cannula (Figure 2). Surveillance and prophylactic antibiotics were administered for 3 days post catheterization, and no complications reported at the 1 year follow-up. Upon the resolution and definite diagnosis of TIF, the angio-computed tomography was examined retrospectively observing presence of contrast medium in the airway tract, which suggests the presence of the fistula.

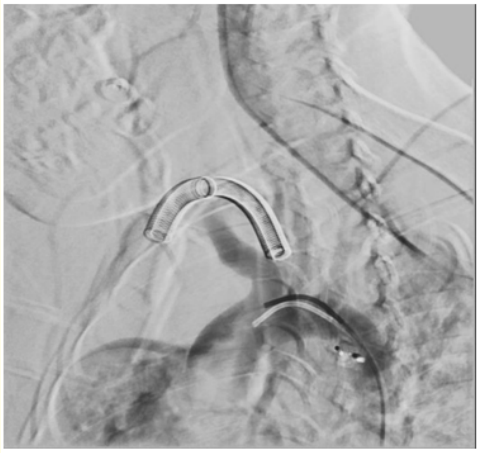

Figure 1: *Innominate artery adjacent to tracheostomy.*

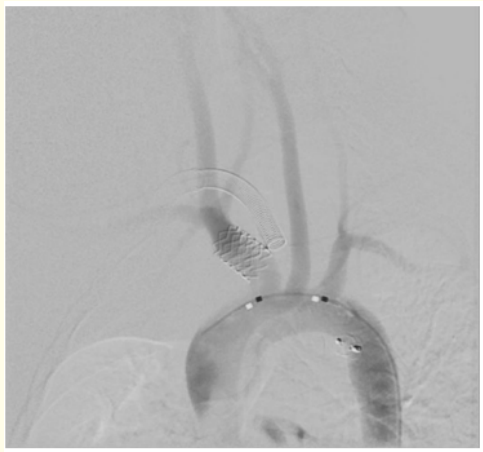

Figure 2: *Stent covering the site where tracheo-innominate artery fistula was bleeding.*

Discussion

Tracheo-innominate artery fistula is a complication reported in 0.3% of patients with tracheostomy that could potentially be fatal [1,4,5]. While the majority (70%) of these cases occur within the first 3 weeks post-tracheostomy, it can occur at any moment [4,5].

Typically, patients present with self-limited bleeding or sentinel bleeding in 35% of the cases and the rest 65% with significant hemorrhage via tracheostomy [6].

Multiple factors that damage the innominate artery contribute to the formation of the tracheo-innominate artery fistula although the exact pathophysiology is unknown. Certain factors predispose to its formation such as the tracheostomy itself, overinflation of the tracheostomy cannula, and overriding innominate artery [4].

Diagnosis is usually done through clinical presentation [7], such as this patient where no evidence was found through bronchoscopy nor angiotomography. However, clinical suspicion remains due to the presence of 1) sentinel bleeding followed by massive hemorrhage, 2) cessation of bleeding by digital compression, 3) homeostasis achieved after stent placement and 4) absence of other etiologies such as prior lesions in that site or coagulopathies [4,8].

The prognosis of patients depends upon timely diagnosis, adequate management of the hemorrhage and infection prevention measures [6]. Currently, studies that compare the two different treatment modalities for the tracheo-innominate artery fistula such as stent placement and surgical intervention do not exist. However, when not diagnosed or stabilized in time, left untreated leads to a mortality rate of 100% [5,7]. Even after surgical intervention, there still exists a high mortality rate among these patients. In a report of 70 patients that underwent surgical correction of the fistula, only 40 survived 2 months post-procedure [4]. Given the substantial risk of surgical intervention, endovascular management could be a promising alternative being less invasive with less operation time, reducing the risk of cerebral ischemia and minimizing the risks associated with dissecting in high complexity areas such as the supra-aortic trunks [7-9].

Conclusion

Tracheo-innominate artery fistula is a rare complication that could be potentially fatal if not diagnosed opportunely. Endovascular management coupled with antibiotic prophylaxis offers a promising alternative to conventional surgical intervention. This is the first successful case of endovascular management of a tracheo-innominate artery fistula in a patient with Cantú Syndrome. This case highlights the potential of endovascular management in treating this pathology.

Bibliography

1. Yogo Aoi., *et al*. "A case of tracheo-innominate artery fistula successfully treated with endovascular stent of the innominate artery". *Journal of General and Family Medicine* 18.4 (2017): 162-164.

2. Grange Dorothy K., *et al*. "Cantú Syndrome". PubMed, University of Washington, Seattle (1993).

3. Daas Falastine., *et al*. "Multiple vascular anomalies and refractory pericardial effusion in a young patient with Cantu syndrome: a case report and review of the literature". *BMC Pediatrics* 23.1 (2023): 644.

4. Allan James S and Cameron D Wright. "Tracheoinnominate fistula: Diagnosis and management". *Chest Surgery Clinics of North America* 13.2 (2003): 331-341.

5. Rodriguez Alonso A and M Garcia Fernandez de Villalta. "Otorrinolaringología En Pacientes Crónicos Complejos". Traqueotomía Pediátrica. Editorial Médica Panamericana (2021).

6. Deguchi J., *et al*. "Successful management of tracheo-innominate artery fistula with endovascular stent graft repair". *Journal of Vascular Surgery* 33.6 (2001): 1280-1282.

7. Javier Fernández Lorenzo., *et al*. "Endovascular treatment of tracheo-innominate fistula". *Angiología* 73.3 (2021): 151-154.

8. Machado AT and Ribeiro Dias Barroso MC. "Tratamento Endovascular de Fístula Tronco Arterial Braquiocefálico-Traqueal: Relato de Caso E Revisão de Literatura". *Jornal Vascular Brasileiro* 17.4 (2018): 348-352.

9. Kakimoto Takayuki., *et al*. "A case of tracheo-innominate artery fistula after tracheostomy successfully treated with a covered stent". *NMC Case Report Journal* 10 (2023): 21-25.

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