Case Report

Single-stage Open Lobectomy and Modified Ravitch Procedure in an Infant with Coexisting Severe Pectus Excavatum and Congenital Cystic Adenomatous Malformation

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Abstract

Coexisting congenital cystic adenomatous malformation of the lungs and severe pectus excavatum (PE) is an uncommon presentation that poses significant management challenges. Conventionally managed in a staged manner, there are increasing reports of superior outcomes with single-staged concurrent repair with minimally invasive techniques (video-assisted thoracoscopic surgery [VATS] and minimally invasive repair of PE [MIRPE]). The outcome of a single-stage open repair for both anomalies has not been previously reported to the best of our search. We report the successful single-stage management of a 9-month-old infant with both anomalies who had an open lobectomy and modified Ravitch procedure. We aim to report the feasibility and safety of a single-stage concurrent repair of both conditions using open techniques, as VATS and MIRPE are not readily available in our environment.

Keywords: Congenital cystic adenomatous malformation, lobectomy, modified Ravitch procedure, pectus excavatum, single stage

Introduction

Severe pectus excavatum (PE) coexisting with congenital cystic adenomatous malformation (CCAM) is relatively uncommon. The concurrent presence of both conditions often poses a significant management challenge for several reasons. They both impair cardiopulmonary function hence these patients are often very ill at presentation. Also, they both require major thoracic surgeries involving significant surgical trauma and prolonged exposure to anaesthesia, especially in infants. Hence, a staged management approach has been traditionally advocated intuitively, with the lobectomy first done, followed by the pectus repair.[1] However, it has been noted that, in the setting of severe PE, there is often no improvement in the respiratory symptoms following lobectomy or correction of other intrathoracic conditions alone as a first-stage procedure, with even reports of worsening of the degree of PE.[2-4] Furthermore, multiple surgical procedures increase the risk of anaesthetic and surgical complications compared to a single-stage procedure.[1,2] Although the optimal approach is still not fully established, there is an increasing number of reports of successful single-stage management of both PE and CCAM, especially with the utilisation of a minimally invasive repair of PE (MIRPE) without increased complications, surgical risk and intraoperative blood loss.[2] The clinical outcome of a single-stage repair of severe PE by a modification of the Ravitch procedure and a concomitant thoracotomy and lobectomy for CCAM is not widely reported. This article aims to report these relatively uncommon coexisting conditions and to share our initial experience with this surgical technique, in which a modification of the Ratvich procedure without sternal osteotomy, using folded polypropylene mesh in place of a pectus bar, and a right upper lobectomy was done in a single stage in an infant with a successful outcome.

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Figure 1: (a) Pre-operative pectus excavatum deformity. (b) Four weeks post-operative outcome

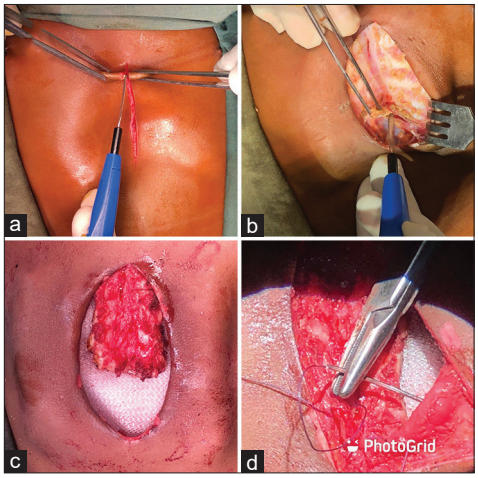


Figure 3: Intraoperative images. (a) Midline longitudinal skin incision. (b) Mobilisation and division of the xiphoid process. (c) Placement of the polypropylene mesh used as sternal support. (d) Layered closure

Case Report

A 9-month-old female infant was referred to us on account of anterior chest wall deformity since birth and recurrent respiratory tract infection that had necessitated four hospital admissions in the preceding 6 months. She also had failure to thrive. There was no clinical feature of any associated cardiac anomaly or syndromes. Examination revealed an underweight (66% expected weight for age), dyspnoeic and tachypneic infant. There was a funnel-shaped deformity of the anterior chest involving the lower sternum and the adjoining costal cartilages and markedly reduced breath sound in the right lower lung zone [Figure 1a]. Chest Computed tomography scan showed features of congenital cystic adenomatous malformation Type I and PE with Haller's index of 3.48. A shown in Figure 2a and b the full blood count revealed a leucocytosis of 17.0 × 10^3/µL, haemoglobin 15.1 g/dL, platelet 347 × 10^3/µL and genotype AA. She subsequently had an open right upper lobectomy through a standard

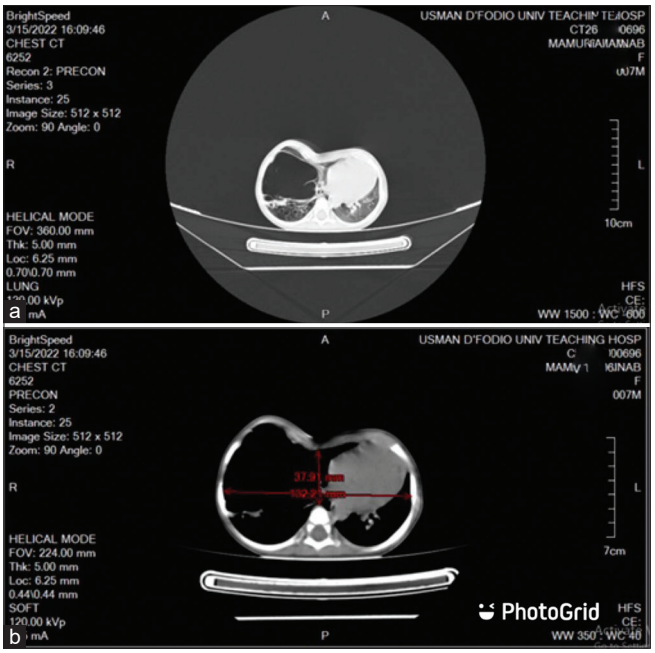


Figure 2: Chest Computed tomography scan of the patient. (a) Areas of cystic adenomatous Malformation. (b) Severity of pectus excavatum deformity and Haller's index



Figure 4: Three months post-operative outcome

posterior thoracotomy and a modified Ravitch procedure. Under general anaesthesia, she was initially placed in the left lateral decubitus position and had a right upper lobectomy in a standard fashion, following which the chest was closed over a chest tube drain with an underwater seal. She was then placed in the supine position and had the modified Ravitch procedure. Intraoperatively, a longitudinal skin incision was made and deepened down to the sternum [Figure 3a]. The xiphoid process was mobilised, separated from the transversus abdominis muscles and excised [Figure 3b]. Pectoral muscle flaps were raised bilaterally, exposing the deformed costal cartilages. Then, subperichondrial/subperiosteal excision of the affected costal cartilages and ribs was done. Due to the pliable nature of the chest wall in this age group, and perhaps,

a less severe form of deformity in this patient, a sterna wedge osteotomy was adjudged unnecessary after the resection of the affected costal cartilages. Rather, a multilayered folded polypropylene mesh was passed retrosternal and anterior to the pericardium and pleura [Figure 3c]. This was used as posterior support to stabilise the sternum and was tautly anchored to the adjoining distal segments of the ribs bilaterally using polypropylene suture, followed by a layered closure of the overlying pectoralis muscles, subcutaneous tissue and skin. She had a left chest tube drain due to a left pleural breach [Figure 3d]. The total operation time was 213 min. Meticulous haemostasis was ensured, and there was no need for blood transfusion. The post-operative course was uneventful, with good analgesic and antibiotic coverage. There was no need for mechanical ventilatory support and was discharged from the intensive care on the 2nd-day post-operative and home on the 10th day. Outpatient follow-up over the last 4 months revealed a complete resolution of symptoms and the mother expressed a satisfactory cosmetic outcome [Figure 1b and Figure 4].

Discussion

The concurrent surgical management of PE and CCAM with a good outcome was first reported by Metzelder *et al*. in 2007.[5] Subsequently, there have been other reported successful outcomes.[2,6] In these series, the roles of minimally invasive approaches to both conditions were emphasised, with most patients having MIRPE with either video-assisted thoracoscopic surgery (VATS) for lobectomy or other thoracic conditions, to minimise the extent of surgical trauma and post-operative complications.[2] In resource-constrained settings like ours, the limited availability of VATS and appropriate sternal support implants, especially for infants and young children, makes concurrent repairs previously unappealing un contemplated. Although the modified Ravitch procedure has been combined with open repair of complex cardiac conditions with excellent outcomes,[7] there are no reports with concomitant open lung resections for CCAM to the best of our search. The use of the 'hammock support' technique in the Ravitch procedure, which has been well described and established to have excellent outcomes, was a suitable choice of technique considering our unique resource constraints.[8,9] Furthermore, a recent meta-analysis comparing both MIRPE and the modified Ravitch procedure showed both have comparable outcomes and complication rates.[10] Although associated with more surgical trauma than MIRPE and VATS, this case report suggests that the single-stage approach of open lobectomy for CCAM and this pectus repair technique can be safely utilised in well-selected patients, even in very young age groups, especially in lower resource settings where the facility and hardware for MIRPE may not be available. The duration of surgery and length of stay in the hospital for this index patient was comparable to those reported for single-stage lobectomy and MIRPE.[2] In addition to the reductions in the overall cost of health-care expenses comparing single-stage versus staged procedures, this reported technique of the modified Ravitch procedure has the additional advantage of using readily available, less expensive means of sternal support/stabilisation, compared to titanium plates and screws. It also does not require the future removal of the pectus bar as in MIRPE.

Conclusion

Single-stage modified Ravitch procedure and open lobectomy for severe PE coexisting with CCAM are feasible and can be successfully done in infants. Larger studies are required to better characterise immediate and long-term outcomes.

Informed consent

A written informed consent was obtained from the parent.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understands that name and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

References

1. Kim YT, Kim JS, Park JD, Kang CH, Sung SW, Kim JH. Treatment of congenital cystic adenomatoid malformation-does resection in the early postnatal period increase surgical risk? Eur J Cardiothorac Surg 2005;27:658-61.
2. Wang H, Wang FH, Liang JH, Liu W, Yang JZ, Guo XH, *et al.* Pectus excavatum and congenital cystic lung lesion: Simultaneous surgery advocated. J Thorac Dis 2018;10:6230-7.
3. Shamberger RC, Welch KJ, Castaneda AR, Keane JF, Fyler DC. Anterior chest wall deformities and congenital heart disease. J Thorac Cardiovasc Surg 1988;96:427-32.
4. Miller DR, Pugh DM. Repair of ascending aortic aneurysm and aortic regurgitation complicated by acute cardiac compression by pectus excavatum in Marfan's syndrome. J Thorac Cardiovasc Surg 1970;59:673-84.
5. Metzelder ML, Ure BM, Leonhardt J, Grigull L, Khelif K, Petersen C. Impact of concomitant thoracic interventions on feasibility of Nuss procedure. J Pediatr Surg 2007;42:1853-9.
6. Bilgi Z, Ermerak NO, Bostancı K, Saçak B, Batırel HF, Yüksel M. Feasibility and complications in concomitant lung resection with minimally invasive repair of pectus excavatum. Ann Thorac Surg 2015;100:707-9.
7. Zaki AL, Vargo PR, Schraufnagel DP, Kalahasti V, Murthy S, Roselli EE, *et al.* Modified ravitch procedure for pectus excavatum combined with complex cardiac surgery. Semin Thorac Cardiovasc Surg 2021;33:1146-53.
8. Robicsek F. Marlex mesh support for the correction of very severe and recurrent pectus excavatum. Ann Thorac Surg 1978;26:80-3.
9. Karagounis VA, Wasnick J, Gold JP. An innovative single-stage repair of severe asymmetric pectus excavatum defects using substernal mesh bands. Ann Thorac Surg 2004;78:e19-21.
10. Jawitz OK, Raman V, Thibault D, Yerokun B, Zwischenberger BA, Kosinski AS, *et al.* Complications after ravitch versus nuss repair of pectus excavatum: A Society of Thoracic Surgeons (STS) general thoracic surgery database analysis. Surgery 2021;169:1493-9.