中文題目：古漢氏病以乳糜胸及多發性骨質溶解和脊椎壓迫性骨折表現之病例報告

英文題目：Gorham disease, a case with chlothorax, multiple osteolysis and spine compression fracture

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Introduction

Gorham disease is a rare disease characterized by the replacement of bone with fibrous tissue. It is also known as massive osteolysis of Gorham, vanishing bone disease, phantom bone disease, or Gorham–Stout syndrome. There are limited case reports with chylothorax and Gorham's disease. Herein, we present a case of multiple osteolytic lesions, spine compression fracture and chlothorax.

Case Report

This 44-year-old male presented with lower back pain aggravated for 2 weeks. His past medical history was chylothorax history post pleurodesis 6 years ago, and left hip Fibrous dysplasia. Initial chest X ray showed expansile lesions of the right 2nd and left 5th ribs and right L1 pedicle, suspect osteolytic lesions. Chest and abdominal CT showed expansile osteolytic lesions at ribs, clavicles, thickened of right pleura, pubic bone, and compression fracture of L2. Thoraco-lumbar spine MRI also showed multiple osteolytic lesions with high signal on T2W and low signal on T1W are identified in the visible thoracolumbar spine, bilateral ribs, and pelvic bone. and vertebral compression fracture at L2. The tumor markers were all within normal range. And there was no hypercalcemia, no kappa, or lambda light chain elevation, no β2-Microglobulin elevation, and no monoclonal or oligoclonal gammopathy. Bones scan showed active bone lesions involving mildly in middle portion of right humeral shaft and slightly heterogeneously in lumbar spine, and a hot spot in right temporal skull.

Due to multiple osteolytic lesion at thoracolumbar spine, bilateral ribs, and pelvic bone, Gorham's disease was impressed. The patient suffered from recurrence of bilateral pleural effusion. We suggested to the patient for video assisted thoracoscopic surgery for tissue proof. However, the patient hesitated.

Discussion

To date, the specific etiology of Gorham disease remains unknown. Gorham and Stout hypothesized the local proliferation of endothelial-lined vessels could induce bone loss by increasing vascularity, lowering local pH, local hypoxia or by exerting mechanical force. Lymphatic vessels are primarily affected in Gorham disease. The uncontrolled growth of fluid-filled lymphatic vessels could cause osteolysis by compressing bone, and may cause pathologic fracture. Diffuse infiltration of lymphangiomas in the lung, bone, and other organs may be seen. Within the thorax, lymphangiomas may be found wide spread in the mediastinum, heart, thoracic duct, lung, and pleura or limited in a single organ. Most authors agree that the manifestations of lymphangiomatosis that facilitate its diagnosis are bone lesions in conjunction with chylothorax.