CASE REPORT

Gorham's Disease: Vanishing Bone Syndrome

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Gorham's disease, also known as massive "osteolysis" or "vanishing bone disease" is an extremely rare bone disease. It is characterized by angiomatosis with adjacent bone resorption. We report an 8-years old boy with the disease who was managed successfully with alpha 2b interferon therapy.

Keywords: *Gorham's disease, Chylothorax, Osteolysis.*

Gorham's disease is characterized by loss of one or more bones associated with swelling or abnormal blood vessel growth (angiomatous proliferation). In about one fifth of the patients, chylothorax occurs and it carries a poor prognosis. Treatment options available are thoracic duct ligation, pleurodesis, radiotherapy, alpha 2b interferon supportive nutrition and biphosphonates.

CASE REPORT

An 8 year old boy weighing 20 kg was admitted with fever and pain in the left hypochondrium for 3 days. The child was febrile, thin built, and had deep set eyes. There was a huge hemangioma occupying left chest and hypochondrium. The air entry was diminished in left base. The blood count and metabolic profile were normal. Chest X-ray showed left lower lobe consolidation with pleural effusion and rarefaction of left lower ribs. He was treated with parentral antibiotics and intercostal drainage (ICD) tube was placed in the left side. Fluid draining was serosanginous in nature and biochemical parameters were suggestive of exudate. The child had been operated at 3 years of age for an axillary cystic hygroma/lymphagioma.

He subsequently developed right-sided chest discomfort and chest X-ray showed right pleural effusion for which a right-sided ICD tube was placed. CT scan chest showed left lower lobe consolidation, bilateral pleural effusion with septations on left side and eroded left lower ribs. A video assisted thoracoscopy was done on left side and fibrous strands were removed, fluid drained and parietal pleura was decorticated. The pleural fluid was chylous and showed high triglyceride level. More than 2 liters of chyle was drained per day from both the ICD tubes. We added parentral octreotide, and hypoproteinemia was corrected with albumin infusions and protein rich diet. A Tc 99m MDP whole body scan revealed increased bone activity. A diagnosis of Gorham's disease was made.

Major treatment options identified were radiation therapy, thoracic duct ligation and interferon therapy. Considering the potential to cause secondary malignancy and high failure rates, respectively, the first two options were not chosen. Child was started on alpha 2b interferon, which has anti-angiogenic properties, and a steady decline in chylous aspirate was noticed over a week. Both the ICD tubes were removed and a repeat radiograph of the chest showed good chest expansion. Child was

started on oral biphosphonates and nutritional supplements were added on discharge. After 6 months, a repeat bone scan showed decrease in tracer activity suggestive of disease under control.

DISCUSSION

Gorham's disease, also referred to as Gorham-Stout syndrome, idiopathic massive osteolysis and vanishing bone tumor, is a rare disorder described first by Gorham and Stout in 1955(1). Less than 200 cases are reported(2). Mandible, ribs, scapula, humerus, pelvis and femur are involved. If the shoulder girdle or thoracic cage is involved, then 17% of patients develop chylothorax which leads on to hypoproteinemia, malnutrition and immunosuppression; this is often fatal with a mortality rate of 64%(3). It is common in adolescents and young adults. There is no gender or racial predilection. The exact etiopathogenesis is not known. A progressive osteolysis is always associated with angiomatosis of blood vessels or lymphatics. The theories offered are silent hamartoma becoming active after minor trauma, neurovascular changes as seen in Sudec's atrophy, primary aberration of vascular tissue in bone inducing hypoxia and acidosis leading to increased local hydrolytic enzymes.

Gorham's syndrome is characterized by monocentric osteolysis origin with hereditary pattern(4). These patients usually present with abrupt or insidious onset of pain in affected site or with trivial injury bringing out the underlying problem.

Death occurs due to malnutrition, lymphocytopenia or infection(1,5). Radiological changes have been classified into four stages as (*i*) radiolucent foci, resembling patchy osteoporosis; (*ii*) shrin-kage of shaft of bones by tapering of the ends ("Sucked candy" appearance); (*iii*) complete resorption of the bone unless there is spontaneous arrest; and (*iv*) progression to adjacent bones and joints(6). There is no biochemical or endocrine abnormality(7). No standard medical therapy is available at present. Surgical options include resection of lesion and reconstruction using bone grafts and prostheses. Gorham's related chylothorax may be treated by pleurectomy, pleurodesis, thoracic duct ligation, radiation therapy, interferon therapy, oral clodronate and bleomycin(1).

Malnutrition and infection issues have to be promptly addressed. Of 22 reported cases of Gorham's related chylothorax between 1960 to 2000, 12 patients survived and 10 died. Of the 12 surviving patients, 8 were surgically managed and 4 were medically managed, all in a trial and error basis(5). We report this case due to its rarity of presentation and management and to create awareness about this disease entity.

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