Diffuse Lymphangiomatosis: Gorham-Stout Syndrome

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Gorham-Stout syndrome (GSS) is a very rare disorder of uncertain etiology. GSS presents with lytic destruction of one or more bones and can include chylous effusion in the pleurae, peritenoum and pericardium and cystic lesions in visceral organs, such as the spleen. A 32-year-old woman was admitted to the emergency department suffering from dyspnea and orthopnea. A chest roentgenogram revealed bilateral pleural effusion (Picture A). Thoracentesis was performed, which led to a diagnosis of chylothorax. Radio-graphs and a computed tomography scan showed multiple lytic lesions in several vertebrae, the cranium (Picture B, arrows) and the pelvis (Picture C, arrows) as well as pleural and peritoneal effusion and cystic lesions in the spleen (Picture D, arrows). We administered combination medical treatment consisting of interferon-α-2b, thalidomide, bevacizumab, zoledronic acid and enoxaparin and argue that this multitargeted approach should be considered in cases of severe GSS. In the first month of treatment, the amount of

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pleural and peritoneal fluid was successfully reduced. A diagnosis of GSS should therefore be suspected in patients who present with chylotorax and lytic bone lesions (1, 2).

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References