POSTER PRESENTATIONS

P11

LANGERHANS CELL HISTIOCYTOSIS – A RARE CASE OF SEVERE PULMONARY INVOLVEMENT

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Background: Langerhans Cell Histiocytosis (LCH) is a dendritic cell disorder, resulting from proliferation of immunophenotypically and functionally immature Langerhans cells with eosinophils, macrophages, lymphocytes and multinucleated giant cells. The incidence of LCH is 2 to 10 cases per million children aged less than 15 years old. Pulmonary LCH is rare in childhood. It typically affects young adults and associated with cigarette smoking. We describe a case of LCH with severe pulmonary involvement.

Case report: A 9-year-old, Chinese, boy, with 3-month history of constitutional symptoms and gum swelling presented with sudden onset of left sided chest pain and difficulty in breathing. His chest X-ray showed bilateral diffuse cystic lung changes and pneumothoraces. His HRCT thorax revealed multiple diffuse, irregular-shaped cystic lesions of varying sizes in both lung fields suggestive of pulmonary LCH. Biopsy of the gum confirmed Langerhans cell histiocytosis. He developed diabetes insipidus secondary to posterior pituitary infiltration, confirmed by MRI pituitary and was started on oral Minirin. He subsequently was started on multisystem risk regime as per LCH III UK Protocol. He received 4 weeks of Prednisolone (60mg/m²/daily) and Vinblastin bolus (6 mg/m²) one week a part, 6 doses in total. It was a challenge for us to manage the recurrent tension pneumothorax which required chest tubes in-situ for prolong period of time. In view of this, he underwent bilateral thoracotomy and mechanical pleurodesis.

Conclusion: We presented a case of severe pulmonary LCH with challenge in managing his recurrent bilateral pneumothoraces.