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TOTAL PLEURECTOMY: THE MECHANICAL PLEURODESION OF CHOICE FOR RECURRENT SECONDARY SPONTANEOUS PNEUMOTHORAX IN A CHILD WITH PULMONARY LANGERHANS CELL HISTIOCYTOSIS; A CASE REPORT

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Background:
Pulmonary Langerhans Cell Histiocytosis (LCH) is a rare entity, characterized by accumulation of Langerhans cells (LC) infiltrating the interstitium of small airways of the lung leading to chronic progressive interstitial lung disease. Patients may present with spontaneous secondary pneumothorax (SSP) which tends to be recurrent and refractory to conventional treatment. In children, the disease is more extensive and will progress despite aggressive chemotherapy with 50% survival rate at 5 years. Pulmonary LCH is a rare cause of SSP and its occurrence in pediatric patients posed great challenge to the choice of surgical management.

Case history:
We report a case of malignant LCH with severe bilateral pulmonary involvement in a 9 year old boy. The boy presented with bilateral recurrent spontaneous pneumothorax which initially responded to multiple thoracic drain insertions. Surgical consult was made following failed conservative treatment with specific request for mechanical pleurodesis. Computed tomography (CT) scan of the thorax confirmed bilateral extensive cystic and bullae lesions. The right lung appeared worse. The patient underwent 2 surgeries. The first was bilateral thoracotomy with partial right pleurectomy and left thoracic pleural abrasion. The patient was symptom-free for only 3 weeks before a huge pneumothorax on the left lung 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 pneumothorax since then.

Discussion and conclusion:
The surgical experience of managing SSP is usually from adult patients with chronic obstructive pulmonary disease (COPD). These patients usually undergo mechanical pleurodesis via pleural abrasions either thoracoscopically or open thoracotomy. Based on our experience, in pulmonary LCH, partial pleurectomy or pleural abrasion are not sufficient to eradicate SSP. We recommend total pleurectomy as the technique of choice for mechanical pleurodesis in patients with extensive pulmonary disease like Pulmonary LCH.

Keywords:
Pulmonary LCH, secondary spontaneous pneumothorax, mechanical pleurodesis, total pleurectomy, children.