Respiratory Medicine Case Reports 44 (2023) 101866

Contents lists available at ScienceDirect

Respiratory Medicine Case Reports

journal homepage: www.elsevier.com/locate/rmcr

Case Report

Airway obstruction caused by achalasia: A case report☆

Tokuo Fujisawa [a], Junji Hatakeyama [a, b, *], Kenichiro Omoto [a, c]

[a] Department of Emergency and Critical Care Medicine, National Hospital Organization Tokyo Medical Center, Japan
[b] Department of Emergency and Critical Care Medicine, Osaka Medical and Pharmaceutical University, Japan
[c] Department of Emergency and Critical Care Medicine, Nippon Medical School Tama Nagayama Hospital, Japan

ARTICLE INFO

Handling Editor: DR AC Amit Chopra

Keywords:
Airway obstruction
Esophageal achalasia
Noninvasive ventilation
Stridor
Tracheomalacia

ABSTRACT

We report a rare case of airway obstruction caused by megaesophagus associated with achalasia. A 78-year-old man was admitted with post meal dyspnea, decreased consciousness, expiratory and inspiratory wheezing, and respiratory distress. Arterial blood gas analysis showed findings of marked acute respiratory acidosis (pH 7.18, $PaCO_2$ 75 mmHg, PaO_2 225 mm Hg, HCO_3^- 22 mmol/L). An emergency laryngoscopy was performed because of a suspected airway obstruction, but no abnormalities were observed from the airway to the glottis. Noninvasive positive pressure ventilation (NPPV) was immediately introduced, and the respiratory rate and breathing pattern was normalized. A chest X-ray showed an enlarged upper mediastinal outline and an ill-defined border of the trachea. A computed tomography (CT) scan showed an enlarged esophagus with a maximum diameter of 9.90 cm, compressing the trachea to the back of the sternal notch. Following removal of the esophageal contents using a nasogastric tube, NPPV was discontinued with no respiratory episodes. After he was stabilized, he was transferred to another hospital for endoscopic myotomy. In a review of the literature, we identified 66 cases of airway obstruction due to achalasia, mainly in older women. None of the patients received NPPV. As a differential diagnosis for acute airway obstruction, achalasia-related airway obstruction should be considered, particularly in older women. Furthermore, since this condition is suspected to involve tracheomalacia, NPPV may be a useful respiratory support therapy.

Author contributions

Tokuo Fujisawa: Writing – original draft Junji Hatakeyama: Writing – review & editing Kenichiro Omoto: Writing – review & editing.

1. Introduction

Esophageal achalasia is a chronic, progressive, functional disease resulting from inadequate relaxation of the lower esophageal sphincter, and impaired peristalsis of the esophageal body, leading to impaired transit [1]. We encountered a case of airway obstruction due to tracheal compression associated with upper esophageal dilatation. This study also reviews previous reports of similar cases.

☆ Institution where the work was performed: National Hospital Organization Tokyo Medical Center, 1-5-2, Higashigaoka, Meguro-ku, Tokyo 152–0021, Japan.

* Corresponding author. Department of Emergency and Critical Care Medicine, Osaka Medical and Pharmaceutical University 2-7 Daigaku-machi, Takatsuki, Osaka, 569-8686, Japan.
E-mail address: junji.hatakeyama@ompu.ac.jp (J. Hatakeyama).

https://doi.org/10.1016/j.rmcr.2023.101866
Received 24 November 2022; Received in revised form 5 May 2023; Accepted 10 May 2023
Available online 11 May 2023

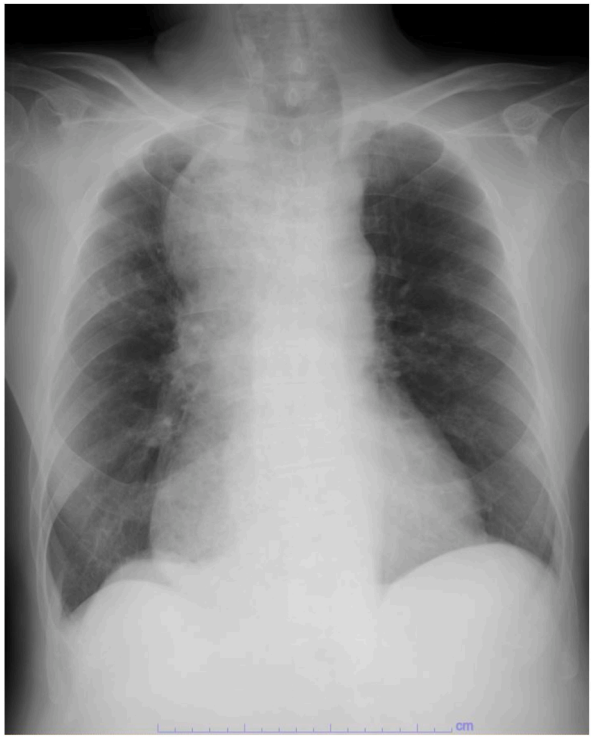

Fig. 1. Chest X-ray film.
Enlarged upper mediastinal outline and ill-defined border of the trachea was seen.

2. Case presentation

A 78-year-old man (height 160 cm; weight 60 kg) was admitted to our hospital with a complaint of dyspnea that suddenly started after dinner. He had a history of esophageal achalasia, hypertension, dyslipidemia, benign prostatic hyperplasia, and emphysema. Esophageal achalasia was diagnosed 11 years earlier but was left untreated. On arrival to the hospital, the patient's level of consciousness was E4V5M6 on the Glasgow coma scale; pulse, 140 bpm; blood pressure, 172/108 mmHg; respiratory rate, 30 bpm; and labored respiratory effort were recorded. On auscultation, a stridor was audible from the neck to the anterior chest, and respiratory sounds were attenuated in both lung fields. Additionally, wheezing and rhonchi were audible in the bilateral superior lung fields. SpO_2 was 100% with a 10 L/min reservoir mask. Arterial blood gas analysis showed marked acute respiratory acidosis (pH 7.18, $PaCO_2$ 75 mmHg, PaO_2 225 mmHg, HCO_3^- 22 mmol/L), and blood tests revealed no significant abnormalities. Chest radiography showed no obvious abnormalities in the lung fields but marked enlargement of the upper mediastinum and opacification of the tracheal shadow (Fig. 1). Emergency laryngoscopy revealed no obvious abnormalities in the airway up to the glottis. Therefore, we introduced noninvasive positive pressure ventilation (NPPV) on suspicion of lower respiratory tract and pulmonary diseases. The patient's respiratory rate improved rapidly to 20 bpm and respiratory effort and wheezing disappeared. The computed tomography (CT) scan showed the dilation of esophagus to a maximum diameter of 9.90 cm and the trachea flattened and nearly obstructed by compression at the thoracic inlet level. There were slight emphysematous changes in the lung fields but no sign of aspiration and no anatomical abnormalities in ribs and spines leading to hypoventilation (Fig. 2). Additionally, there were no neurological abnormalities suggestive of neuromuscular diseases. Therefore, airway obstruction due to esophageal achalasia was diagnosed. After admission to the intensive care unit, the esophageal contents were removed through a nasogastric tube. Arterial blood gas analysis rechecked 7 h later showed improvement to pH 7.38, $PaCO_2$ 49 mmHg, PaO_2 88 mmHg, HCO_3^- 28 mmol/L (continuous positive airway pressure: 4.0 cmH_2O, FiO_2: 0.25) without any treatment for chronic obstructive pulmonary disease and NPPV was discontinued. After the respiratory status stabilized, he was transferred to another hospital for endoscopic myotomy.

3. Discussion

Esophageal achalasia occurs in approximately 1 in 100,000 people annually [1] regardless of sex, with primary symptoms of dysphagia, heartburn, esophageal reflux, vomiting, and noncardiogenic chest pain. Respiratory symptoms can include coughing, asthma symptoms, chronic aspiration, hoarseness, and sore throat. We found only 66 adult case reports of airway obstruction due to dilated esophagus since 1950 (Table 1). A review of these cases revealed that 85.1% of the patients were women with a mean age of 71.3 years for women and 44.8 years for men, indicating that airway obstruction is more common in older women. Acute dyspnea and stridor were the most common symptoms, occurring in approximately 90% and 70% of the cases, respectively, with other symptoms in-

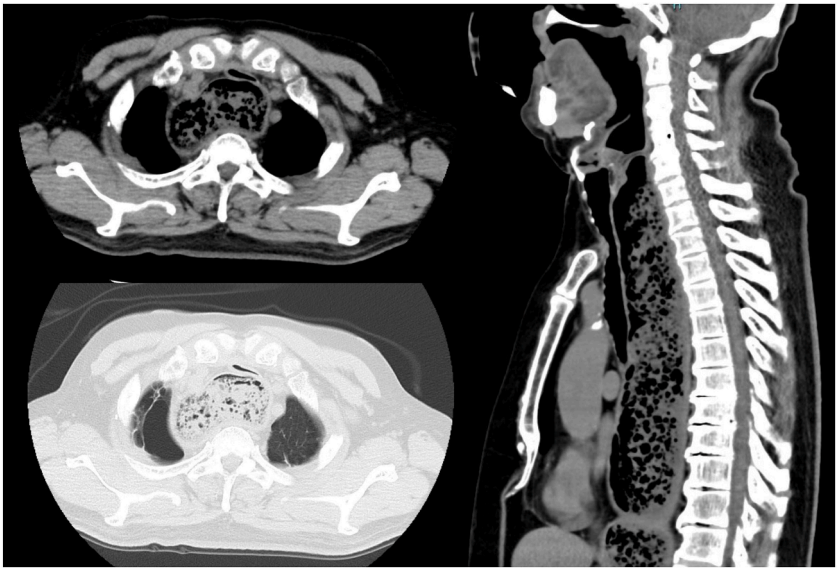

Fig. 2. Chest computed tomography (CT) scan.
Axial plane at the level of the sternoclavicular joint (left) and sagittal plane (right). A flattened and occluded trachea caused due to dilated esophagus was observed.

cluding dysphagia, neck swelling, cyanosis, and wheezing. Approximately 44% of the patients underwent tracheal intubation or tracheostomy, and three patients experienced cardiac arrest, indicating the urgent need for diagnosis and treatment.

Several hypotheses have been proposed for the pathogenesis of megaesophagus caused by achalasia [2]. Esophagus dilation is thought to occur due to a combination of factors, including one-way valve formation by the twisted esophagus behind the cricopharyngeal muscle leading to trapping of air, inadequate relaxation of the upper esophageal sphincter [3,4], and difficulty in air expulsion due to loss of the burp reflex [5]. Moreover, the pathogenesis of airway obstruction is reported to involve tracheomalacia [2], which has been confirmed by bronchoscopy and CT in several cases [6,7]. In tracheomalacia, due to the fragility of the trachea, negative pressure in the airway causes collapse of the trachea outside the thoracic cavity during inspiration, and positive pressure in the thoracic cavity causes collapse of the trachea inside the thoracic cavity during expiration [8], which is consistent with the physical findings in our case. Therefore, it is theoretically effective to maintain positive pressure in the airways to prevent tracheal collapse. This case demonstrated its effectiveness, although there have been no case reports on using NPPV. If the risk of vomiting and aspiration is considered and indications are carefully examined, NPPV may be a useful treatment for airway obstruction due to achalasia, as it leads to the avoidance of highly invasive airway interventions, such as tracheal intubation and tracheostomy.

4. Conclusions

We encountered a rare case of airway obstruction associated with achalasia. A literature review revealed that this phenomenon is more common in older women. NPPV may be effective for these cases because its pathogenesis involves tracheomalacia.

Funding

The authors did not receive any specific grant from funding agencies in the public, commercial, or not-for-profit sectors.

Informed consent

Written informed consent was obtained from the patient for the publication of this case report and the accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal upon request.

Declaration of competing interest

All authors of this manuscript have no conflicts of interest, and have no commercial, financial or other relationships related to the subject of this article, according to the ICMJE conflict of interest guidelines.

Table 1
Reported cases of airway obstruction caused by a dilated esophagus in patients with achalasia.

Author, year	Age	Sex	Presenting symptoms	Artificial Airway	Definitive treatment	Achalasia previously diagnosed
Natesh et al., 2013	94	F	acute dyspnea, stridor, neck swelling, hoarseness, dysphagia, chest pain	tracheal intubation, tracheostomy	not reported	no
Medici et al., 2013	93	F	acute dyspnea, stridor, cough, wheeze		not reported	no
Hay et al., 2000	90	F	acute dyspnea, stridor, dysphagia, retrosternal discomfort		not reported	no
Westbrook et al., 1992	90	F	acute dyspnea, cyanosis, neck swelling		not reported	no
Arcos et al., 2000	89	F	acute dyspnea, stridor, dysphagia, cyanosis, weight loss	tracheal intubation	botulinum toxin	no
Hoshino et al., 2020	88	F	acute dyspnea, stridor, neck swelling, cyanosis		pneumatic dilation	yes, 8 years earlier
Tomlinson et al., 2016	88	F	acute dyspnea, stridor, cough, sputum	tracheal intubation	not reported	no
Kurimoto et al., 2014	88	F	acute dyspnea, stridor, cough, nausea, vomiting	tracheal intubation	pneumatic dilation	no
Dunlop et al., 1997	87	F	acute dyspnea, stridor, cough, cyanosis, distention of jugular veins, neck swelling		pneumatic dilation	no
Ali et al., 1995	87	F	acute dyspnea, stridor, dysphagia, neck swelling	tracheal intubation	myotomy	yes, 18 years earlier
Evans et al., 1982	87	F	acute dyspnea, stridor		pneumatic dilation	no
Wichramasinghe et al., 1988	85	F	acute dyspnea, hoarseness, stridor, cyanosis, dysphagia, weight loss, regurgitation	tracheostomy	not reported	no
Davis et al., 1992	83	F	acute dyspnea, stridor, neck swelling		not reported	no
Wagh et al., 2004	82	F	acute dyspnea, stridor, wheeze, chest discomfort, neck swelling		pneumatic dilation	no
Berrisford et al., 1998	82	F	acute dyspnea, stridor, cyanosis, dysphagia	tracheal intubation	airway stent	no
Molena et al., 2016	81	F	acute dyspnea, stridor, wheeze, neck swelling, distention of jugular veins		botulinum toxin	yes, unknown
Thomas et al., 2009	81	F	acute dyspnea, stridor, dysphagia		not reported	no
Aubignat et al., 2020	80	F	acute dyspnea, syncope, cardiopulmonary arrest	tracheal intubation	botulinum toxin	no
Wechalekar et al., 2008	80	F	chest pain, stridor, distention of jugular veins, dysphagia, weight loss	tracheal intubation	pneumatic dilation	no
Requena et al., 1999	80	F	acute dyspnea, stridor, dysphagia, wheeze	tracheal intubation	botulinum toxin	yes, 8 month earlier
Doshi et al., 2009	79	F	periodic dyspnea, dysphagia, regurgitation, weight loss, dizziness, neck swelling, stridor, wheeze		not reported	yes, 9 years earlier
Brujins et al., 2009	79	F	respiratory arrest	tracheal intubation	botulinum toxin	no
Ho et al., 2008	78	F	acute dyspnea, stridor, chest pain, cyanosis, dysphagia	tracheal intubation	myotomy	no
Khan et al., 2007	78	F	acute dyspnea, stridor, wheeze, cyanosis, chest discomfort, vomiting, weight loss, cough		pneumatic dilation	no
Maclachlan et al., 2005	78	M	dyspnea, stridor		myotomy	yes, 3 years earlier
Carlsson-Nordlander et al., 1987	78	F	acute dyspnea, stridor, distention of jugular veins, neck swelling	tracheal intubation	not reported	yes, unknown
Hatakeyama et al., 2010	77	F	acute dyspnea, stridor, cyanosis, consciousness disturbance	tracheal intubation	pneumatic dilation	no
Campbell et al., 1995	77	M	stridor, cyanosis, distention of jugular veins, neck swelling		myotomy	yes, 3 months earlier
Leeaphorn et al., 2013	76	F	acute dyspnea, stridor, neck swelling	tracheal intubation	not reported	no
Bello et al., 1950	75	F	hoarseness, neck swelling, dysphagia, thoracic tightness		pneumatic dilation	no
Zikk et al., 1989	74	F	acute dyspnea, stridor, cough, dysphagia		pneumatic dilation	no
Moloney et al., 1987	74	F	acute dyspnea, stridor, neck swelling		not reported	no
Basavarajaiah et al., 2011	73	F	acute dyspnea, dysphagia, weight loss, stridor, distention of jugular veins, neck swelling		botulinum toxin, pneumatic dilation	no
Travis et al., 1981	73	F	acute dyspnea, stridor, cyanosis, distention of jugular veins, consciousness disturbance	tracheal intubation	myotomy	no
Aslam et al., 2007	72	F	acute dyspnea, dysphagia, neck swelling, vomiting, stridor, hoarseness		pneumatic dilation	yes, unknown
Tasker et al., 1995	72	F	cough, stridor, chest pain, nausea, vomiting, dysphagia, chest discomfort, neck swelling		pneumatic dilation	no
Tasker et al., 1995	71	F	acute dyspnea, stridor, chest pain, chest discomfort, dyspepsia, vomiting, neck swelling, distention of jugular veins, regurgitation		pneumatic dilation	no
McLean et al., 1976	70	F	acute dyspnea, stridor, wheeze, cyanosis, neck swelling, distention of jugular veins	tracheal intubation	myotomy	yes, unknown

(*continued on next page*)

Table 1 (*continued*)

Author, year	Age	Sex	Presenting symptoms	Artificial Airway	Definitive treatment	Achalasia previously diagnosed
Kendall	68	F	acute dyspnea, stridor, cyanosis, consciousness disturbance		pneumatic dilation	no
Barr et al., 1989	65	F	acute dyspnea, stridor, cyanosis	tracheostomy	myotomy	yes, unknown
Brock et al., 1986	65	F	acute dyspnea, wheeze, distention of jugular veins, neck swelling	tracheal intubation	myotomy	no
King et al., 1979	65	F	acute dyspnea, dysphagia, neck swelling		myotomy	yes, unknown
Blaney et al., 2020	63	F	acute dyspnea, dysphagia, throat pain, nausea, frequent belching		pneumatic dilation	no
Saoraya et al., 2018	59	F	acute dyspnea, stridor, bradycardia	tracheal intubation	not reported	no
Turkot et al., 1997	59	F	acute dyspnea, hoarseness, stridor, cyanosis, distention of jugular veins	tracheal intubation	pneumatic dilation	no
Dominguez et al., 1987	59	F	acute dyspnea, hoarseness, stridor, cyanosis, dysphagia, neck swelling		myotomy	yes, unknown
Suffoletto et al., 2008	56	F	stridor		myotomy	yes, unknown
Becker et al., 1989	56	F	acute dyspnea, stridor, wheeze, cyanosis, dysphagia, neck swelling	tracheal intubation	not reported	yes, unknown
Collins et al., 1984	55	F	acute dyspnea, dysphagia, neck swelling	tracheal intubation	myotomy	no
Giustra et al., 1973	55	F	acute dyspnea, dysphagia		died prior to intervention	no
Hifumi et al., 2013	53	F	acute dyspnea, cardiopulmonary arrest, consciousness disturbance, dysphagia, vomiting	tracheal intubation	pneumatic dilation	no
Miyamoto et al., 2011	52	F	acute dyspnea, stridor, difficulty in belching, neck swelling, wheeze		not reported	yes, 12 years earlier
Lindenmann et al., 2012	51	F	chest tightness, acute dyspnea, wheeze, cyanosis, consciousness disturbance	tracheal intubation	esophagectomy	yes, 11 years earlier
Giustra et al., 1973	51	F	acute dyspnea, stridor, wheeze		myotomy	no
Genc et al., 2014	50	M	acute dyspnea, stridor, wheeze, neck swelling, dysphagia, regurgitation		pneumatic dilation, myotomy	yes, 10 years earlier
Healy et al., 2007	50	F	acute dyspnea, dysphagia, stridor, cyanosis, bradycardia	tracheal intubation	myotomy	yes, 12 years earlier
Desprez et al., 2019	49	F	acute dyspnea, wheeze		per-oral endoscopic myotomy	no
Adamson et al., 2013	47	F	acute dyspnea, weight loss, dysphagia, neck swelling	tracheal intubation	botulinum toxin	no
Mabvuure et al., 2014	40	M	acute dyspnea, consciousness disturbance, cardiopulmonary arrest	tracheal intubation	esophagectomy	yes, 2 months earlier
Chew et al., 2015	38	F	acute dyspnea, pulmonary arrest	tracheal intubation	myotomy	no
Chijimatsu et al., 1980	36	M	regurgitation		myotomy	no
Gomez-Larrauri et al., 2017	34	F	acute dyspnea, cough, fatigue, wheeze, stridor, sputum		myotomy	no
Suarez et al., 2018	25	M	acute dyspnea, stridor, cyanosis, wheeze, consciousness disturbance	tracheal intubation	botulinum toxin	no
Kaths et al., 2015	23	M	dysphagia, weight loss, chest pain, dyspnea, regurgitation		myotomy	no
Panzini et al., 1993	23	M	acute dyspnea, stridor, wheeze		myotomy	yes, unknown
Aydin et al., 2013	18	M	cough, dyspnea, wheeze, regurgitation		myotomy, esophagectomy	no
Present case, 2020	78	M	acute dyspnea, stridor, wheeze	noninvasive ventilation		yes, 11 years earlier

References

[1] J.E. Pandolfino, A.J. Gawron, Achalasia: a systematic review, JAMA 313 (2015) 1841–1852, https://doi.org/10.1001/jama.2015.2996.

[2] J. Hatakeyama, T. Takei, T. Ito, M. Takemoto, Airway obstruction in a patient with achalasia: a case report and review of the literatures, J. Jpn. Assoc. for Acute Med. 21 (2010) 377–382 https://doi.org/10.3893/jjaam.21.377 (Japanese).

[3] R.S. Dudnik, J.A. Castell, D.O. Castell, Abnormal upper esophageal sphincter function in achalasia, Am. J. Gastroenterol. 87 (1992) 1712–1715.

[4] F. Yoneyama, M. Miyachi, Y. Nimura, Manometric findings of the upper esophageal sphincter in esophageal achalasia, World J. Surg. 22 (1998) 1043–1046, https://doi.org/10.1007/s002689900514.

[5] B.T. Massey, W.J. Hogan, W.J. Dodds, R.O. Dantas, Alteration of the upper esophageal sphincter belch reflex in patients with achalasia, Gastroenterology 103 (1992) 1574–1579, https://doi.org/10.1016/0016-5085(92)91180-c.

[6] A. Gomez-Larrauri, S. Galloway, R. Niven, Achalasia with massive oesophageal dilation causing tracheomalacia and asthma symptoms, Respir Med Case Rep 23 (2018) 80–82.

[7] M. Aubignat, P.A. Roger, A. Dernoncourt, V. Salle, A. Smail, C. Gourguechon, et al., Acute airway obstruction and cardiopulmonary arrest due to tracheomalacia caused by megaesophagus compression secondary to achalasia, Case Rep Pulmonol 2020 (2020) 5946985, https://doi.org/10.1155/2020/5946985.

[8] P. Leong, P.G. Bardin, K.K. Lau, What's in a name? Expiratory tracheal narrowing in adults explained, Clin. Radiol. 68 (2013) 1268–1275, https://doi.org/10.1016/j.crad.2013.06.017.