Case Report

Type III Congenital Pulmonary Airway Malformation Associated with Oesophageal Atresia and Tracheoesophageal Fistula. A Case Report and Review of Literature

Nurul Hafiez Fijasri[1], Nur Asmarina Muhammad Asri[1], Mohd Shahrulsalam Mohd Shah[1], Mohd Ridzuan Abd Samad[1], Norsuhana Omar[2]

[1]Department of Surgery, School of Medical Sciences, Universiti Sains Malaysia, Kubang Kerian, [2]Department of Physiology, School of Medical Sciences, Universiti Sains Malaysia, Kubang Kerian, Kelantan, Malaysia

Abstract

Congenital pulmonary airway malformation (CPAM) together with oesophageal atresia and tracheoesophageal fistula (TOF) is a very rare condition in neonates. We presented a case of an infant with Gross type C oesophageal atresia with TOF coexisting with Stocker Type III CPAM in our centre. It is interesting to know that TOF associated with type III CPAM has never been reported in the literature. The child was delivered through caesarean section, and because of respiratory distress post-delivery, endotracheal intubation was carried out immediately. CPAM was diagnosed by a suspicious finding from the initial chest X-ray and the diagnosis was confirmed through computed tomography scan of the chest. The patient was initially stabilised in a neonatal intensive care unit (NICU), and after the successful ligation of fistula and surgical repair of TOF, lung recruitment was started by high flow oscillatory ventilation. The patient recovered well without complications and able to maintain good saturation without oxygen support through the stay in the neonatal unit. Early recognition of this rare association is essential for immediate transfer to NICU, the intervention of any early life-threatening complications, and for vigilant monitoring in the postoperative period.

Keywords: Congenital pulmonary airway malformation type III, thoracopulmonary anomaly, tracheoesophageal fistula type C

Introduction

The coexistence of tracheoesophageal fistula (TOF) with CPAM is rare in neonates. However, a high index of suspicion of this association should always be practised since this condition may contribute to serious complications. Worsening respiratory distress with abnormal lesion from a chest X-ray and computed tomography (CT) scan thorax may bring to the diagnosis of this condition.

Treatment of symptomatic infant with CPAM is early surgical resection of the affected part of the lungs. However, in asymptomatic infant, management is still controversial since either operative and nonoperative approaches to treatment may be employed. Patient with this association should be managed in a neonatal intensive care unit (NICU) with the involvement of multidisciplinary teams such as the paediatric surgeon, neonatologist, radiologist, paediatric anaesthetist and physiotherapist.

Case Presentation

This is a case of premature infant girl delivered by caesarean section at 35 weeks of gestational age due to fetal distress. Prenatally, the mother was diagnosed with gestational diabetes mellitus. Routine prenatal ultrasound of the mother showed no evidence of polyhydramnios or any obvious anomalies. Her birth weight was 1.75 kilograms with an APGAR score of 8 at 1st min and 10 at 5 min. Soon after delivery, she had respiratory distress requiring ventilatory support and subsequently nursed in the NICU. X-ray of the chest showed coiling of the Ryles tube at the mid-thoracic level; thus, the impression of the

Address for correspondence: Dr. Mohd Shahrulsalam Mohd Shah, Paediatrics and Neonatal Surgery Unit, Department of Surgery, School of Medical Sciences, Universiti Sains Malaysia, Kubang Kerian, Kelantan. E-mail: shahrulsalam@usm.my

Received: 16-01-2021 **Revised:** 13-08-2021 **Accepted:** 08-09-2021 **Available Online:** 14-02-2023

Access this article online

Quick Response Code:

Website: https://journals.lww.com/AJPS

DOI: 10.4103/ajps.AJPS_10_21

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How to cite this article: Fijasri NH, Asri NA, Shah MS, Samad MR, Omar N. Type III congenital pulmonary airway malformation associated with oesophageal atresia and tracheoesophageal fistula. A case report and review of literature. Afr J Paediatr Surg 2023;20:245-8.



Figure 1: X-ray chest abdomen. X-ray shows left lung opacity suggestive of congenital pulmonary airway malformation (CPAM)



Figure 2: Coiling of the Ryles Tube can be seen in initial X-ray for oesophageal atresia



Figure 3: Axial view of computed tomography scan thorax. Multiple thin-wall cystic lesion of left lower lobes with adjacent lung parenchymal consolidations more at posterior part suggestive of Type III CPAM by stoker classification

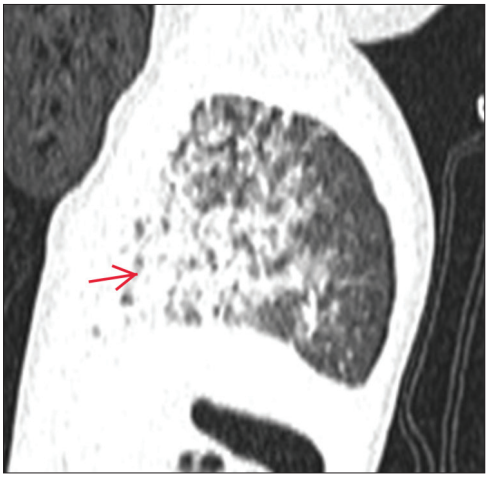


Figure 4: Sagittal view of computed tomography scan thorax. Left lower lobe consolidations with cystic lesions involving both superior and inferior lobes

oesophageal atresia was made. Following that, a Replogle suction catheter was inserted with continuous low pressure suctioning. Apart from that, we noted from the chest X-ray that there were remarkable lung opacities over the left side of the lower hemithorax [refer to Figure 1 and 2]. Due to the suspicion of congenital pulmonary airways malformation (CPAM), CT of the chest was done.

From CT scan thorax, we noted multiple ill-defined thin-walled cystic lesions ranging from 0.2 to 0.5 cm at the left lower lobe with consolidative changes at the adjacent lung parenchyma suggestive of congenital pulmonary airway malformation, Type III according to Stoker classification. Besides, CT scan also showed the coiling of the nasogastric tube at the level of T1/T2 vertebra with abnormal fistulous communication between the distal oesophagus and the trachea at the level of the carina, suggestive of Gross Type C classification of TOF [refer to Figures 3 and 4].

We subjected the patient to TOF ligation and oesophageal anastomosis a day after she was stabilised in NICU. Postoperatively, the patient was ventilated by high frequency oscillatory ventilation for lung recruitment and progressively showed improvement in the next few days. No immediate lung complication such as spontaneous pneumothorax was detected, and subsequently, we were able to wean her off the ventilatory support. Feeding was established successfully without complication, and the child was discharged well on day 24 of life.

Discussion

Oesophageal atresia is a congenital anomaly with an incidence of one in 2500–3000 birth and 90% of cases, and it is associated with TOF.[1] TOF is also commonly associated with one or multiple anomalies involving musculoskeletal, cardiovascular, gastrointestinal, and urogenital systems in 50%–67% of cases.[1] However, the association of TOF with

thoracopulmonary anomaly is rare with an incidence of only 2.1%.[2]

The incidence of congenital pulmonary airway malformation is also rare between 1:11000 and 1:35000 births with males are predominantly affected compared to the female gender.[3] The association of TOF and CPAM has been reported in the literature in very limited numbers. There is a report of TOF with type I CPAM[4] and another two publications of TOF with Type II CPAM.[5] In our case, this is the first reported case of TOF with Type III CPAM due to the findings of solid, homogenous, space-occupying mass of the lesion.

CPAM is a spectrum of a localised lung lesion composed of malformed pulmonary tissue that mimics different anatomic compartments of the lungs.[6] In other words, it is a hamartomatous lesion of the lungs due to anomalous fetal development of the terminal respiratory structures.[6] The morphology of the lesion may predominantly bronchial type epithelium or mesenchyme to predominantly cuboidal or alveolar type.[6]

Historically, CPAM was first described as a separate entity by Chin and Tang in 1949.[7] Then, in 1975, Stoker *et al*. described 38 cases of CPAM and the first to categorise the lesion into three types depending on cyst diameter and predominant cell types.[8] Type I lesion has a large cyst that is more than 2 cm causing mass effect and the cysts are mostly lined by ciliated pseudostratified columnar epithelium. In type II, the lesions are smaller with diameter <1 cm, multiple in numbers, and histologically resembled dilated terminal bronchioles lined by mixed columnar and cuboidal cells. In Type III, the lesion is non-cystic, bulky, forming a solid mass occupying lung lobe causing a mediastinal shift. It has a composition of alveolus-like structures lined by ciliated cuboidal epithelium.[8]

Following this, Stoker expanded their classification into five types based on the site of origin of the malformation, namely tracheal, bronchial, bronchiolar, alveolar ducts and alveolar. Each subgroup is numbered by 0–4 representing the progression of the location of the lesion along the airway.[8] Other classification has been described by ultrasound findings (microcystic or microcytic lesion) and by histopathogenesis (bronchiolar-type epithelial versus acinar alveolar epithelial differentiation).[9] In our case, the pulmonary lesion could be classified as Type III according to Stoker Classification evidenced by CT scan findings of solid, bulky, space-occupying mass in left lung found on the CT scan [refer to Figures 3 and 4].

Recent evidence has looked into a genetic basis associating CPAM and TOF. Some suggest that tracheoesophageal malformation and congenital pulmonary malformation share common embryologic origin thus should not be viewed as a separate entity but as a spectrum of an anomaly.[9] While it is well described in the literature that failure in the septation between the oesophagus and trachea causing TOF, there is evidence that CPAM was due to the anomaly in the early phase of lung development related to bronchial tree branching and differentiation process of surrounding mesenchyme.[8]

At the molecular level, a study has shown that there is an increase in proliferation index that is the Mib-1-labelling index in CPAM lesion compared to adjacent normal lung tissue.[9] There is also the lack of vascularity of the lesion resembling early gestational tissues in CPAM tissue.[8] The genetic basis of CPAM is explained by the findings by Minoo *et al*.[10] demonstrating homozygous deletion of TTF-1 gene (gene-regulating lung development) of a murine model of TOF, causing the arrest of lung embryogenesis at early pseudoglandular stage.[8]

An infant with CPAM may be asymptomatic or may present with tachypnoea with hypoxaemia and carbon dioxide retention, poor feeding and respiratory failure requiring invasive or non-invasive ventilatory support.[3] Imaging studies such as X-ray will confirm the lesion, and CT scan will confirm the size and the presence of mediastinal shift. It is also useful to note that asymptomatic patients may become symptomatic if it is complicated by an intercurrent chest infection and pneumothorax.[3] As in our case, from chest X-ray, we noted a large area of opacity occupying the left lung with mediastinal shift to the right. This lesion is better visualised from the CT, which showed left long consolidations with multiple thin-wall cystic lesions involving both superior and inferior of the left lung [Refer to Figures 3 and 4].

In general, early surgical resection is preferred for symptomatic CPAM patient, especially those with severe impairment on respiratory function. Parenchyma-saving resection is a gold standard to avoid an extensive resection for better long- and short-term result.[11] However, management of an asymptomatic infant with CPAM remains controversial and needs to be tailored by case basis. Some experts argue that leaving the lesion may increase the risk of subsequent malignancy such as bronchoalveolar carcinoma, risk of an enlargement of the lesion inhibiting lung growth and spontaneous pneumothorax.[3] However, in another point of view, a conservative approach may be adopted since there is a likelihood of further regression with increasing age. The complication is not always occurred in asymptomatic patient since only 10% develops chest infection or pneumothorax over 3 years.[3] Furthermore, the risk of malignancy is only in those with bilateral CPAMs who have a genetic susceptibility to CPAMs and malignancy.[3] As in our case, since there is no immediate complication or persistent respiratory compromise, thus we opted for a conservative approach with regular follow-up after successful TOF ligation and repair.

Conclusion

Early identification of associated anomalies in infants with TOF and CPAM should not be overlooked since it may affect the child's direction of treatment and prognosis. We recommend

long-term follow-up until adulthood to look into the natural course and delayed complication of CPAM with TOF.

Financial support and sponsorship

The authors are thankful to the Department of Surgery, School of Medical Sciences, Universiti Sains Malaysia, Kelantan, Malaysia. This paper was supported by the Universiti Sains Malaysia Short-Term Grant (304/PPSP/6315411). We acknowledge the financial support from USM School of Medical Sciences key Performance Index Allocation.

Conflicts of interest

There are no conflicts of interest.

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