Case report

Pulmonary hypertension and congenital bronchial atresia: A time factor association

Prince A. Alebna, David H. Kim, Raghav Chaudhary, Matthew Tavares

A B S T R A C T

Congenital bronchial atresia is rarely symptomatic in adults. Recurrent lung infection and pneumothorax are the feared complications of this otherwise benign condition. The objective of this article is to present a case of congenital bronchial atresia manifesting as pulmonary hypertension in a 66 year-old patient. While doing so, we highlight the relevant knowledge accrued in medical literature with regards this rare condition. Finally, with the revelation that congenital bronchial atresia may cause severe pulmonary hypertension later in life, perhaps a less conservative approach to management may be warranted in younger adults and children with this condition.

1. Introduction

Congenital bronchial atresia (CBA), marked by complete focal occlusion of part of the bronchial tree lumen, is often an asymptomatic benign condition with people who have CBA living well into adulthood unaware they have the condition. Recurrent lung infection is the main complication reported in literature on CBA. This case highlights a potential long-term complication of CBA that was hitherto undокументed.

2. Case description

A 66 year old female with a history of hypertension, coronary artery disease and atrial fibrillation status post ablation presented with dyspnea on exertion for two days. She did not have orthopnea, paroxysmal nocturnal dyspnea, ankle swelling, snoring or excessive daytime sleepiness. Breath sounds were diminished in her left upper lung zone and she had fine bibasilar crackles. The first imaging modality, a postero-anterior chest Xray (Fig. 1) was initially reported as unremarkable. Computerized tomography (CT) chest with contrast ruled out pulmonary embolism but revealed evidence of right heart strain, interstitial edema and hyperlucent left upper lobe consistent with CBA (Fig. 2). Left ventricular ejection fraction was 50–55% on echocardiography. Left heart catheterization revealed patent coronary arteries and right heart catheterization revealed severe pulmonary hypertension (PH) with a mean pulmonary arterial pressure (mPAP) of 48 mmHg and pulmonary capillary wedge pressure (PCWP) of 24mmHg. Vasodilator testing was deferred as the patient was deemed too ill. Inflammatory markers were not elevated and autoimmune disease screen was negative. The core of her management was based on intravenous diuresis. Despite resolution of all the clinical signs of pulmonary edema, she still had some degree of symptomatic limitation to moderate activity. She was eventually discharged with oral loop diuretic on the fourth day of admission and symptoms have been stable at outpatient follow-up.

3. Discussion

Congenital bronchial atresia (Fig. 3) is a rare condition which affects the lobes, segmental or subsegmental bronchi. Though the exact etiology is unknown, it is postulated that CBA is consequent to an ischemic event which may have occurred during the developmental stages of the lungs in utero leading to complete occlusion of the affected segment [1]. The respiratory tree beyond the lesion is often not affected and develops normally. CBA is classified under the broad category of bronchopulmonary anomalies in contrast to vascular anomalies [2]. Combined anomalies of both the bronchopulmonary and vascular structures may occur in the same patient. Clinically, it is more relevant to be cognizant of the potential complications resulting from the very close pathophysiological relationship between the airways and the vasculature. Only a few cases of CBA in adults are reported in literature with most write-ups originating from Asia and Europe. With a male preponderance, the prevalence is estimated at 1.2 per 100,000 males [3,4]. Severe cases are often detected in the pediatric age group and will not be addressed in this paper. Asymptomatic cases may go undiagnosed until chest imaging is performed for some other reason in adulthood, though, with advancement in diagnostic technology,
prenatal diagnosis of CBA is now possible via sonography [5]. CBA occurs most frequently in the apicoposterior segmental bronchus of left upper lobe [6].

Clinically, the only diagnostic clue is diminished breath sounds on auscultation of the affected lung zone during physical examination. CT of the chest is the most sensitive diagnostic modality [7]. The main finding on CT chest is hyperlucency which is due to hyperinflation of affected alveoli and compression of adjacent tissues. Bronchoscopy is only indicated in doubtful cases.

The hyperinflation observed on CT chest results from unidirectional airflow via collateral canals called pores of Kohn and canals of Lambert [8,9]. The emphysematous changes in the post-atretic airways lead to the creation of a physiological dead space. Additionally, neighboring parenchyma is compressed by the hyperinflated segment or lobe further compromising blood flow in this zone. While anterograde air flow into the affected alveoli is absent, arterial supply is maintained. The hyperinflated lobe induces a ventilation/perfusion mismatch resulting from physiological vasoconstriction [10]. The level of vascular resistance due to all the factors mentioned above, depends on the size of the affected area of lung. Referring to the index case, the elevation of her mPAP was comparatively way out of proportion compared to the elevation in her PCWP. A strong linear relationship exist between mPAP and PCWP. This relationship is mathematically represented as follows: mPAP = [0.94 + (1.15 x mean PCWP) + (0.03 x age) + (0.07 x heart rate)] mmHg [11]. A constant value of 1.34 is added for females. Hence, with a PCWP of 24 mmHg, the estimated mPAP should have averaged 37 mmHg. Therefore, although she had left heart failure, her PH was in part caused by the sequelae of the CBA, as no other lung pathology was noted. We believe that after several years and decades the increased vascular resistance created by the presence of CBA will eventually manifest as PH. In contrast to pathologies such as massive pulmonary embolus which cause resistance in series to the right ventricles resulting in acute onset of symptoms, CBA causes resistance in parallel which, depending on the size of affected vasculature, may take decades to cause symptomatic PH (Fig. 4).

Recurrent infections and pneumothorax are the major known complications of CBA. There is also a single reported case of non-tuberculous mycobacterial infection associated with CBA [12]. Prior to this paper CBA had not gained the reputation as a potential etiology of PH. Several developmental lung abnormalities have been identified as etiologies of PH [13]. This case highlights how CBA, a rare congenital condition causes PH.

With regards management of CBA in patients with complications the consensus is surgery. Surgical options include local resection or lobectomy via either open thoracotomy or thoracoscopic resection [14,15]. However, there are no societal guidelines for the management of CBA in adult patients. While surgery is widely accepted as appropriate in patients with known complications, opinions are divided on surgery in asymptomatic patients. Transplant surgery may improve the quality of life and survival in patients who have developed severe PH.

Fig. 1. Postero-anterior view of her chest Xray illustrating an increased cardiotoracic ratio. Left upper translucency is apparent on the Xray.

Fig. 2. (A) Transverse view of the CT chest showing hyperlucent left upper lobe and the main pulmonary artery which is larger than the aorta suggestive of pulmonary hypertension. (B) Coronal view of the CT chest showing hyperlucent left upper lobe (red arrow). (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)

Fig. 3. An illustration of the bronchovascular anatomy in CBA. (A) Pulmonary artery (B) Pulmonary vein (C) Capillary bed around the alveolus (D) Constricted blood vessel around a post-atretic hyperinflated alveolus (E) Focal atresia of the bronchial tree (F) Hyperinflated alveolus (G) Depiction of collateral airflow through Canal of Lambert and Pores of Kohn (H) Normal alveolus.
from CBA. However, multiple comorbidities, advanced age, lack of insurance plan and lack of a strong social support system may hinder access to transplant surgery as was the case in the index case.

4. Conclusion

Given the risk of developing pulmonary hypertension later in life, early surgical intervention in children and young adults diagnosed with CBA may be beneficial. More research is needed to guide the management of CBA.

5. Learning objectives

- Chest CT is the gold standard for diagnosing CBA.
- Recognize PH as a late complication of CBA
- Early diagnosis and surgical intervention in both symptomatic and asymptomatic may prevent later development of PH.

Funding

None.

Conflicts of interest

None of the authors have any conflicts of interest to disclose.

Authorship

All authors had access to the case and participated in preparation of this manuscript.

Disclosure

All authors have participated in the work and have reviewed and agree with the content of the article. This paper has not been published in any journal and is not under consideration for publication in any other journal.

References

[1] R.L. Meng, R.J. Jensik, L.P. Faber, G.R. Matthew, C.F. Kittle, Bronchial atresia, Ann. Thorac. Surg. 25 (3) (1978) 184–192, https://doi.org/10.1016/S0003-4975(10)63519-9.
[2] C.J. Zylak, W.B. Eyler, D.L. Spizarny, C.H. Stone, Developmental lung anomalies in the adult: radiologic-pathologic correlation, Radiographics (2013), https://doi.org/10.1148/ radiographics.22.suppl_1.g02oc26s25.
[3] T.J. Shipchandler, K. Geelan-Hansen, P.R. Krakovitz, Complete bronchial stricture and airway management challenges, Am. J. Otolaryngol. Head Neck Med. Surg. 29 (3) (2008) 195–197, https://doi.org/10.1016/j.amjoto.2007.04.009.
[4] K. Paithakhi, S. Lachanis, C. Kotoulas, et al., The prevalence of congenital bronchial atresia in males, Monaldi Arch chest Dis = Arch Monaldi per le Mal del torace/Fond Clin del Lav (BCCS [and] Ist di Clin Tisiti e Mal Appar Respir Univ di Napoli, Second ateneo 61 (1) (2004) 28–34.
[5] N.J. Hall, M.P. Stanton, Long-term outcomes of congenital lung malformations, Semin. Pediatr. Surg. (2017), https://doi.org/10.1055/s-0037-1610001.
[6] E. Vancamp, R. Salgado, T.H. Mulhens, J.L. Termote, P.M. Parizel, Bronchial Atresia, JBR-BTR, 2015, https://doi.org/10.5334/jbr-btr.808.
[7] C. Beigelman, N.R. Howarth, C. Chartrand-Lefebvre, P. Grenier, Congenital anomalies of tracheobronchial branching pattern: spiral CT aspects in adults, Eur. Radiol. (1998), https://doi.org/10.1007/s003300050343.
[8] S. Rajagopala, U. Devaraj, S. Swamy, A. Kumar, Mucoid impaction in a 15-year-old with bronchial atresia, Respir. Care 57 (3) (2012) 471–473, https://doi.org/10.4187/respcare.01196.
[9] U. BUCHER, L. REID, Development of the intrasegmental bronchial tree: the pattern of branching and development of cartilage at various stages of intra-uterine life, Thorax 16 (1961) 207–218, https://doi.org/10.1136/thx.16.3.207.
[10] S. Intagliata, A. Rizzo, Physiology, Lung Dead Space, StatPearls, 2018, http://www.ncbi.nlm.nih.gov/pubmed/29494107.
[11] R. Ihsr, B.P. Bailey, T.A. Dobbins, D.S. Gelermaier, The relationship between pulmonary artery and pulmonary capillary wedge pressure for the diagnosis of pulmonary vascular disease, Heart Lung Circ. 19 (1) (2010) 38–42, https://doi.org/10.1016/j.hlc.2009.08.001.
[12] K. Matsuoaka, M. Ueda, Y. Miyamoto, Mycobacterium avium infection complicating congenital bronchial atresia, Asian Cardiovasc. Thorac. Ann. 24 (8) (2016) 808–810, https://doi.org/10.1111/act.12463.
[13] G. Simonneau, M.A. Gatzoulis, I. Adatia, et al., Updated clinical classification of pulmonary hypertension, J. Am. Coll. Cardiol. 62 (25 Supplement) (2013), https://doi.org/10.1016/j.jacc.2013.10.029 D34 LP-D41.
[14] Y. Wang, W. Dai, Y. Sun, X. Chu, B. Yang, M. Zhao, Congenital bronchial atresia: diagnosis and treatment, Int. J. Med. Sci. 9 (3) (2012) 207–212, https://doi.org/10.7150/ijms.3690.
[15] M. Kamiyoshitahara, A. Onuki, T. Nameki, et al., Congenital bronchial atresia treated with video-assisted thoracoscopic surgery; report of a case, Kyobu Geka 57 (7) (2004).