Rare disease

Total pleurectomy as the surgical treatment for recurrent secondary spontaneous pneumothorax in a child with severe pulmonary Langerhans cells histiocytosis

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Summary

Pulmonary Langerhans cell histiocytosis (LCH) in children is more extensive and is a rare cause of spontaneous secondary pneumothorax (SSP) which tends to be recurrent and refractory to conventional treatment. Its occurrence in paediatric patients posed great challenge to the choice of surgical management. Surgery in the form of pleurodesis is only considered if SSP does not improve after chemotherapy and after considering all relevant risk and benefits of surgery to patients. Chemical pleurodesis will not give the expected effect to eradicate SSP in this patient. Therefore mechanical pleurodesis is the treatment of choice. There are various techniques to perform mechanical pleurodesis; from pleural abrasion to pleurectomy. In the authors' experience, bilateral total pleurectomy provided the best outcome for this 9-year-old patient with persistent respiratory distress from SSP due to extensive pulmonary LCH.

BACKGROUND

The authors would like to share their experience at managing this difficult case of a 9-year-old child with persistent respiratory distress due to secondary spontaneous pneumothorax from extensive bilateral pulmonary Langerhans cells histiocytosis (LCH) refractory to chemotherapy. A lot of input from different field of expertise including oncology, pulmonology, intensive care and surgery both locally and internationally was sought before surgical procedure was done on this child. The surgery that was carried out that is, bilateral total pleurectomy was indeed an aggressive surgery but with very good outcome. The authors would like to highlight that multi-disciplinary discussion, in this case via e-discussion across the continents was extremely useful and helpful in this case. Surgical option was the last resort. They would like to share the dilemma at managing this case for benefit of clinicians with similar case in the future.

CASE PRESENTATION

The authors report a case of malignant LCH with severe bilateral pulmonary involvement in a 9-year-old boy. The boy initially presented with 3 months history of lethargy, loss of appetite, loss of weight, gum swelling and intermittent pruritic skin lesions on the trunk, back and extremities. The skin lesions were described as initially pruritic and erythematous, subsequently transformed to hypopigmentation and scaly. On the day of admission, he suddenly had respiratory distress. He was severely tachypnoeic with subcostal, suprasternal and intercostals recessions. Examination of the chest revealed unilateral hyperinflation and marked reduced breath sounds on the left side. Clinical diagnosis of left pneumothorax was made. Left

chest tube insertion was immediately performed. At day 4 of admission, he developed a right-sided pneumothorax, a right chest tube was inserted.

Unfortunately, while waiting for diagnosis to be made he developed another episode of left pneumothorax even when the earlier tube was still bubbling. A second chest tube was inserted into the left thorax. Both tubes functioned for a while and provided short relief. Extensive blood and radiological investigations including bone marrow aspiration and tissue biopsy were carried out. Chest x-ray (figure 1) and CT scan of the thorax (figure 2) confirmed bilateral extensive cystic and bullae lesions. The right lung appeared worse.

Diagnosis of LCH was made from histopathological examination of a tissue from his gum. He was started on chemotherapy immediately. During the first 6 weeks on chemotherapy, another chest tube on the right side had to be inserted. All four chest tubes continued to bubble continuously. The patient would be in immediate respiratory distress even when one tube was clamped. E-discussion with several paediatric oncologists and cardiothoracic surgeons from different part of the world through an international-online-working group, including experts from UK and North America advised that mechanical pleurodesis be considered if patient's respiratory condition did not get better or worsened after a second course of chemotherapy. Surgical consult was finally made following failed conservative treatment with specific request for mechanical pleurodesis.

The patient underwent two open surgeries. Minimally invasive technique was not considered in this case. The first was bilateral thoracotomy with right partial pleurectomy and left thoracic pleural abrasion using diathermy-scraper-pad. The patient was symptom-free for only 3 weeks

Figure 1 Chest x-ray of patient with pulmonary langerhans cells histiocytosis; note the bizarre honeycomb appearance brought by bilateral diffuse cystic changes on the lungs with left-sided pneumothorax.

Figure 2 CT thorax of pulmonary langerhans cells histiocytosis presurgery. Note the extensive bullae with compressed lung tissues and evidence of pneumothorax.

before a huge pneumothorax on the left side and a small pneumothorax on the right recurred. A second surgery was then performed; this time bilateral thoracotomy with right completion pleurectomy and left total pleurectomy were carried out. The patient's condition improved. There was no further episode of symptomatic pneumothorax since then although a minimal residual pneumothorax was seen on postoperative CT scan (figure 3). Six months after surgery another CT scan was carried out; patient's bilateral lung tissues had expanded further (figure 4). He continued to be on oncology follow-up with no more respiratory issues.

INVESTIGATIONS

At presentation, patient full blood count was normal that is, white cell count: $10.8 \times 10^9/l$, haemoglobin 10.4 g/dl, platelets $585 \times 10^9/l$. C-reactive protein was minimally raised that is, 1.51 mg/dl. Erythrocyte sedimentation rate was raised at 83 mm/h. Chest x-ray revealed bilateral diffuse cystic changes on the lungs with left sided pneumothorax. Investigations for pulmonary tuberculosis and atypical infection were negative. High resolution CT thorax showed multiple diffuse, irregular-shaped cystic lesions of varying sizes in both lung fields, with thickened

Figure 3 CT thorax of pulmonary langerhans cells histiocytosis post pleurectomy. Note both lung tissues have expanded. A small residual pneumothorax is seen on the apical left zone (patient remained asymptomatic despite this finding).

Figure 4 CT thorax of the same patient at 6 months post surgery. Note both lung tissues have expanded further.

bronchial walls, suggestive of pulmonary Langerhans cell histiocytosis (LCH). Investigations to look for extension of the LCH were also carried out; skeletal survey revealed no evidence of lytic lesion or bony involvement. Bone marrow aspiration and trephine biopsy revealed no evidence of LCH. Biopsy of the gum confirmed presence of LCH which was positive for S-100 protein.

TREATMENT

The patient underwent two surgeries. Surgery was only considered after the patient had undergone chemotherapy regime as per LCH III UK protocol group 1. Despite being on therapy for over 12 weeks, there was no improvement with his respiratory condition. A repeat CT thorax after 6 weeks of medical treatment showed worsening of both

Figure 5 Appearance of right lung langerhans cells histiocytosis at surgery.

Figure 6 Appearance of left lung langerhans cells histiocytosis at surgery. Note lesions in the left lung is less severe than right.

lung tissues and persistent pneumothorax despite multiple insitu chest tubes. The international work-group's advice for mechanical pleurodesis included discussion on whether the procedure would interfere with lung transplantation in the future. Experienced adult lung transplant surgeons both locally and internationally were of the opinion that mechanical pleurodesis would not be a contra-indication for future lung transplant.

For this patient, each surgery involved bilateral thoracotomy and mechanical pleurodesis. During the first surgery, as the right lung looked worse (figure 5), the author decided to do partial pleurectomy; and for the left lung which looked less severe (figure 6); the author opted to do pleural abrasion using diathermy- scraper-pad-which is the commonest method of mechanical pleurodesis performed. Three weeks later, the spontaneous secondary

pneumothorax (SSP) recurred on both sides, this time the SSP on the left lung was worse. The author had to do a second surgery through the same wounds that is, completion pleurectomy for the right side and total pleurectomy for the left side. The patient continues to receive chemotherapy for pulmonary LCH and repeated admission into intensive care unit during the course of treatment.

OUTCOME AND FOLLOW-UP

The patient had the best outcome after the second surgery. After bilateral total pleurectomy, the lung expanded better than before and the patient no longer had SSP although the post surgery CT scan showed minimal residual pneumothorax on the left side (figure 5). Six months post surgery another CT thorax revealed a better picture that is, bilateral expanded lung tissues (figure 6). The child remained asymptomatic.

DISCUSSION

Pulmonary LCH (PLCH) can occur in isolation or as part of multisystem disease manifestation.^{1 2} The definition of PLCH is often associated with cigarette smoking in adults whereas in children there is no association at all.³ The lung in adults is the most commonly and solely affected organ, whereas in children, a single organ involvement is extremely rare.^{1 4} It has been reported that in children, when lung is involved, it is severe and even lethal and usually a manifestation of multisystem disease.^{3 4}

Histologically, both paediatric and adult populations with LCH have identical manifestations.^{5 6} However imaging studies have found differences in high resolution CT findings between them. In adults, the bilateral nodules and thin- or thick-walled cysts typically involved upper and middle lung zones with sparing of the costophrenic angles.^{7 8} The cysts, fibrosis and nodules that are spared at the sub pleural parenchyma and costophrenic recess in adults are however not spared in the paediatric population.⁹ In children with multisystem disease, the 5 year survival rate is around 50% despite aggressive chemotherapy.¹⁰

The main complication of pulmonary LCH is the occurrence of SSP with subsequent recurrence and persistence despite conservative management.¹¹ Patients usually present to the emergency department with shortness of breath associated with pleuritic chest pain of sudden onset.

Patients with SSP due to PLCH tend to be quite refractory to conventional treatment due to underlying fibrotic lungs which are more fragile and tend to collapse easily. This poses a great challenge to surgeons with regard to the best choice of surgical management and technique. Based on the author's experience, in extensive pulmonary LCH, partial pleurectomy or pleural abrasion was not sufficient to eradicate SSP fully. Could the authors have avoided the second surgery if they had waited longer? As there was not enough clinical experience elsewhere, the threshold to perform re-surgery was lower, especially when the patient was still symptomatic. In this patient total pleurectomy proved to be a successful treatment. The latest CT thorax

(figure 6) confirmed success of this procedure. The authors recommend, for any child with extensive lung disease, such as PLCH presenting with SSP, when surgery becomes the only choice, total pleurectomy should be considered as an option.

Learning points

- ▶ Surgical referral for SSP in children is not common. Chemical pleurodesis is the method of choice for SSP from other lung pathology but not for extensive pulmonary LCH. Mechanical pleurodesis is superior to chemical pleurodesis as the treatment of choice for SSP in a child with extensive pulmonary LCH.
- ▶ Surgical referral must only be considered after thorough discussion with colleagues in relevant specialties. E-discussion proved to be very beneficial in managing rare cases.
- ▶ Partial pleurectomy or pleural abrasion may not be sufficient to eradicate SSP in extensive pulmonary LCH.
- ▶ Bilateral total pleurectomy was a successful treatment for this child with extensive pulmonary LCH and persistent SSP.

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Competing interests None.

Patient consent Obtained.

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