

# Pneumonectomy for Congenital Isolated Unilateral Pulmonary Artery Agenesis

J Britton, MBBS\*, A Sachithanandan, FRCSI (CTh)\*\*, L Srinivasan, FRCS\*, S Ghosh, FRCS (CTh)\*

\*Department of Cardiothoracic Surgery, University Hospital North Staffordshire NHS Trust, Stoke-on-Trent, ST4 7LN, United Kingdom, \*\*Department of Cardiothoracic Surgery, Hospital Serdang, Jalan Puchong, 43000 Kajang Selangor, Malaysia

## SUMMARY

**Unilateral pulmonary artery agenesis (UPAA) is a rare congenital anomaly usually diagnosed in infancy due to associated cardiovascular malformations. We report a rare case of isolated right UPAA that presented atypically in adulthood with massive haemoptysis requiring a pneumonectomy. This case highlights the importance of maintaining a high clinical suspicion, the role of CT angiography and a multi disciplinary approach. Optimal management is often surgical however bronchial artery embolization (BAE) remains a useful adjunct.**

## KEY WORDS:

*Unilateral pulmonary artery agenesis, Haemoptysis, Pneumonectomy*

## INTRODUCTION

UPAA is a rare congenital anomaly, typically left sided and often diagnosed and treated in infancy due to associated cardiovascular malformations. Patients with isolated UPAA however, can present much later. We report a young adult with isolated right sided UPAA who required a pneumonectomy for recurrent massive haemoptysis.

## CASE REPORT

A 21-year-old female presented with an episode of massive haemoptysis. She had a childhood history of two isolated episodes of minor haemoptysis at age 8 and 16 but was otherwise well. An initial chest radiograph (CXR) showed moderate loss of right lung volume with compensatory hyperexpansion of the left lung. A subsequent computed tomogram (CT) showed fibrosis and honeycombing of the right lung, thought to be acquired following a childhood infection.

At age 17 she had three further episodes of small volume haemoptysis. Bronchoscopy and spirometry were normal. A repeat CT (with 3-D reconstruction) excluded an arteriovenous malformation but demonstrated complete absence of the right pulmonary artery (Figures 1 & 2). The right lung was perfused systemically via extensive mediastinal collaterals; a combination of hypertrophied bronchial arteries arising directly from the aorta, prominent intercostals and sub-diaphragmatic collaterals from the celiac axis.

At this stage however, her symptoms were deemed merely

troublesome and not life threatening hence intervention was thought unnecessary. Four years later she presented to our emergency department with massive haemoptysis and haemodynamic compromise. Initial treatment was supportive with blood transfusion, oxygen and intravenous tranexamic acid. BAE was excluded as a therapeutic option as our interventional radiology team did not think it was possible to selectively identify and safely isolate the culprit bronchial vessel due to extensive collaterals.

The patient proceeded to surgery. At thoracotomy we encountered prominent intercostal vessels with abundant collaterals and a rich plexus on the diaphragm. The right lung was small and cystic in appearance. She had a right pneumonectomy and went home five days later. The patient remains well, haemoptysis-free on follow-up four months later.

## DISCUSSION

UPAA is a rare congenital abnormality usually associated with other cardiovascular anomalies including Fallots tetralogy, septal defects, truncus arteriosus and total anomalous pulmonary venous drainage<sup>1</sup>. The majority are diagnosed and treated in the first year of life. Isolated UPAA is particularly rare and haemoptysis (usually minor and self-limiting), an uncommon presentation in only 10-20% cases<sup>1</sup>. UPAA, first described by Fraentzel in 1868, arises from mal-development of the ipsilateral 6th aortic arch ventral bud<sup>2</sup>. In the absence of an inflow source, the pulmonary circulation is derived from systemic segmental vessels. These submucosal collaterals hypertrophy with time and may rupture causing haemoptysis.

The diagnosis of isolated UPAA requires a high clinical suspicion in any patient with unexplained persistent respiratory symptoms. More often CT or pulmonary angiography is diagnostic, however typical CXR findings include ipsilateral cardiac and mediastinal displacement, elevation of the ipsilateral hemi diaphragm, reduced ipsilateral pulmonary vasculature and contralateral hyperinflation.

In patients with UPAA, the entire cardiac output will be diverted to the contralateral normal lung which may predispose to pulmonary hypertension (PHT) in the long term with associated complications including PA aneurysmal dilatation, dissection and even rupture<sup>3</sup>. A meta analysis by

*This article was accepted: 8 August 2011*

*Corresponding Author: Anand Sachithanandan, Department of Cardiothoracic Surgery, Hospital Serdang, Jalan Puchong, 43000 Kajang Selangor, Malaysia Email: anandsachithanandan@yahoo.com*



**Fig. 1:** CT thorax scan demonstrating a congenitally absent right pulmonary artery (marked "A" with arrow) with hypoplasia of the right lung.



**Fig. 2:** 3-D reconstruction CT thorax scan illustrating complete agenesis of the right pulmonary artery.

Ten Harkel et al suggests almost half of all isolated UPAA patients will develop PHT due to a progressive increase in the pulmonary vascular resistance<sup>1</sup>. Early surgical intervention may result in regression of PHT before it becomes irreversible and thus prevent right ventricular failure. Optimal management requires a multi-disciplinary approach involving a cardiothoracic surgeon, respiratory physician and interventional radiologist. Treatment options include BAE, surgery or a conservative approach with surveillance depending on symptomatology. In this case an expectant approach was taken until the patient re-presented with massive haemoptysis.

BAE is less invasive and may carry less procedural risk than a pneumonectomy however limitations include lack of expertise, technical difficulty for safe coil release and higher long term recurrence rates (up to 25%) due to the extensive collateralization<sup>4</sup>. Furthermore BAE may take longer, potentially compromising patient safety in an acute scenario. The contribution of the affected lung to functional gas exchange is usually minimal which negates a lung preserving procedure. Our patient had an essentially non-functional hypoplastic cystic, fibrotic right lung. At early follow up our patient appears to have benefited from lung resection with no further haemoptysis and an increased exercise tolerance. BAE however should be considered in less fit patients or as an adjunct to surgery to reduce subsequent surgical blood loss by devascularising the pleural space in preparation for a thoracotomy<sup>5</sup>.

### CONCLUSION

We report a rare case of congenital isolated right UPAA that presented atypically with life-threatening massive haemoptysis in adulthood, treated successfully with a pneumonectomy. This case highlights the importance of maintaining a high clinical suspicion in patients with persistent unexplained respiratory symptoms. CT or pulmonary angiography is usually diagnostic. Optimal management requires a multi disciplinary approach with surgery (pneumonectomy) often the definitive treatment.

### REFERENCES

1. Ten Harkel AD, Blom NA, Ottenkamp J. Isolated unilateral absence of a pulmonary artery: a case report and review of the literature. *Chest* 2002; 122: 1471-7.
2. Thomas P, Reynaud-Gaubert M, Bartoli J-M, Auge A, Garbe L, Giudicelli R et al. Exsanguinating hemoptysis revealing the absence of left pulmonary artery in an adult. *Ann Thorac Surg* 2001; 72 (5):1748-50.
3. Heper G, Korkmaz ME. High- pressure pulmonary artery aneurysm and unilateral pulmonary artery agenesis in an adult. *Tex Heart Inst J* 2007;34(4): 425-30.
4. Chun JY, Belli AM. Immediate and long-term outcomes of bronchial and non-bronchial systemic artery embolisation for the management of haemoptysis. *Eur Radiol* 2010; 20(3): 558-65.
5. Redmond KC, Mansfield L, Griffin N, Mullen M, Dusmet M. Coiling of major aortopulmonary collateral arteries with agenesis of the left pulmonary artery prior to pneumonectomy. *Ann Thorac Surg* 2009; 87(1): 312-4.