A Rare Case of Chylothorax Manifestation of Gorham's Disease, Lymphangioma and Tuberculosis

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Abstract

Background: Chylothorax is a rare condition caused by accumulation of chyle in the pleural cavity (2-3%). More rare causes are Gorham's disease and tuberculosis
Case: A 31-year-old male complained of swelling left arm and shortness of breath with recurrent pleural effusion. Pleural fluid analysis showed chylous. Bone survey showed osteolytic lesions of multiple bones as Gorham's disease. Thoracic CT showed left lung mass, atelectasis, massive fluidothorax, ipsilateral supraclavicular lymphadenopathy, destructive left scapula. Needle aspiration of left humerus revealed lymphangioma. Expert Mtb-Rif examination revealed Mtb detected. We assessed Gorham's disease, lymphangioma and lung tuberculosis with complication recurrent chylothorax and was treated anti-tuberculosis, anti-osteolytic and thorax catheter insertion.
Conclusion: Difficult and rare case of chylothorax in one patient with Gorham's disease and tuberculosis, is a poor prognosis.
Keywords: Chylothorax, Gorham's Disease, Tuberculosis

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1. Introduction

Chylothorax is a rare condition, it represent about 2-3% of all pleural effusions.[1] Gorham's disease (vanishing bone disease, phantom bone disease, or idiopathic massive osteolysis) is a rare bone disorder characterized by progressive osteolysis with lymphatic and vascular proliferation. The incidence is very rare, with only a few hundred case reports described in literature.[2] Lymphangiomas are focal proliferations of well-differentiated lymphatic tissue presenting as multicystic or sponge-like accumulations. It can lead to chylous effusion.[3] Tuberculous chylothorax is a rare infectious disease that occurs when the thoracic duct is obstructed.[4] The case is interesting, it includes very rare cases,

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recurrent massive chylothorax manifestations of Gorham's disease, lymphangioma and pulmonary tuberculosis

2. Case

A 31year old male, complained of swelling left arm and shortness of breath with recurrent massive pleural effusion sinistra. Pleural fluid analysis showed chylothorax (Figure 1). Thoracic CT showed left lung mass, atelectasis, massive fluidothorax, ipsilateral supraclavicular lymphadenopathy, destructive left scapula. Bone survey showed osteolytic lesions of multiple bones as Gorham's disease (Figure 2 A, B, C). Needle aspiration of left humerus benign cyst lesion revealed lymphangioma (Figure. 2 D). Xpert Mtb-Rif examination showed Mtb detected very low. We assessed Gorham's disease, lymphangioma with superimposed infection lung tuberculosis and complication recurrent massive chylothorax sinistra. Patient was treated 1st category anti-tuberculosis, anti-osteolytic agent, pleural fluid evacuation (75 L) and thorax catheter insertion.

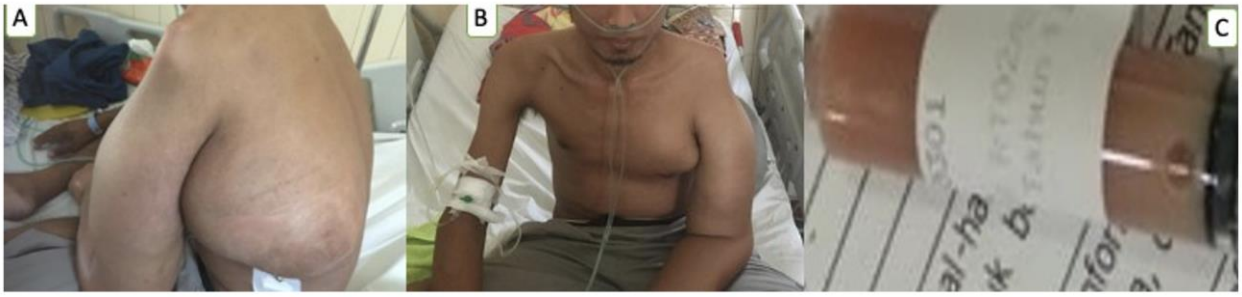

Figure 1. A. B. Clinical presentation show left arm and back edema. C. Chylous pleural fluid with milkyappearance. Pleural fluid analysis showed triglycerides 801 mg/dL and total cholesterol 59 mg/dL.

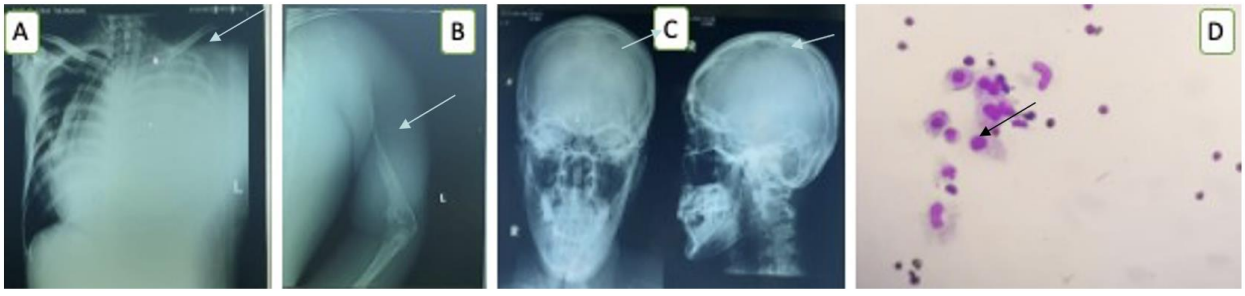

Figure. 2. A Massive pleural effusion sinistra B and C. Multiple bone lytic lesion proximal left humerus and skull D. Hypocellular consisting of the spread of several cyst macrophage, polymorphonuclear and mononuclear Inflammatory cells, aswell as some spindle cells with oval nuclei, fine chromatin, spread between erythrocyte backgrounds.

3. Discussion

Chylothorax is a rare condition caused by obstruction or injury to thoracic duct leading accumulation of chyle in pleural cavity. First line therapy consists of treatment of underlying conditions, pleural drainage to control symptoms and dietary modifications. Therapeutic thoracentesis or indwelling pleural catheter may be used to control shortness of breath during initial treatment. Continuous drainage allows the lung to re-expand and thus optimized pulmonary function.[5]

Gorham's disease is a very rare disorder that characterized by local osseous invasion by angiomatous vascular mass without skip area, eventually causing of the affected bone. Bone scan is considered useful in excluding it. Gorham's disease should be included in differential diagnosis of chylothorax, especially when it is associated with osteolytic lession.[6] Management of Gorham's disease is challenge for clinicians. The medical treatment includes radiation therapy, anti-osteoclastic agent (bisphosphonate) and alpha-2b interferon.[7] However, pulmonary involvement with chylothorax may mean a poor prognosis.[8]

Bone involvement has been described in almost 75% of the patients, being more common of the skull, ribs, pelvis, tibia, humerus dan vertebrae. Lymphangioma is known that with the concomitant involvement of the osseous and thoracic system, the prognosis is usually poor. Lymphangioma is a definitive diagnosis with biopsy and in this case lymphangioma as part of Gorham's disease.[9] In cases of chylothorax associated with tuberculosis; it observed in sputum, fluid or tissues and caseous necrosis are used for diagnosis.[10] In this patient found Mtb detected. In patients with immunodeficiency can be easily infected with Tb with atypical manifestations. Treatment for tuberculosis includes anti-tuberculous therapy.[4] It can be a challenge to diagnose patients with Gorham's disease on the clinical, histological and radiological feature. In clinical practice, Gorham's disease is often a diagnosis by exclusion, after ruling out inflammatory, infectious, metabolic and neoplastic diseases.[2]

4. Conclusion

The case report reported a 31-year-old patient with chylothorax manifestations of Gorham's disease, tuberculosis and lymphangioma. This case is a rare condition. This requires a multidisciplinary discussion for the diagnosis and management of patients. Because of these cases and literacy that are still rare, further research is needed in terms of diagnosis and management of patients with similar complaints.

References

1. Jose M. P. Pulmonary Advisor: Chylothorax and Cholesterol Effusion. 2016.
2. De Keyser CE, Saltzherr MS, Bos EM, Zillikens MC. A large skull defect due to Gorham-Stout disease: case report and literature review on pathogenesis, diagnosis, and treatment. Frontiers in Endocrinology. 2020:37.
3. Riley LE, Ataya A. Clinical approach and review of causes of a chylothorax. Respiratory Medicine. 2019 Oct 1; 157:713.
4. Rodríguez-Hidalgo LA, Concepción-Urteaga LA, Cornejo-Portella JL, Alquizar-Horna ON, Aguilar-Villanueva DA, Concepción-Zavaleta MJ, Ruiz-Caballero DC. A case report of tuberculous chylothorax. Medwave. 2019 Jun 19;19(05).
5. Dhakal P, Bastakoti S, Rayamajhi S. Chylothorax: a review of management options. Clinical Pulmonary Medicine. 2017 Mar 1;24(2):66-8.
6. Lee WS, Kim SH, Kim IH, Kim HK, Lee KS, Lee SY, Heo DS, Jang BS, Bang YJ, Kim NK. Chylothorax in Gorham's disease. Journal of Korean Medical Science. 2002 Dec 1;17(6):826-9.
7. Saify FY, Gosavi SR. Gorham's disease: A diagnostic challenge. Journal of Oral and Maxillofacial Pathology: JOMFP. 2014 Sep;18(3):411.
8. Anomynous. National Center for Advancing Translational Sciences, 2021. Gorham’s disease.
9. Uribe R, Isaza S, Prada V, Cadavid L, Quiceno W. Lymphangiomatosis in a 14-year-old female presenting with chylothorax and multiple cystic lesions. Radiology Case Reports. 2018 Aug 1;13(4):782-7.
10. Cansiz KA, Tug T, Konuk S, Kilicgun HA, Aydin S. A Case with Tumor, Tuberculosis and Chylothorax. Archives of Clinical and Medical Case Reports. 2018;2(3):68-74.