HAEMOPTYSIS IN PREGNANCY – IS IT UNILATERAL PULMONARY ARTERY AGENESIS (UPAA)?

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Background

Unilateral pulmonary artery agenesis (UPAA) is a rare congenital pulmonary-vascular abnormality with a prevalence of 1 in 200, 000 to 300, 000[1]. As of 2011, 419 cases of UPAA were reported[2]. It can occur either in isolation or together with other congenital cardiovascular defects such as tetralogy of Fallot, patent ductus arteriosus (PDA) and right sided aortic arch[2]. UPAA can lead to pulmonary hypertension in pregnancy which has high mortality and morbidity rate.

Aims

To contribute to the literature and discuss symptoms, workup and management of UPAA in pregnancy given the low incidence rate.

Case

In 2020, a 29-year-old primiparous lady with UPAA presented to a metropolitan hospital at 35 weeks gestation with haemoptysis with coughing and tachycardia. She was diagnosed with UPAA at 11yo in India after an episode of haemoptysis but had no follow-up. Her only symptoms were haemoptysis and tachycardia which recurred annually, particularly in winter. When she migrated to Australia in 2018, an episode of haemoptysis led to an extensive work-up of her condition. She is otherwise healthy, has good exercise tolerance and has no significant past history. The pregnancy itself was uncomplicated. All her first trimester investigations were unremarkable including the aneuploidy screening and the morphology scan showed clear visualisation of the foetal pulmonary arteries and veins with no abnormalities.

Investigations pre-pregnancy in 2018

- CT chest - atresia of the right pulmonary artery, serpiginous collateral vessels to the right lung hilum, moderate to extensive ground glass appearance of the right lower lobe and a compensatory expansion of left lung.
- Respiratory lung function test - mildly reduced carbon monoxide transfer with normal vital lung capacity and total lung capacity.
- Echocardiogram - normal cardiac parameters and a patent ductus arteriosus (PDA).
- CT angiography - absent right pulmonary artery, a hypoplastic right lung with middle lobe less opaque, systemic collateral arteries of up to 7 mm and no evidence of pulmonary hypertension. A 12x9 mm diverticulum arising from the ascending aorta likely corresponds to the small PDA.

Investigations during pregnancy

- Echocardiogram - normal LV and systolic function, normal right heart appearance and function and a small PDA not affecting the pulmonary artery pressure.
- Chest x-ray – Tracheal and mediastinal shift to the right and expansion of the left lung (not new).
- Sputum culture for mycobacterium and other respiratory organisms – negative.

Management

The patient was transferred to a major tertiary centre whereby she was looked after by a multi-disciplinary team. The cardiology team repeated her echocardiogram and given the stable findings, was happy for a trial of vaginal birth with an early epidural, limited second stage and a dose of antibiotic in labour. The anaesthetic team repeated the CTPA prior to her induction at 38 weeks because of persistent small amount of haemoptysis which also demonstrated stable findings. She had an induction at 37+6 but due to failed ripening, she had an uncomplicated caesarean section.

Discussion

People with UPAA can present with pulmonary hypertension, limited exercise tolerance, dyspnoea, recurrent chest infection and haemoptysis. Around 13% of cases are asymptomatic. A case report published in 2002 by Harkel et al. demonstrated a mortality rate of 7% associated with UPAA with the cause of death being secondary to pulmonary hypertension, right heart failure, pulmonary haemorrhage or high altitude pulmonary edema[3]. Angiogram is the gold standard modality but it is recommended for people requiring surgical intervention and CT and MRI should be used instead for diagnosis. There is no prescriptive guidelines and management such as endovascular embolization, pneumonectomy or surgical revascularisation are usually dependent on the severity and frequency of the signs and symptoms and anatomy. There is also no clear guidelines regarding management of pregnant women with UPAA and recommended mode of delivery.

This case demonstrates the importance of using a multidisciplinary approach to safely care for pregnant patients with a rare congenital condition. A world literature review might help develop a guideline regarding diagnosis and management of UPAA in pregnancy to provide high quality patient-centred care.

References:

1. Fraentzel O et al. Virchows Arch A Pathol Pathol Anat. 1868;43:420.
2. Bouros D et al. The varied manifestation of pulmonary artery agenesis in adulthood. Chest. 1995;108(3):670-6
3. Ten Harkel et al. Isolated unilateral absence of a pulmonary artery: a case report and review of the literature. Chest. 2002;122(4)

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