Research Paper Pediatrics

Isolated Bronchial Stenosis with Pulmonary Artery Hypoplasia Presenting with Recurrent Wheezing

Anupama Mauskar Professor (Additional) Department of Pediatrics, LokmanyaTilak Municipal Medical College and General Hospital, Mumbai – India

Ronak Sheth Senior Registrar, Department of Pediatrics, LokmanyaTilak Municipal Medical College and General Hospital, Mumbai – India

Santosh Prasad Senior Registrar Department of Pediatrics, LokmanyaTilak Municipal Medical College and General Hospital, Mumbai – India

KEYWORDS Isolated bronchial stenosis, Recurrent wheezing.

Introduction

Broncho-pulmonary Vascular Malformations (BPVM) are rare congenital abnormalities involving the airways and pulmonary vascular tree; worldwide incidence varies from 30 to 42 cases per 100,000 population or 0.06% to 2.2% of all patients admitted to general hospitals.(1,2). They present commonly as recurrent pneumonias and respiratory distress in infancy. We present a case of congenital BPVM who presented after infancy with recurrent wheezing as the predominant manifestation.

Case Summary

We report a case of two and half year old female child presented with repeated episodes of cough, breathlessness and wheezing treated with salbutamol nebulization and oral antibiotics every time. At the age of 18 months, an episode with severe breathlessness and some hemoptysis required admission and treatment with oxygen, bronchodilators and steroids. She had persistently reduced air entry in left mammary and infra clavicular area with recurrent wheezing episodes for which she was thoroughly investigated.Bronchoscopy, computerized tomography chest with pulmonary angiography, revealed a 9mm long stenotic segment of the left main bronchus, with minimum diameter of 0.8 mm; with marked paucity of sub segmental pulmonary arterial branches in the left lung(Fig-1)

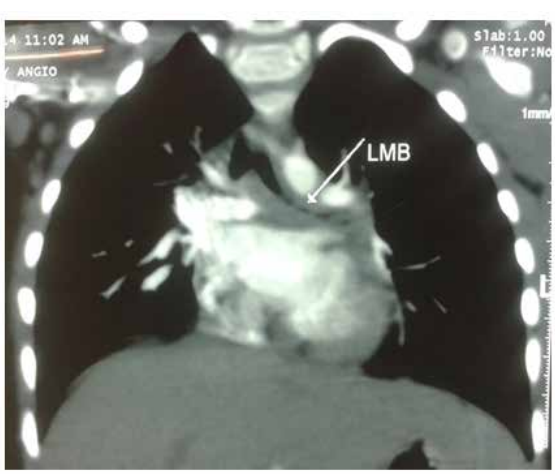

Fig-1 High Resolution Computerized Tomography Chest with Pulmonary angiography showing narrowing of Left main bronchus & paucity of segmental Arterial supply

There was no evidence of aberrant vessel pressing on the bronchial wall externally. This supported the diagnosis of congenital BPVM. Her work up for tuberculosis was essentially negative. The child was treated conservatively with Budesonide MDI to which she responded and there were no further wheezing episodes on follow up after 3 months.

Discussion

Congenital pulmonary malformations can be grouped into a continuous spectrum ranging from pure pulmonary parenchymal anomalies to isolated vascular anomalies, with an intermediate group in which both pulmonary and vascular components are intertwined.(3,4,5).To the best of our knowledge, this is a first of its kind case reported in literature, with isolated left main bronchus segmental stenosis (without tracheal involvement) with associated paucity of pulmonary arterial branches with preserved pulmonary parenchymal structure. There is only 1 other case of isolated bronchial stenosis in literature; Chang et al in 1968 had reported a 5 month old child with isolated right main stem bronchial stenosis (symptomatic since neonatal period) successfully treated with surgical excision of the stenotic segment. Patients may be diagnosed antenatally, or may remain clinically silent till adult life. Most common is the presentation in early life with recurrent chest infections. Associated anomalies are common.

CT pulmonary angiography is the desired modality for investigating such cases as CECT chest may be entirely normal in the absence of any structural abnormality. In some cases, Digital Subtraction Angiography (DSA) may be required for complete evaluation, and for pre-operative assessment, in cases where the lesions are amenable to surgical correction (6, 7). Asymptomatic or mildly symptomatic patients can be managed conservatively. Those with severe progressive respiratory morbidity or ventilator dependence should be aggressively considered for surgical intervention as surgical outcomes are overall good (8). Focal pulmonary hypertension is a known complication in adults with such congenital pulmonary vascular anomalies. Hence regular screening for pulmonary hypertension should be done in this group of patients.

Conclusion

In children who present with recurrent wheezing, focal chest findings are most commonly due to mucous plugging of the hyper-reactive airways. But persistence of focal findings or development of an unusual symptom like hemoptysis or unusual ventilator dependence should prompt a thorough workup as foreign body and congenital BPVM are important differentials, once common things like tuberculosis and asthma are ruled out. A high index of suspicion is essential for prompt diagnosis and timely management.

Acknowledgement:
we are thankful to the dean DrSuleman Merchant for permitting us to publish this manuscript.

REFERENCES

1. Sylvester KG, Albanese GT. Bronchopulmonary Malformations. In: Ashcraft KW, Holcomb GW, Murphy JP, Ashcraft's pediatric surgery. Philadelphia: Saunders Elsevier; 2005. p. 276-89. | 2. Costa Júnior Ada S, Perfeito JA, Forte V. Surgical treatment of 60 patients with pulmonary malformations: what have we learned? J Bras Pneumol. 2008; 34(9):661-6. | | 3. Lee M-L, Lue H-C, Chiu I-S, Chiu H-Y, Tsao L-Y, Cheng C-Y, et al. A Systematic Classification of the Congenital Bronchopulmonary Vascular Malformations: Dysmorphogeneses of the Primitive Foregut System and the Primitive Aortic Arch System. Yonsei Medical Journal. 2008; 49(1):90-102. | 4. Clements BS, Warner JO. Pulmonary sequestration and related congenital bronchopulmonary-vascular malformations: nomenclature and classification based on anatomical and embryological considerations. Thorax. 1987; 42(6):401-8. | 5. Lee ML, Tsao LY, Chaou WT, Yang AD, Yeh KT, Wang JK, et al. Revisit on congenital bronchopulmonary vascular malformations: a haphazard branching theory of malinosculations and its clinical classification and implication. Pediatric pulmonology. 2002; 33(1):1-11. | 6. Paterson A. Imaging evaluation of congenital lung abnormalities in infants and children. Radiologic clinics of North America. 2005; 43(2):303-23. | 7. Watarai F, Takahashi M, Hosoya T, Murata K. Congenital lung abnormalities: a pictorial review of imaging findings. Japanese journal of radiology. 2012; 30(10):787-97. | 8.Salles M, Deschildre A, Bonnel C, Dubos JP, Bonnevalle M, Devismes L, et al. [Diagnosis and treatment of congenital bronchopulmonary malformations. A review of 32 cases]. Archives de pediatrie: organe officiel de la Societe francaise de pediatrie. 2005; 12(12):1703-8.