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Shell-Like Thorax with Intercostal Space Ossification Complicating a Child with Empyema Thoracics and Underlying Rhizomelic Chondrodysplasia Punctata

See WS[1*], S H Syed Rasul[1], J V Simon[1]
Cardiothoracic Department, Hospital Sultanah Aminah, Johor Bahru, Johor, Malaysia

***Corresponding author**
See WS

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Abstract: Rhizomelic chondrodysplasia punctata (RCDP) is very rare. It is an autosomal recessive metabolic disorder which is associated with multiple orthopaedic disorders due to its related chondrodysplasia and abnormal calcification. Here, we report a child with RCDP presented with empyema thoracis. A thoracotomy was attempted for drainage and decortication. However, it failed due to the ossified intercostal spaces. The fixed right shell was inpenetrable. The procedure was abandoned in view of the risk of destruction to the chest wall from excessive traction and to avoid excessive bleeding. The clinical presentations, anaesthetic and surgical challenges will be discussed. To date, there is no similar case reported.

Keywords: Intercostal Space Ossification, Thoracic Empyema, Rhizomelic Chondrodysplasia Punctata, RCDP.

INTRODUCTION

Rhizomelic chondrodysplasia punctata (RCDP) is very rare. It is an autosomal recessive metabolic disorder. There is a deficiency of plasmalogens and deficient activity of the peroxisomal enzyme acyl-CoA dihydroxy-acetone-phosphate acyltransferase (DHAP-AT) [1]. This resulted in low level of plasmalogen and accumulation of phytanic acid [2].

The clinical features include symmetric rhizomelic shortening of limbs, dwarfism, foot deformities, bowing of proximal limbs, flat face, microcephaly, micrognathia, cleft palate, ichthyosis, congenital heart disease, seizures, repeated respiratory infections, congenital cataracts, deafness, and joint contracture [3, 4]. Due to skeletal malformation, multiple orthopaedic problems such as cervical spine stenosis, long bone epiphyses calcification and shortening are reported in RCDP [5-7]. Here, we report a child with RCDP presented with empyema thoracis. A thoracotomy was attempted for drainage and decortication. However, it failed due to the ossified intercostal spaces. The clinical presentations, anaesthetic and surgical challenges will be discussed. To date, there is no similar case reported.

CASE PRESENTATION

This is a 2 years old girl (weighed 4.8kg) presented at the age of 4 months with shortening of upper limbs, scissoring of lower limbs, fixed flexion deformity of knees, hips joint contractures. Skeletal surveys showed shortening of humerus and femur, punctate calcification over shoulders joints, hips and knees. There were metaphyseal flaring of the distal humerus and femur and proximal tibia. Coronal clefting were identified over the vertebral bodies. At the age of 1 year, she had lensectomy done for bilateral congenital cataract.

Currently, she presented with fever and rapid breathing for 1 day. On presentation, she was in respiratory distress with grunting dependent on high flow oxygen. Chest x-ray showed a right pleural effusion with collapse consolidation of right lung (Figure 1).

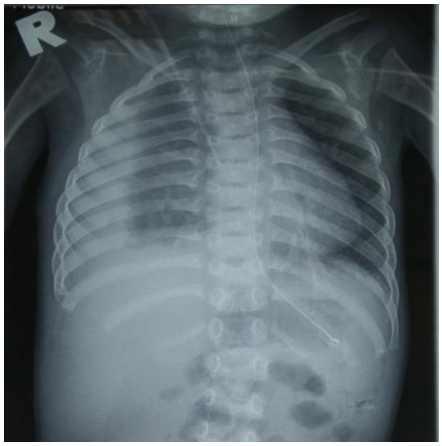

Fig-1: Chest x-ray on presentation

She was ventilated in high dependency ward and started on intravenous ceftriaxone. Blood culture grew streptococcus pneumonia. An ultrasound chest showed grade 3 empyema thoracis. Right chest tube was inserted 4 days following presentation. It drained total of 350 hemoserous fluid over a period of 5 days.

Echocardiography showed global pericardial effusion with maximum diameter of 9.2mm. Computed tomography of thorax done 3 weeks from presentation reported a right pleural collection with highest density at lower zone (34-62 Hounsfield Units). The surrounding pleural is thickened and enhanced. Right lower lobe collapse and consolidation were identified with air bronchograms (Figure 2).

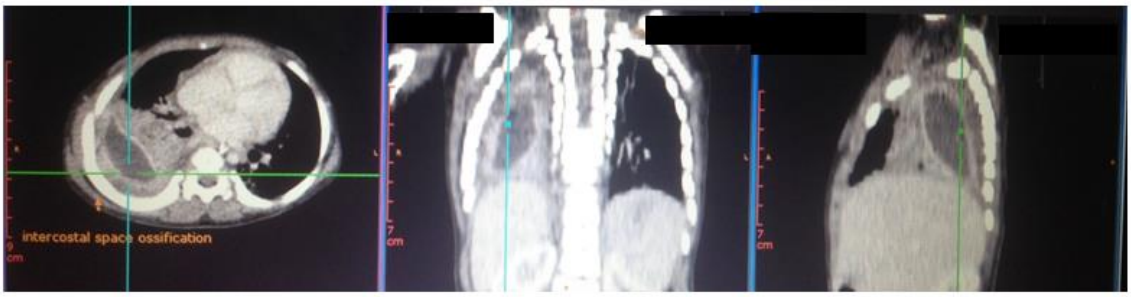

Fig-2: Preoperative CT Thorax which showed a large right pleural collection with collapse right lung

Three weeks following a course of antibiotic therapy and chest tube drainage, she improved and wean off from oxygen. A right posterolateral thoracotomy was performed due to complex right pleural effusion with trapped right lung. Due to the stiff neck, intubation with laryngoscopy was with great difficulty.

During the thoracotomy, we discovered that intercostal spaces were ossified. The rib spaces were inpenetrable either with diathermy or mosquito artery holder from 2nd till 6th intercostal spaces. (Figure 3) It was not reported in the CT scan previously.

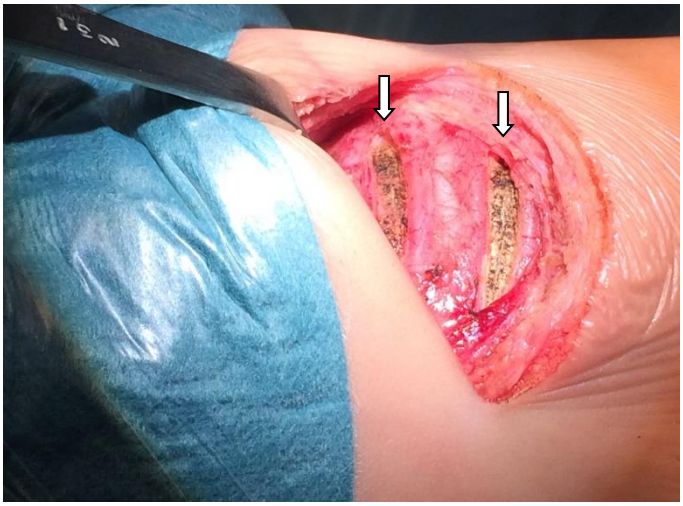

Fig-3: Intraoperative picture of right thoracotomy. 2 intercostal spaces shown were charred following failed attempt of entry (shown by white arrow)

Justifying the risk of excessive bleeding and destruction of right chest wall bony framework in a young child, right pleural space was not explored. The procedure was abandoned. Patient was extubated after surgery and progressed well with antibiotic treatment. She was discharged home two weeks after surgery and planned for surgery if symptoms recur.

DISCUSSION

Child with RCDP presents a challenge to both anaesthetist and surgeons when present with a surgical problem such as empyema thoracic with trapped lung. The association of cervical spine stenosis, joint stiffness or contracture pose a difficult manoeuvrability of the airway [2]. Preoperative anaesthetic evaluation in current patient identify the features of difficult airway such as stiff neck and limited mouth opening. Instruments such as glide scope, video assisted laryngoscopy were standby. A secondary plan of tracheostomy was informed to the parents. Fortunately, the experienced anaesthetist was able to intubate the child with deep cricoid pressure using laryngoscopy despite a finding of Cormack-Lehane 3, avoided the extra procedures.

The chest wall with fixed right shell in current case is the key issue to be highlighted. It has never been reported previously. It made the pleural cavity inaccessible for a proper evacuation of infective debris and decortication. (Figure 4) Attempts to enter the pleural cavity were fraught with bleeding and unnecessary trauma to the chest wall bony framework in a young child. Hence, the thoracotomy was abandoned. The child was treated conservatively at this moment with close monitoring.

The ossified intercostal spaces were found incidentally during surgery, which was not reported in the pre-operative CT scan. Retrospective review of the pre-operative CT thorax with 3D reconstruction revealed a shell-like ossified right intercostal space. (Figure 4) In this case, we would like to highlight the risk of failed thoracotomy in patients with rhizomelic chondrodrodysplasia punctata. Up to date, we were unable to explain the mechanism of the intercostal ossification.

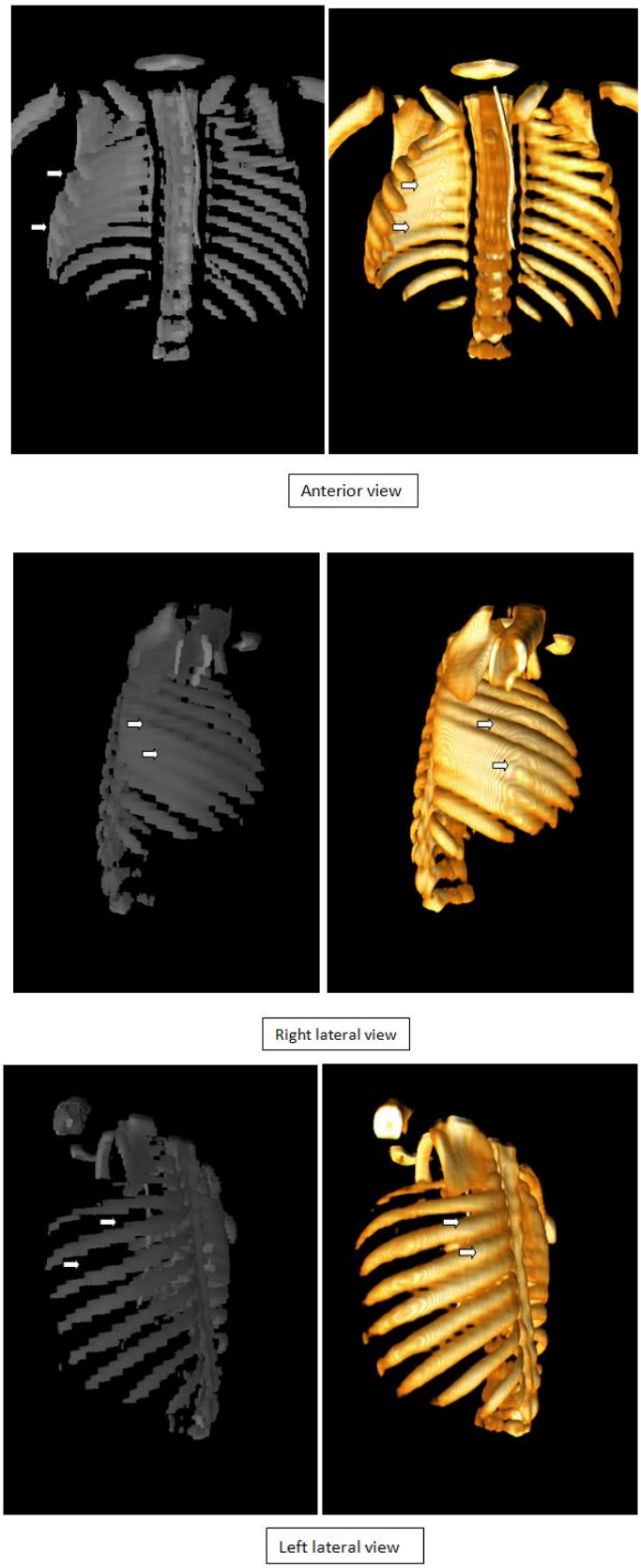

Fig-4: 3D reconstruction of the images from CT thorax showed the absence of intercostal spaces (white arrow) due to intercostal ossification over the right thoracic cage, in comparison with the normal left thoracic cage

One of the perpetuating factors may be due to the crowding of thickened ribs at the affected side, which subsequently lead to the fusion of the costa causing the shell-like feature of the thorax [9]. The underlying inflammatory process is the leading cause for this calcification process.

CONCLUSION

In a child with Rhizomelic Chrondrodysplasia Punctata, empyema thoracis may complicate with an ossification of intercostal spaces resulting in a rigid shell like chest wall, which is a great challenge for both the anesthetists and cardiothoracic surgeons for surgical intervention.

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