Obstetric Anaesthesia
Annual Scientific Meeting 2021 goes virtual
10-11 June 2021

Birt-Hogg-Dubé syndrome: a rare case of pneumothorax

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Introduction

Birt-Hogg-Dubé syndrome is a rare and complex genetic condition with multi-organ consequences. As a disease of folliculin, the manifestations can include cutaneous lesions, renal tumours, pulmonary cysts and a predisposition to cancer. We present a case of a woman who developed a spontaneous pneumothorax during the third trimester of pregnancy.

Case report

A 37 year-old woman (G2P1) of 35 weeks' gestation presented to the Emergency Department with a short history of chest pain and shortness of breath. She had a known diagnosis of Birt-Hogg-Dubé syndrome with a history of pneumothoraces. She had no other features of her chronic condition and kept well. The patient was tachycardic, breathless and requiring supplemental oxygen. Chest x-ray showed radiological evidence of a large left -sided pneumothorax (Fig. 1)

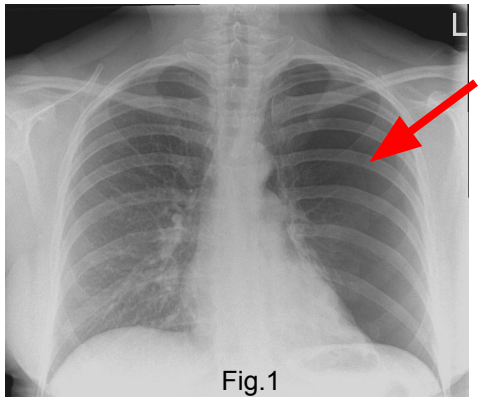


Fig.1

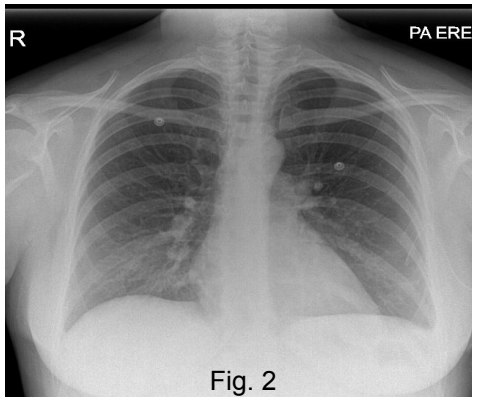


Fig. 2

Obstetric history and examination were unremarkable. The patient was admitted under the respiratory physicians with input from obstetric and anaesthetic teams. An intercostal chest drain was inserted with successful re - inflation of the lung (Fig.2) and symptomatic improvement. However, the pneumothorax reaccumulated the following week and an MDT was organised to discuss expeditious delivery. Advice was sought from the thoracic surgeons regarding definitive treatment of the recurrent pneumothoraces.

Semi-elective caesarean section was organised with the intercostal drain in situ and the patient underwent successful delivery of a live singleton female under a spinal anaesthetic. She was transferred to surgical high dependency where she remained stable, and the intercostal chest drain was able to be removed 24 h following delivery.

The patient was discharged home awaiting thoracic follow up. She has subsequently undergone a VATS pleurodesis and bullectomy and remains well.

Discussion

Birt-Hogg-Dubé syndrome is an autosomal dominant condition infrequently encountered in obstetric practice. The mutation within the FLCN gene codes for the protein folliculin, which impacts on cell division and growth. This commonly manifests as skin tumours, renal lesions and pulmonary cysts of variable severity.

Over 80% of Birt-Hogg-Dubé syndrome patients have pulmonary bullae which may spontaneously rupture leading to an abnormal collection of air within the pleural space.[1] The resulting pneumothorax is a rare and potentially life-threatening condition and must be managed promptly to prevent decompensation of both mother and foetus.

We elected to deliver this patient operatively with the chest drain in situ to mitigate rises in intra -thoracic pressure associated with labour, which may have resulted in expansion of the pneumothorax or secondary rupture of other known pulmonary cysts. Meticulous history and examination were necessary to delineate any other manifestations of this woman's chronic condition.

Multidisciplinary involvement from obstetric, midwifery, anaesthetic, surgical and respiratory teams was key in coordinating a successful outcome for both mother and baby.

References

1. A Gupta N, Kopras EJ, Henske EP et al. Spontaneous pneumothoraces in patients with Birt-Hogg-Dube Syndrome. Ann Am Thorac Soc 2017; 14: 706–13.

Acknowledgements: The authors would like to thank the patient for her consent to publication and the obstetric and midwifery teams involved in her care.