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Case report

Management of an elderly patient with respiratory failure due to double aortic arch

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ABSTRACT

Vascular rings are congenital malformations of the aortic arch. A double aortic arch (DAA), the most common type of vascular ring, results from the failure of the fourth embryonic branchial arch to regress, leading to an ascending aorta that divides into a left and right arch that fuse together to completely encircle the trachea and esophagus. The subsequent DAA causes compressive effects on the trachea and esophagus that typically manifests in infancy or early childhood. Adult presentations, particularly in the elderly, are exceedingly rare. Historically such patients have a long-standing history of dyspnea on exertion and dysphagia, with many assumed to have obstructive lung or intrinsic cardiac disease. We describe a case of an elderly woman who presented with respiratory failure due to DAA. In her case, surgery was not feasible and we describe our experience with airway stenting.

1. Introduction

Vascular rings are a common congenital anomaly of the aortic arch. A double aortic arch (DAA), the most frequent type of vascular ring, results from the failure of the fourth embryonic branchial arch to regress. The subsequent ascending aorta divides into a left and right arch that fuse together to completely encircle the trachea and esophagus [1,2]. The resultant DAA exerts compressive effects on the trachea and esophagus, which typically manifest with dyspnea and dysphagia during infancy or early childhood. Surgical correction is first-line treatment when possible [3,4].

Symptomatic presentation of DAA in adulthood is extremely rare, particularly in the elderly. Adult patients often have a long-standing history of dyspnea on exertion and dysphagia, with many assumed to have asthma, chronic obstructive pulmonary disease (COPD), or intrinsic cardiac disease. In many instances, similar to childhood cases, operative management is attempted [5–7].

We report our case of an elderly patient who presented with a DAA, which has been rarely described in the literature. We aimed to discuss the key clinical elements of her care, particularly the treatment challenges that arose. In her case surgery was not feasible, and we describe our experiences with airway stenting.

2. Case presentation

Our patient was a 79 year-old caucasian woman with a past medical history significant for diastolic heart failure, asthma, COPD, and “pulmonary fibrosis” who was admitted for worsening shortness of breath. She was treated for both a heart failure exacerbation with diuresis, achieving a net negative fluid balance of over 4 L, and for a COPD exacerbation with oral steroids and scheduled bronchodilators. However, despite these aggressive measures, her respiratory status continued to deteriorate to where she required transfer to our step-down unit for high-flow nasal cannula.

Her pulmonary history revealed that she was diagnosed with asthma as a child, where she only required intermittent inhaler use and was never hospitalized. She complained of dyspnea on exertion for most of her adult life as she always experienced shortness of breath when ambulating far distances. She was eventually started on bronchodilator therapy for presumed COPD given her airway obstruction on pulmonary function tests (PFTs), which provided minimal benefit. Prior to coming to our institution, she was additionally diagnosed with pulmonary fibrosis and was started on continuous oxygen. She was also given a continuous positive airway pressure (CPAP) machine that she adamantly refused to use. On review of systems she endorsed a long-standing history of difficulty swallowing with a sensation of choking that worsened with

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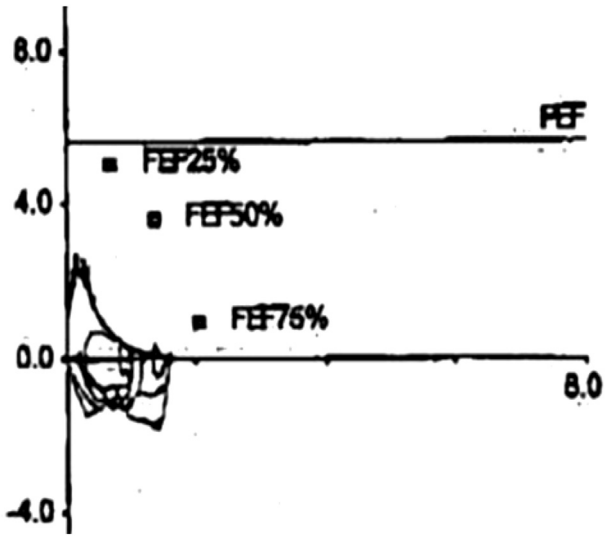

Fig. 1. Airway obstruction detected on patient's flow-volume loop.

activity.

On examination she was short of breath after walking from the bed to her chair. There was no evidence of stridor, wheezes, or crackles. A review of her PFTs revealed fixed obstruction (Fig. 1). Her chest CT (Fig. 2A and B) showed a DAA and a dilated esophagus without evidence of pulmonary fibrosis. Her barium esophagram (1C) showed narrowing of the esophagus at the site of the DAA. Reconstructed images (1D) demonstrated a complete ring that encircled the trachea and esophagus. We performed flexible bronchoscopy to define the tracheal narrowing seen on her chest CT. At the level of her DAA she had a fixed anterior tracheal compression and dynamic near-complete collapse of the trachea from malacia (Fig. 3).

We concluded that her dyspnea was multifactorial. In addition to pulmonary edema due to heart failure, she had significant radiographic compression of her trachea and esophageal narrowing. This likely caused wheezing, which contributed to her prior diagnoses of asthma and COPD. Additionally, her esophageal narrowing with proximal distention likely contributed to chronic aspiration. We suspect this led to some bronchiectasis, ostensibly misinterpreted as pulmonary fibrosis.

We discussed several treatment possibilities, including surgery, which is the treatment of choice. However she declined surgical intervention, although her co-morbidities and clinical condition would have likely rendered her an inoperable candidate as well. Moreover, during surgery, her aortic calcification would make vascular clamping and over-sewing technically challenging while also increasing her risk of distal calcific embolization. Tracheobronchoplasty was also considered for her tracheobronchomalacia. While reported to be successful, we deemed that the risks of the procedure, including the procedure length (373 ± 93 min), complications (infection, pulmonary embolism, atrial fibrillation, recurrent intubation, and aspiration), and mortality rate (3.2%), exceeded potential therapeutic benefits [8]. Furthermore, tracheobronchoplasty did not address her DAA; concurrent surgical correction of a vascular ring has never been reported.

Given her deteriorating respiratory status, we deemed airway stenting her only viable option. Although tubular stenting would be feasible for isolated compression, her diffuse severe malacia required tracheobronchial Y-stent placement. We performed rigid

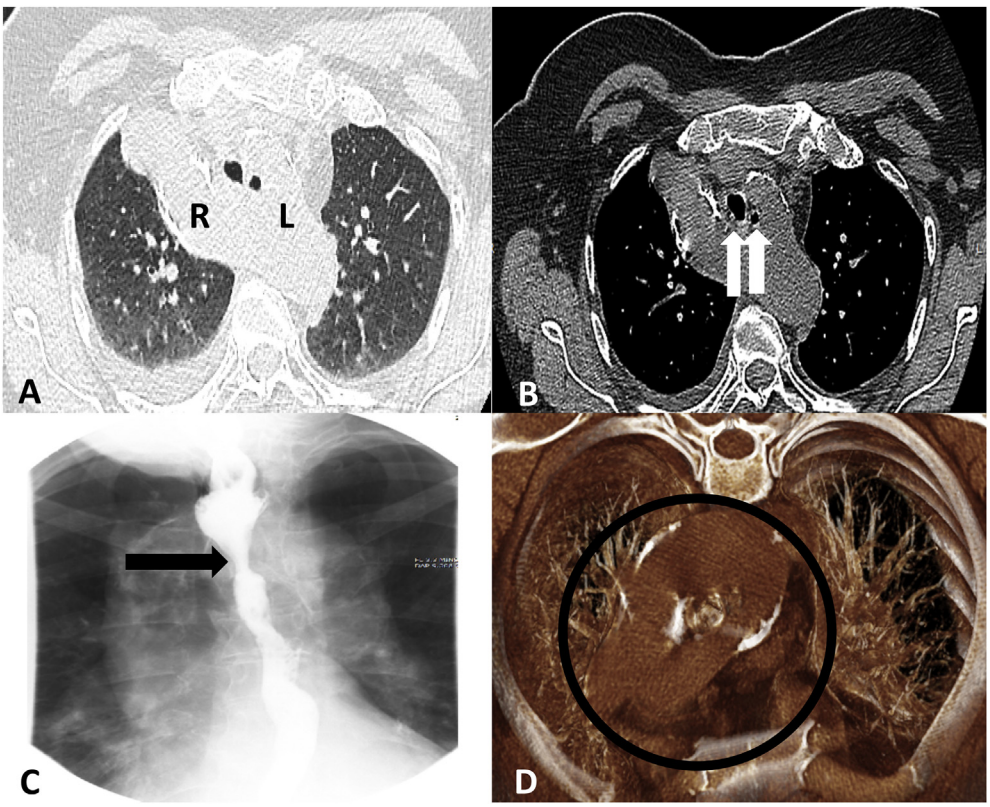

Fig. 2. Radiographic images demonstrating DAA. The right (R) and left (L) arch are seen (A), along with the trachea and esophagus (B, arrows). Esophagram (C) demonstrates obstruction and reconstructed images (D, circle) highlight the DAA. Her aortic calcification is also readily evident.

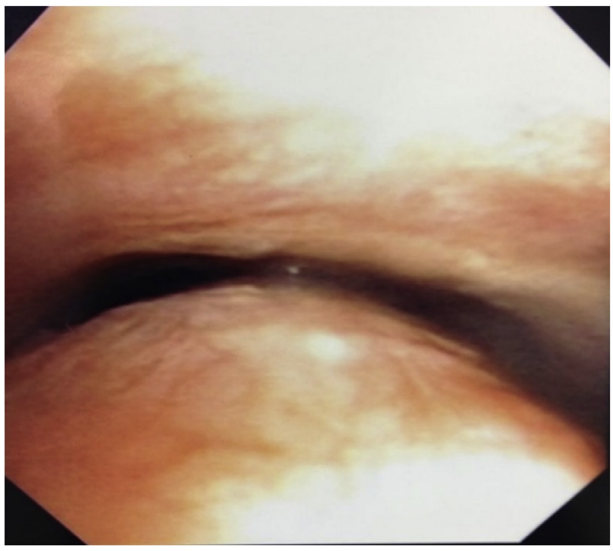

Fig. 3. The anterior trachea, as seen during bronchoscopy, is flattened. Significant expiratory collapse is seen of the posterior membranous tracheal wall.

bronchoscopy with placement of a 15 × 12 × 12 mm Y-stent that immediately stabilized her trachea and mainstem bronchi.

For her dysphagia, we considered esophageal stenting. However, she successfully passed pills during a barium esophagram, and the gastroenterology consultants felt she did not have critical obstruction meriting stent placement.

Following stent placement she developed recurrent episodes of severe mucus obstruction despite aggressive airway clearance techniques with scheduled bronchodilators and chest physiotherapy. Her respiratory status continued to worsen and her family requested comfort measures only, and she eventually died.

3. Discussion

Adult presentations of vascular rings are exceptionally rare. Dyspnea on exertion is the most common symptom. A consequence of normal aging results in aortic dilatation of 0.1 cm per decade of life so that the obstructive effects of the DAA are not appreciated until late adulthood. Hypertension and atherosclerosis can accelerate this process and further increase the tortuosity of the aorta. Finally, age-related changes in the vertebrae and thorax limit the dimensions of the mediastinum, exacerbating the compressive effects of the DAA [6]. In our patient, we postulate that early in life, when her DAA was more compliant, she only manifested mild symptoms. Subsequent age-related changes, including significant calcification of the aorta, led to vascular stiffening causing her debilitating symptoms.

In addition to airway compromise from extrinsic obstruction, many patients develop malacia at the site of the arch. This likely develops from acquired airway wall weakness secondary to long-standing compression and failure of normal development [2].

Airway stenting for vascular anomalies has rarely been described. Although bronchial [9] and tracheal stents [10] have been reported, their efficacy remains uncertain and limited to unusual cases.

By discussing therapeutic strategies for symptomatic vascular rings in adulthood, namely DAA, we aimed to increase an understanding of these unique causes of dyspnea. It is imperative to perform the appropriate testing with a multidisciplinary evaluation to determine the best management in adults with vascular rings.

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