

LETTERS TO THE EDITOR

Gorham's syndrome vs generalized lymphangiomatosis: A close call

Sir,

The article on generalized lymphangiomatosis by Putta *et al.*^[1] was very interesting, and I would like to make a few observations regarding the article. Among the differentials mentioned by the authors – the entity of Gorham Stout syndrome is conspicuously absent – an entity which can account for almost all features with which the patient presented. Gorham Stout syndrome is a rare disease usually presenting in young individuals below 40 years of age.^[2] It is characterized by dilated vascular channels involving different organs of the body. The common presentations include osteolytic lesions in bones (most commonly involving pelvis, spine, etc.) and chylothorax, though other organs may be less commonly involved.^[3] Having a high index of suspicion is crucial and biopsy of the lesions (most commonly the bone) is diagnostic. Hefez *et al.*^[4] had suggested a diagnostic criteria for the same: Biopsy finding of angiomatous disease, absence of cellular atypia, minimal osteoclastic reaction with no dystrophic calcification, evidence of local bone progressive resorption, nonexpansile and nonulcerative lesion, absence of visceral involvement, osteolytic radiographic pattern, absence of hereditary/infective/malignant/metabolic, and immunologic etiology. Treatment of the disease is not well-established, however, the different modalities used includes radiotherapy, bisphosphonates, interferon alpha 2b etc.

The patient described by the authors showed most of the features described above. The patient was systematically worked up by the treating team and appropriate treatment was instituted. Whether treatment like bisphosphonates or interferon would have changed the patient's clinical course is difficult to speculate. Chylothorax (as seen in this patient) itself is a poor prognostic marker in Gorham's syndrome, hence, the sudden demise of the patient seems to reflect the severity of the involvement of the underlying disease. The terminal event of the patient – increased shortness of breath, metabolic acidosis, and seizure could have been due to sepsis (infection of bone, meningitis etc.) – which has been described in this syndrome. Finally, the difference between generalized lymphangiomatosis and Gorham syndrome is somewhat blurred with some authorities considering them to be a part of a spectrum.^[5] The implication of this

essentially is that more work needs to be done to further characterize this enigmatic disease.

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Conflicts of interest

There are no conflicts of interest.

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