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CLINICAL CASE

Double aortic arch and aortotracheal fistula as cause of fatal massive bleeding: a rare and dangerous association

Doble arco aórtico y fistula aortotraqueal como causa de hemorragia masiva fatal: una asociación rara y peligrosa

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What do we know about the subject matter of this study??

Double aortic arch is a rare congenital malformation whose diagnosis is usually challenging. It is associated with tracheal and esophageal compression where prolonged use of endotracheal and esophageal tubes favors the development of conditions that can be fatal such as aortotracheal or aortoesophageal fistulas.

What does this study contribute to what is already known??

We present a case report associating double aortic arch and aortotracheal fistula in a premature newborn. This report reinforces the need for a high index of suspicion in neonates with lower airway obstruction secondary to massive aerodigestive tract bleeding without focus, in order to achieve diagnosis and treatment, given the poor prognosis of this entity.

Abstract

Vascular rings are unusual congenital malformations. Among them, double aortic arch (DAA) is often difficult to diagnose due to its low incidence of symptoms. DAA can be associated with tracheal or esophageal compression and, in severe cases, could require tracheal intubation or chronic use of a nasogastric tube. This scenario favors the development of aortotracheal fistulas (ATF) or aortoesophageal fistulas (AEF). **Objective:** To present a clinical case with an unusual association of DAA with ATF and to reinforce the importance of maintaining high diagnostic suspicion in patients with massive aerodigestive bleeding without an obvious source. **Clinical Case:** A 32-week preterm newborn who required prolonged mechanical ventilation and presented intermittent episodes of massive oropharyngeal bleeding with hemodynamic compromise associated with lower airway obstruction without pulmonary hemorrhage. The patient underwent upper endoscopy and exploratory laparotomy without evidence of bleeding. Flexible nasopharyngolaryngoscopy and direct laryngoscopy also showed no abnormalities. A CT angiography showed complete DAA with indentation of the left dominant arch over the trachea, without severe stenosis or evidence of a fistula. AEF was suspected,

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so exploratory surgery was considered. However, the patient died before surgery due to a massive pulmonary hemorrhage. The autopsy revealed the presence of ATF. **Conclusions:** In patients with massive aerodigestive bleeding without an obvious source, the presence of DAA and possible AEF/ATF should be considered. Imaging studies have a poor performance for this diagnosis, so surgery should be considered for diagnosis and treatment in these patients.

Introduction

Vascular rings are rare and account for approximately 1% of all congenital cardiovascular malformations[1]. Among them, double aortic arch (DAA) can more frequently present as tracheal and/or esophageal compression[1]. Due to the low incidence of symptomatic cases, diagnosis can be delayed, omitted, or incidental[1,2]. Given the above, DAA can cause compression of the trachea and esophagus, often associated with tracheomalacia[2,3]. More severe cases require prolonged mechanical ventilation (MV), often associated with multiple endotracheal intubations and nasogastric feeding[3,4]. Long-term use of endotracheal and/or nasogastric tubes can favor the development of aortotracheal fistula (ATF) or aortoesophageal fistula (AEF), both associated with high mortality[5].

The diagnosis of AEF/ATF is difficult and carries a poor prognosis and may not be considered in cases of hematemesis or hemoptysis and undiagnosed DAA[2]. In the literature, there is a strong association between DAA and AEF[1-6].

The objective of this report is to present a case with an unusual association of DAA with ATF and to reinforce the importance of maintaining a high diagnostic suspicion in patients with massive aerodigestive tract bleeding without an evident focus.

Clinical Case

Preterm newborn of 32 weeks of gestational age, born from a twin pregnancy induced due to intrauterine growth restriction and altered fetal Doppler ultrasound in this twin. She presented with neonatal respiratory distress syndrome, required MV, and developed bilateral intracranial hemorrhage. Enteral feeding was initiated through an orogastric tube.

Weaning from MV was difficult, with three extubation failures associated with episodes of lower airway obstruction (LAO). A flexible nasopharyngoscopy was performed, which showed pharyngeal hypotonia and mild laryngomalacia without other pathological findings.

At 30 days of life, the patient presented recurrent and massive episodes of spontaneous hematemesis with acute hemodynamic compromise requiring multiple saline boluses and blood transfusions. Upper gastrointestinal bleeding was suspected, so she was transferred to our institution for an upper endoscopy at 40 days of life.

Upper endoscopy showed blood clots in the gastric fundus without underlying lesions. Due to the absence of an obvious source of bleeding, an exploratory laparotomy was performed, which showed no lesions in the stomach or distal esophagus.

The patient remained on MV with intermittent episodes of massive oropharyngeal hemorrhage and severe hemodynamic compromise, associated with LAO, with no signs of pulmonary hemorrhage.

A new nasopharyngoscopy followed by a rigid endoscopy and direct laryngoscopy showed no anatomical abnormalities that could explain the massive bleeding. Nasal and oropharyngeal packing was placed, reducing bleeding episodes for 48 hours.

A CT angiography showed a complete DAA with indentation from the dominant left arch in the posterior wall of the intrathoracic trachea, without significant stenosis or evidence of fistula (Figure 1). Given these findings, an AEF associated with DAA was considered as the cause of bleeding. Based on this, surgical thoracic exploration was planned; however, before surgery, the patient died due to a massive pulmonary hemorrhage.

Due to the characteristics of the case, an autopsy was requested. The pathological examination confirmed the presence of a DAA. The posterior arch was in close contact with a segment of the trachea, with a 2 mm ATF at that level (Figure 2).

The cause of death was attributed to a hemorrhage of the lower respiratory tract secondary to an ATF. There was no evidence of AEF or other sources of bleeding. Figure 3 depicts the clinical reasoning and steps followed throughout the case.

Discussion

The diagnosis of AEF/ATF is difficult and carries a poor prognosis and may not be considered in cases of hematemesis or hemoptysis and undiagnosed DAA (2). Massive oropharyngeal hemorrhage episodes without

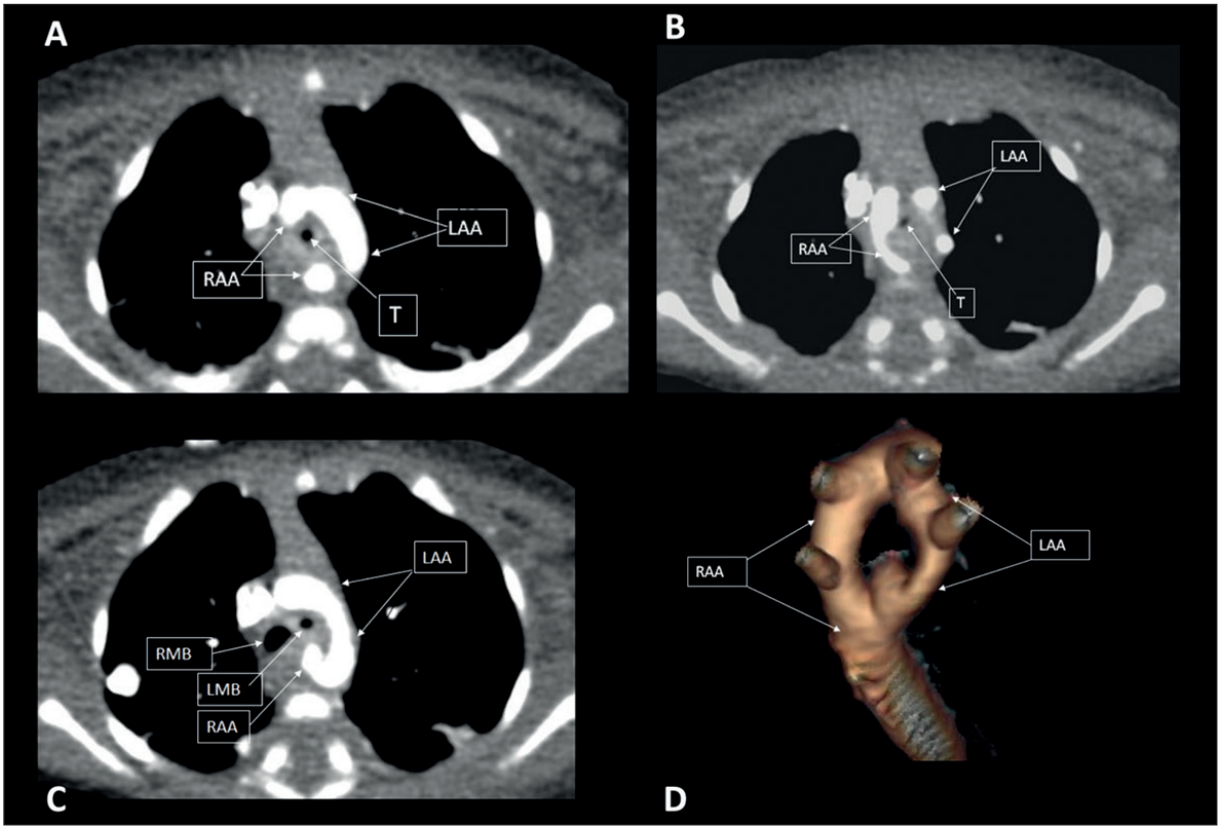

Figure 1. Axial CT-Angiography of the patient (**A, B, C**); 3-D reconstruction (**D**). Dominant LAA compression and narrowing of T and LMB, without fistula. RAA: right aortic arch; LAA: left aortic arch; T: Trachea; RMB: right main bronchus; LMB: left main bronchus.

a clear source or cause can confuse the medical team and delay the diagnosis of DAA and AEF/ATF[3]. A CT angiography or MRI are the imaging studies of choice, both with high sensitivity and specificity in diagnosing vascular rings but may not detect an AEF or ATF[4]. This is mainly because bleeding may be intermittent and not observed at the time of imaging. Other vascular rings such as the right aortic arch associated with aberrant left subclavian artery can also produce tracheal and esophageal compression but have not been related to AEF/ATF, except in cases associated with esophageal foreign bodies (e.g., batteries/coins)[6]. Since imaging studies and complementary tests to diagnose AEF/ATF are often futile, early open- or endovascular surgery should be considered once the diagnosis of DAA is confirmed in these patients, as diagnosis and treatment[3].

Clinical suspicion, active search, and early diagnosis are crucial for timely diagnosis and treatment of this condition with a poor prognosis[2,3]. In the case presented, the diagnosis was further complicated, as it was a premature newborn with complicated DAA caused by an ATF, in contrast to cases previously documented in the literature, which generally correspond to AEF in full-term or older newborns[2,5,7-11]. The presence of ATF in this case was associated with a worse prognosis.

Table 1 presents a review of pediatric cases reported in the literature with DAA associated with ATF/AEF not related to foreign bodies in the pharynx/esophagus (except for nasogastric, orogastric, and endotracheal tube). Upon analyzing the cases presented in the literature, it is noteworthy that the presence of DAA along with chronic invasion such as nasoenteral or endotracheal tube has a higher risk of presenting ATF/AEF, as well as patients who undergo surgical intervention.[2,3,5-16].

Massive oropharyngeal hemorrhages, as described in this report, should lead to suspicion of possible vascular malformations such as DAA and underlying ATF/AEF.

In this case, other complications of prematurity could have contributed to the difficulty of diagnosing DAA. Therefore, a high index of clinical suspicion and early treatment are relevant, especially in the presence of episodes of lower airway obstruction suggestive of external compression of the airway. Al-

though an ATF was confirmed, we cannot rule out the possibility of a transient AEF, considering the presence of massive oropharyngeal hemorrhage without evidence of bleeding from the endotracheal tube and the high prevalence of AEF associated with DAA among published cases[4].

In conclusion, we recommend a high index of suspicion for the presence of vascular rings and possible ATF/AEF in patients presenting with episodes of lower airway obstruction with hemorrhage from the aerodigestive tract without an evident focus. Along with this and given the low performance of imaging studies in the diagnosis of AEF/ATF, an early surgical approach should be considered as diagnosis and treatment[4].

Ethical Responsibilities

Human Beings and animals protection: Disclosure the authors state that the procedures were followed according to the Declaration of Helsinki and the World Medical Association regarding human experimentation developed for the medical community.

Data confidentiality: The authors state that they have followed the protocols of their Center and Local regulations on the publication of patient data.

Rights to privacy and informed consent: The authors have obtained the informed consent of the patients and/or subjects referred to in the article. This document is in the possession of the correspondence author.

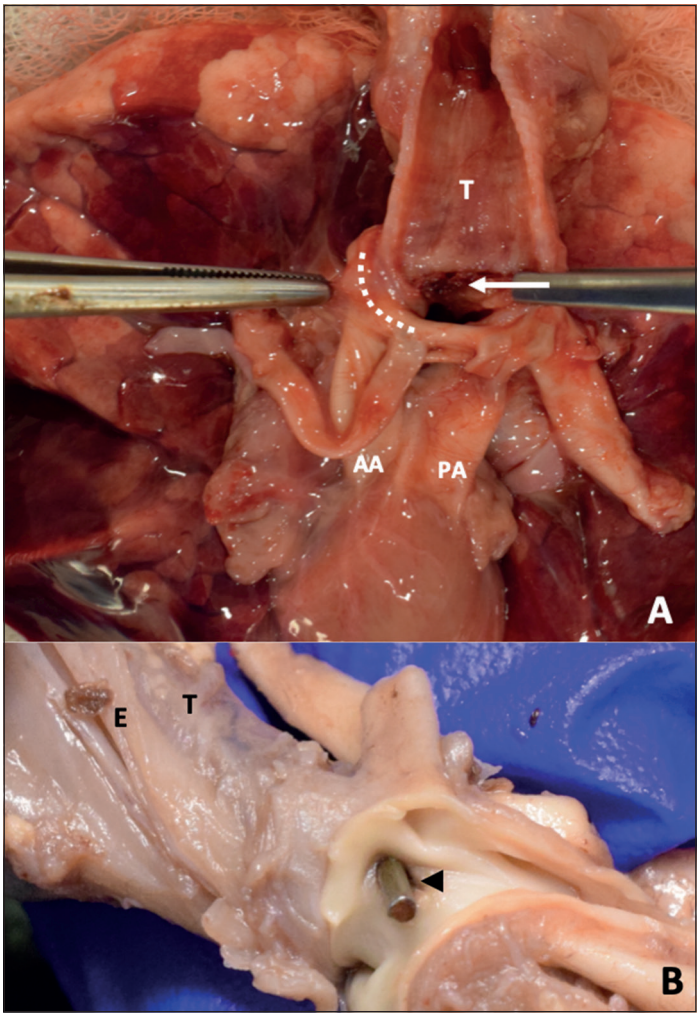

Figure 2. (**A**) Vascular ring (dashed line) surrounds the trachea with an ATF at the posterior tracheal wall (arrow). (**B**) ATF communicating tracheal lumen with the VR (arrowhead). AA: Ascending aorta; PA: Pulmonary Artery; T: Trachea; E: Esophagus

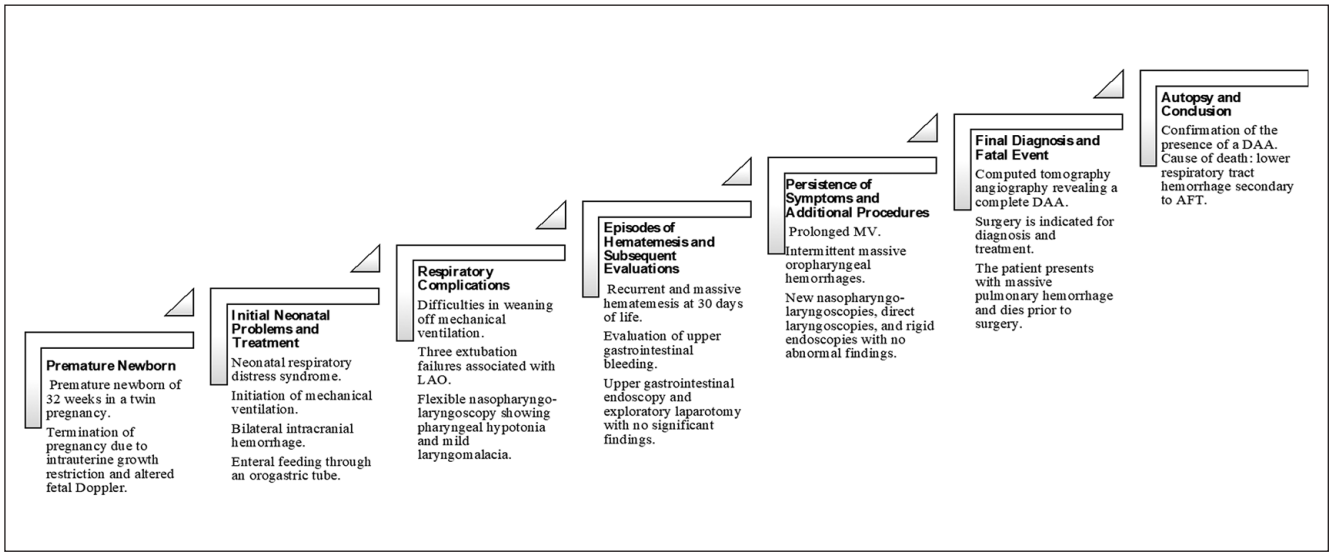

Figure 3. Flowchart of clinical reasoning in the management of the case. LAO: lower airway obstruction. DAA: double aortic arch; ATF: aortotracheal fistula.

Table 1. Double aortic arch associated to aortoesophageal and/or aortotracheal fistula, literature case report summary.

Study	Year	Population	Language	Nº	Age	Results
Arciniegas et al.[7]	1979	AEF in a child with DAA re-cently repaired, secondary to prolonged use of nasogastric tube and MV.	Inglés	1	4 months	Prolonged use of me-chanical ventilation (MV) and nasogastric tube prior to surgical repair of DAA. On postoperative day 9, re-intervention due to parae-sophageal ab-scess and aortic su-ture dehiscence. On postoperative day 19, massive hemorrhage due to AEF. Man-aged with esophageal defunctionali-zation and aortic suture re-pair. Transit reconsti-tution 1 year later without incidents.
McKeating J et al.[8]	1990	AEF in a child with DAA secondary to nasogastric tube.	Inglés	1	3 months	Right thoracotomy showed a right aortic arch that had eroded into the right posterolateral wall of the upper thoracic esoph-agus. Patient suffered cardiac arrest and died.
Yahagi N et al.[12]	1992	Tetralogy of Fal-lot, DAA and AEF.	Inglés	1	4 day of life (premature 32-week patient)	DAA and AEF surgery was uneventful.
Heck Ha Jr[5]	1993	AEF in two pa-tients with undiag-nosed DAA, asso-ciated to pro-lon-ged MV and nasogastric tube.	Inglés	2	6 weeks old 3 weeks old	Patient 1: Upper airway compromise associated with prolonged MV and difficulty in weaning. DAA surgical repair, de-veloped bloody stools and emergency surgery exploration showed AEF. Unsuccessful attempt to repair, the patient died of exsanguination. Patient 2: Intermittent apneic episodes. DAA surgi-cal repair showed contained posterior esophageal perforation that was repaired. Une-ventful posto-perative course.
Mizushima A et al.[11]	1995	Tracheomalacia and fatal AEF due to unrecog-nized DAA, associated to prolonged MV and nasogastric tube.	Japonés	1	3 months	Prolonged MV and naso-gastric tube after cardiac surgery, developed fatal upper gastrointestinal hemorrhage. Postmortem examination showed an AEF.
Biemann Othersen H et al.[13]	1996	AEF in two pa-tients with DAA	Inglés	2	2 days old 2 months	Patient 1: DAA surgical repair. Developed respira-tory distress and massive gastrointestinal bleeding. Emergency thoracotomy for AEF repair. Successful outcome. Patient 2: DAA diagnosis, prolonged MV and naso-gastric tube. Developed massive hematemesis. DAA and AEF repair was uneventful.
Hill JG et al.[14]	1999	AEF in two pa-tients with DAA	Inglés	2	5 weeks old 2 months	Patient 1: Initial DAA surgical repair. Prolonged MV and nasogastric tube, 9th day after surgery, the patient developed respira-tory distress and hema-temesis. Emergency sur-gery showed an AEF that was repaired. Uneventful postoperative course. Patient 2: Intermittent apnea and massive he-matemesis. DAA was diagnosed. Surgical DAA and AEF repair. Unevent-ful postoperative course.
Angelini A et al.[2]	2002	AEF in two pa-tients with DAA secondary to na-sogastric tube	Inglés	2	3 months 3 months	Patient 1: Prolonged MV and difficulty in weaning. After replacement of nasogastric tube present-ed fatal profuse hemor-rhage from mouth and nose. Patient 2: Prolonged MV and difficulty in weaning. After nasogastric tube placement presented with profuse hematemesis with no identified source, de-veloped disseminated intravascular coagulation and died.

Chaikitpinyo A et al.[10]	2004	AEF with DAA after prolonged MV and nasogas-tric tube	Inglés	1	2 months	Emergency thoracotomy with DAA and AEF repair. Uneventful postoperative course.
Van Woerkum et al.[15]	2006	AEF with DAA and prolonged nasogastric tube.	Inglés	1	9 weeks old	Surgical repair of DAA. Post operatively devel-oped an AEF due to na-sogastric intubation. Na-sogastric intubation was discontinued, feeding gastrostomy was provid-ed. No surgical interven-tion was per-formed with no recurrent bleeding.
D'Angelis et al.[16]	2006	AEF in a child with DAA secondary to DAA surgi-cal re-pair	Inglés	1	3 months	DAA surgical repair was uneventful. 4 weeks later the patient developed respiratory distress and massive hematemesis. An AEF was suspected. Emer-gency thoracotomy was performed, an AEF was confirmed and repaired. The surgery was success-ful, and the recovery was uneventful and without bleeding.
Atsumi et al.[3]	2015	AEF fistula with DAA and pro-longed MV. Pa-tient develops an ATF fistula after AEF surgical re-pair.	Inglés	1	2 months	Left thoracotomy for DAA and AEF repair. The surgery was successful with no residual bleeding. Five hours after the oper-ation, massive bleeding from the endotraqueal tube. Surgical exploration showed ATF that was repaired. The patient died from hypoxia 6 hrs after surgery.
Hoshina T et al.[9]	2015	AEF with undiagnosed DAA	Japonés	1	7 weeks old	Severe dyspnea, pro-longed MV and use of naso-gastric tube. Devel-oped massive hemateme-sis and hemorrhagic shock. Surgical DAA and AEF repair with unevent-ful postoperative course.

AEF: Aortoesophageal fistula; DAA: Double aortic arch; MV: Mechanical ventilation; ATF: Aortotracheal fistula.

Conflicts of Interest

Authors declare no conflict of interest regarding the present study.

Financial Disclosure

Authors state that no economic support has been associated with the present study. Epta sit, ne niet ut ut duciiscim quam voluptatqui accus dem sum nobis eatis int adis aut everum etum quid es pa demporem as is sam, omnimpos eosae excerumqui temperibus, to tem num ipsum nos magnatquatem quidiciae alis reperib eribusapicil ilitibus moluptat volluptatur, et faciet vid unturep ellenim fuga. Ut voloribus iuste repel eat earciis dolorrovid milis et et eosserum facestia se od et doluptati veliquamusam delest faciis pliqui in consequos dolorecti cullaborum et, veliam, atistes reicaep udicimodis rest, susam dist fuga. Ut porenih illupti idendandae rerum sequid modia volupic ipidelent laborei ciistium reperiti omnissimaio. Et volut ut quo iusam, omnimetur, omnihita dunt aritatiis ni deles rernam aliti nost, sequam exerum, istrum reremquas ratium eosame nosti rerum eates quam exceat.

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