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Case Report

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Langerhans cell histiocytosis (LCH) presenting as pneumothorax and neck lymphadenopathy

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Abstract

Langerhans Cell Histiocytosis (LCH) is a rare clonal disease characterized by dendritic cells infiltration in various main organs. We report a 6-month-old infant with severe respiratory distress, cervical lymphadenopathy, and reddish skin rashes on face. Diagnostic work up revealed spontaneous pneumothorax on Chest X-ray (CXR). Multiple thin-walled lung cysts and emphysema was discovered on spiral chest CT scan. Lung tissue and cervical lymph node biopsy revealed dendritic cells infiltration. Broncho alveolar lavage (BAL), cervical lymph node and lung tissue biopsy was performed and histopathology was assessed. It was suggestive of LCH with positive immunohistochemistry markers including CD1a, CD68 and S100.

Introduction

LCH is a rare clonal disease that could occur at any age. The most common age distribution of LCH is between one to four years of age. LCH can involve various organs such as skin, bones, lymph nodes, lungs, liver and other organs and systems. The clinical course of disease may present as a self-limited condition to a rapidly progressive and life- threatening disease. LCH in different organs could have various manifestations. Vesicles and bullae on skin (most commonly in early infancy), bone involvement presenting as lytic lesions of temporal and orbital bones or long bones, cavitated pulmonary nodules, jaundice and diabetes incipitus are some manifestations of LCH.

The diagnosis of LCH is based on clinic pathologic and immunophenotipic study of lesions in involved tissues based on histological criteria by the histiocyte society in 1987 [1, 2]. In recent years, LCH is classified in to three categories: single-system, single-site (SS-s), single-system, multi-sites (SS-m) and multi system (MS) types [3]. The most common category is SS with prevalence of roughly 65% [4-7].

The incidence of LCH in children is declared approximately 2-9 cases in one million births per one year. LCH is more prevalent in male gender with male to female ration of 1.2 to 1.4 [7-9]. We came across a case of LCH with bilateral cervical lymphadenopathy, severe respiratory distress and skin rashes on face in a six month-old infant.

Case History

We report a previously healthy 6-month-old male infant that was hospitalized with four days of fever, cough and progressive respiratory distress with the primary diagnosis of pneumonia. He was hemodynamically stable, but tachypnea and tachycardia with decreased O_2 saturation (Spo_2 more than 94%) was reported at the time of admission. He was admitted in Paediatric Intensive care Unit (PICU) due to progressive and severe respiratory distress.

Pneumothorax was discovered in chest radiography (CXR) and multiple bilateral thin walled lung cysts and unilateral pneumothorax was observed in high resolution CT scan (HRCT). On physical examination, bilateral cervical lymphadenopathy (without hepatosplenomegaly and generalized lymphadenopathy) and reddish skin rashes on face mostly on forehead were detected simultaneously.

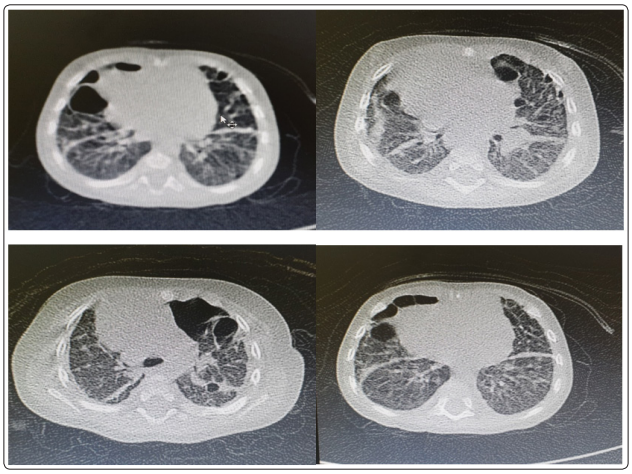

Image 1: pneumothorax and multiple bilateral thin walled lung cysts were observed in high resolution CT scan (HRCT)

He was intubated due to deterioration of respiratory distress and decreased Spo_2. Chest tube was inserted by paediatric surgeon in PICU. He underwent thoracotomy surgery in order to resect the involved lobe of lung because of persistent air-leak in chest tube. He was extubated during the second week. But, following the third week of admission, he again developed pneumothorax in the contralateral lung.

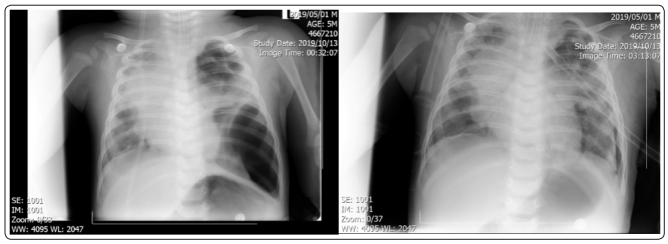

Image 2: Pneumothorax was discovered in chest radiography(CXR). Chest tube was inserted by paediatric surgeon

Biopsy was performed to confirm the diagnosis by evaluation of cells histopathology. Broncho alveolar lavage (BAL), cervical lymph node and lung tissue biopsy was performed and histopathology was assessed. It was suggestive of LCH with positive immunohistochemistry markers including CD1a, CD68 and S100.

Discussion

LCH has a wide spectrum of organ involvement, ranging from a self-limited condition to a rapidly progressive organ involvement resulting in death. LCH could commonly involve skin, bone, lymph nodes, central nervous system (CNS), liver, lungs, thyroid and other organs. Single system disease, multisystem disease, multisystem disease with risk organ involvement are stages of LCH disease. Our case had respiratory system involvement, lymphadenopathy and skin rashes, which could be classified as multisystem disease. Isolated lung involvement in LCH is not considered recently as a risk organ, because it could unlikely results in death exception for complications of mechanical ventilation like uncontrolled pneumothorax, cardiopulmonary arrest and multiple bullae cysts resulting in recurrent pneumothorax. Pneumothorax should be managed by chest tube insertion. Uncontrolled pneumothorax must be treated with pleurodesis [10-12]. Our patient was a future candidate for pleurodesis in case of persistent and recurrent massive pneumothorax [10, 11].

In 2017, Satyaki et al. published two cases of LCH with skin rashes and respiratory distress. The second case has been treating as tuberculosis (TB) since recent months due to bilateral military shadows on CXR suggestive of TB. But, no significant improvement was observed in follow-ups. During further assessment to evaluate his persistent respiratory distress, pneumothorax and subcutaneous emphysema were reported in CXR and physical examination [13]. This case had similar presentation to our case. Respiratory distress and CXR mimicking pneumonia is the usual presentation of LCH in children.

Another case study by Aggarwal et al. in 2010, a 4 month-old infant was introduced with nodular skin rashes and lung infiltration. However, spontaneous regression was considered in this case during follow-ups (congenital self-healing LCH) [14]. A case of LCH was introduced by Pooja Abbey et al. In 2014. A ten month-old boy with multisystem LCH presenting with thin-walled lung cysts and pulmonary interstitial emphysema without pneumothorax [15]. This case was similar to our case report. In our case, bullous lesions expanded to large sizes, merged together and finally caused pneumothorax in bilateral lungs. Madasu et al reported a case of LCH in a 5-month-old female infant with a presentation similar to lymphoma. She was admitted with severe respiratory distress, generalized lymphadenopathy, wide mediastinum and hepatosplenomegaly. She was treated as T-cell lymphoma with no medical response to chemotherapy. Finally, biopsy confirmed the diagnosis of LCH [16]. Lymphadenopathy and respiratory distress in this case was similar to our case.

In another case, report published from Iran LCH was reported in a two-month-old infant with simultaneous bilateral and spontaneous pneumothorax. Due to destructive changes in in lungs, pneumothorax could happen in 10% of LCH cases with respiratory involvement [17]. In 2005, Nakhla et al. reported a previously healthy 16 years old girl with sudden death. She developed sudden shortness of breath and loss of consciousness. Bilateral pneumothorax was discovered in post-mortem CXR. Lung biopsy revealed the diagnosis of LCH. Bilateral emphysematous bullae was discovered throughout the pulmonary tissue on autopsy [18].

Seely JM et al. In 2012 studied a series of seven children and twelve adults with LCH. In all cases, CT findings were similar to each other [19]. According to studies conducted in recent years, children with no suitable response to chemotherapy, involvement of various organs (MS category of LCH), neurodegeneration in CNS and diabetes insipidus have the worst prognosis ever. Recurrence of LCH after induction of treatment has been reported in patients with bone involvement especially in cases with multiple bone lesions [20]. Recurrence is not common in LCH with other organ involvements. In a study in 2017, Asilsoy S et al. concluded that all children with chronic respiratory problems should be evaluated in terms of LCH [21].

References

1. Minkov M, Grois N, McClain K (2009) Langerhans cell histiocy- tosis – Histiocyte Society Evaluation and Treatment Guide- lines, Protocol.
2. Bechan GL, Egeler RM, Arceci RJ (2006) Biology of Langerhans cells and Langerhans cell histiocytosis. Int Rev Cytol 254: 1-43.
3. Haupt R, Minkov M, Astigarraga l, Eva Schäfer, Vasanta Nanduri, et al. (2013) Langerhans cell histiocytosis (LCH): guideline foe diagnosis, clinical work-up, and treatment for patients till the age of 18 years. Pediatr Blood Cancer 60: 175-184.
4. Berry DH, Gresik M, Maybee D, Marcus R (1990) Histiocytosis X in bone only. Med Pediatr Oncol 18: 292-294.
5. Roger G, Dupre M, Leboulanger N, Godey B, Thomas C, et al. (2009) Cholesteatoma secondary to temporal bone involvement by Langerhans cell histiocytosis: A complication amenable to curative surgery. Otol Neurotol 30: 190-193.
6. Ronceray L, Potschger U, Janka G, Gadner H, Minkov M, et al. (2012) Pulmonary involvement in pediatric-onset multisystem langerhans cell histiocytosis: Effect on course and outcome. J Pediatr 161: 129-133.
7. Vassallo R, Ryu JH, Colby TV (2000) Pulmonary Langerhans'-cell histiocytosis. N Engl J Med 342: 1969-1978.
8. Braier J, Latella A, Balancini B, Rosso D, Chantada G, et al. (2004) Outcome in children with pulmonary Langerhans cell histio- cytosis. Pediatr Blood Cancer 43: 765-769.

9. Grois N, Fahrner B, Arceci RJ, Henter JI, McClain K, et al. (2010) Central nervous system disease in Langerhans cell histiocytosis. J Pediatr 156: 873-881.
10. Morren MA, Broecke KV, Vangeeebergen L, Sillevis-Smitt JH, Hauben E, et al. (2016) Diverse cutaneous presentations of Langerhans cell histiocytosis in children: a retrospective cohort study. Pediatr Blood Cancer 63: 486-492.
11. Morimoto A, Ishida Y, Suzuki N, Ohga S, Shioda Y, et al. (2010) Nationwide survey of single-system single site Langerhans histiocytosis in Japan. Pediatr Blood Cancer 54: 98-102.
12. Alston RD, Tatevossian RG, McNally RJ, Kelsey A, Birch JM, et al. (2007) Incidence and survival of childhood Langerhans cell histiocytosis in North- west England from 1954 to 1998. Pediatr Blood Cancer 48: 555-560.
13. Das S, Deepshikha, Giri PP (2017) Skin rash with respiratory distress: A report of two cases. Indian J Paediatr Dermatol 18: 241-244.
14. Aggarwal V, Seth A, Jain M, Krishnamurthy S, Chandra V, et al. (2010) Congenital Langerhans cell histiocytosis with skin and lung involvement: Spontaneous regression. Indian J Pediatr 77: 811-812.
15. Abbey P, Narula MK, Anand R, Chandra J (2014) Persistent pulmonary interstitial emphysema in a case of Langerhans cell histiocytosis. Indian J Radiol Imaging 24: 121-124.
16. Madasu A, Noor Rana A, Banat S, Humad H, Mustafa R, et al. (2015) Langerhans cell histiocytosis in an infant mimicking a lymphoma at presentation. Case Rep Hematol 2015: 670843.
17. Alavi S, Ashena Z, Paydar A, Hemmati N (2007) Langerhans cell histiocytosis manifesting as reccurent simultaneous bilateral spontaneous pneumothorax in early infancy. Pediatr Int 49: 1020-1022.
18. Nakhla H, Jumbelic MI (2005) Sudden death of a patient with pulmonary Langerhans cell histiocytosis. Arch Pathol Lab Med 129: 798-799.
19. Seely JM, Salahudeen S, Cadaval-Goncalves AT, Jamieson DH, Dennie CJ, et al. (2012) Pulmonary langerhans cell histiocytosis: a comparative study of computed tomography in children and adults. J Thorac Imaging 27: 65-70.
20. Jezierska M, Stefanowicz J, Romanowicz G, Kosiak W, Lange M (2018) Langerhans cell histiocytosis in children—a disease with many faces. Recent advances in pathogenesis, diagnostic examinations and treatment. Postepy Dermatol Alergol 35: 6-17.
21. Asilsoy S, Yazici N, Demir S, Erbay A, Koçer E, et al. (2017) A different cause for respiratory disorder in children: cases with Langerhans cell histiocytosis. Clin Respir J 11: 193-199.