SciMedCentral

Annals of Clinical Cytology and Pathology

Case Report

Gorham Disease Involving Thoracic Spine: A Case Report

Mohsen Esfandbod[1], Nasim Khajavirad[2]*, Marziyeh Galamkari[2], and Mahdi Khatuni[2]

[1]*Department of Hematology-Oncology, Tehran University of Medical Sciences, Iran*
[2]*Department of Internal Medicine, Tehran University of Medical Sciences, Iran*

***Corresponding author**
Nasim Khajavirad, Department of Internal Medicine, Tehran University of Medical Sciences, Tehran, Iran, Tel: 98-2166-939-922, Email: nkhajavirad@gmail.com

Submitted: 28 August 2017
Accepted: 26 September 2017
Published: 28 September 2017
ISSN: 2475-9430

Keywords
- Gorham disease
- Osteolytic lesion
- Spinal mass

Abstract

Gorham disease is an extremely rare bone disorder in musculoskeletal system. The pathologic process is the replacement of normal bone by an aggressively expanding vascular tissue. It may affect the appendicular or the axial skeleton. Common sites of involvement are the shoulder & the pelvic. It rarely involves spine that would cause serious neurologic complication. This case report shows a 51-year-old woman with Gorham disease who had paraparesia due to thoracic vertebral involvement.

Gorham disease should be in differential diagnosis of any osteolytic lesion in skeleton after excluding other common causes of osteolysis.

INTRODUCTION

Gorham disease, also known as Gorham-Stout syndrome, massive osteolysis, disappearing/vanishing bone disease, or phantom bone disease [1]; is an extremely rare bone disorder of musculoskeletal system. It was first reported by in 1838 [2]. In 1955 Gorham & his colleagues provided a more comprehensive report on massive osteolysis disease with histologic feature of intraosseous proliferation of hemangiomatosis or lymphangiomatosis tissue. Therefore the condition became known as "Gorham disease" [3].

Gorham disease is a non-hereditory disorder most affected before 40 years old. Its etiology &pathophysiology is poorly understood [4]. The pathological process is the replacement of normal bone by an aggressively expanding but non-neoplastic vascular tissue [1]. Also it has been suggested that an increase in the sensitivity of osteoclast precursors to humoralfactors, promote osteoclast formation & bone resorption [4].

Gorham disease may affect the appendicular or the axial skeleton. The most common sites of involvement are the shoulder & the pelvis [1]. Other sites which may be involved are the humerus, scapula, clavicle, sternum, mandible,ulnar, maxillofacial skeleton & hand [5-8]. Chylosis pericardial & pleural effusions may be present due to extension of the involved vertebra, scapula, rib and sternum to mediastinum [1,9]. It rarely involves spine that would cause serious neurologic complications [10,11].

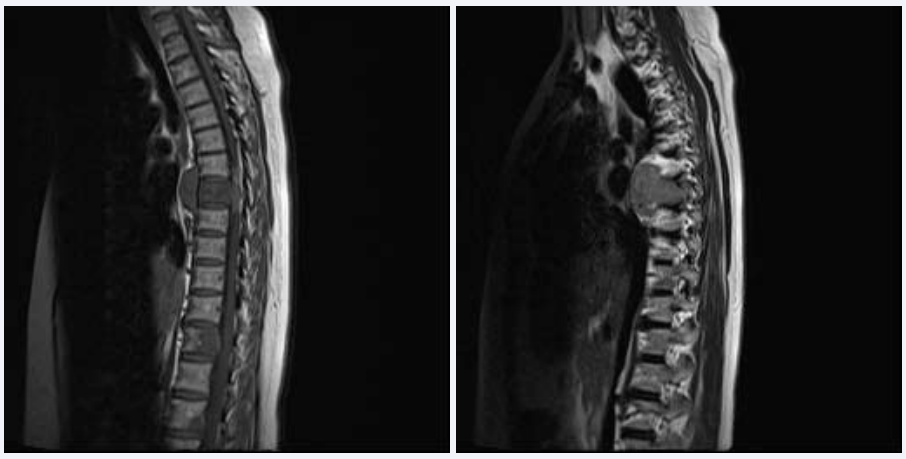

Figure 1 MRI shows mass involving T7 vertebral body.

Here we report a patient with Gorham disease who had neurological deficit due to thoracic vertebral involvement.

CASE REPORT

A 51 year old woman had a 3 month history of progressive low back pain & a week history of paraparesia & parestesia. She was a married house wife from Tehran & didn't have any history of trauma or radiation. She didn't mention any specific medical disorder except long term hypertension & impaired glucose tolerance. She was treated with losartan & metformin since 5 years ago. Her family & habit history was unremarkable.

On physical examination localized tenderness at 7-8 thoracic vertebraes was significant. Paraparesia (muscle force 2/5) with increased knee DTR & upward plantar reflex was detected. She also had sensory level at mid-chest wall.

The patient's lab data was acceptable except a low 25OH Vit D level (10ng/ml). Spinal magnetic resonance imaging (MRI) revealed a mass involving most of the T7 vertebral body (Figure 1); associated with a large lobulated paravertebral soft tissue mass mainly in right anterolateral portion. This was extended into epidural space at T7 level & causes local CSF blocks (Figure 2). Tc 99m MDP bone scintigraphy revealed increased uptake at T7 vertebral body (Figure 3). SPECT CT scan confirmed bones scan (Figure 4).

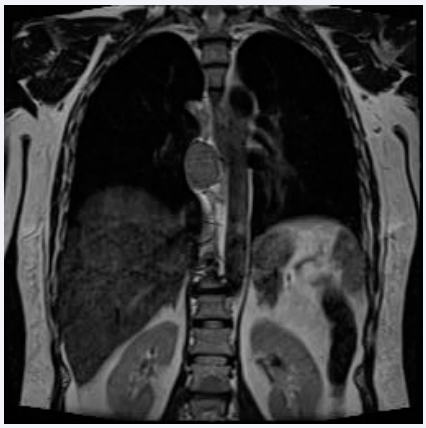

Figure 2 MRI shows large lobulated paravertebral mass in right anterolateral T7.

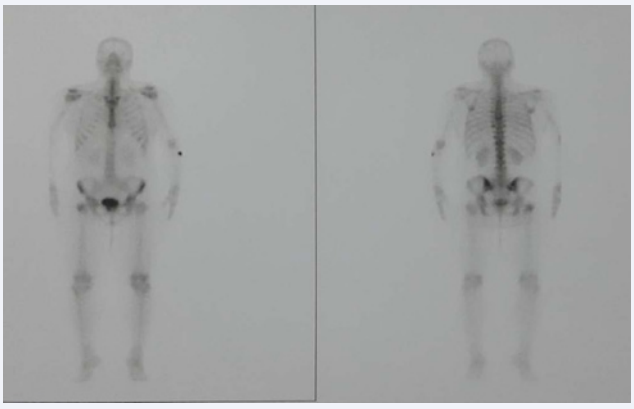

Figure 3 Bone scan revealed increased uptake at spinal thoracic level.

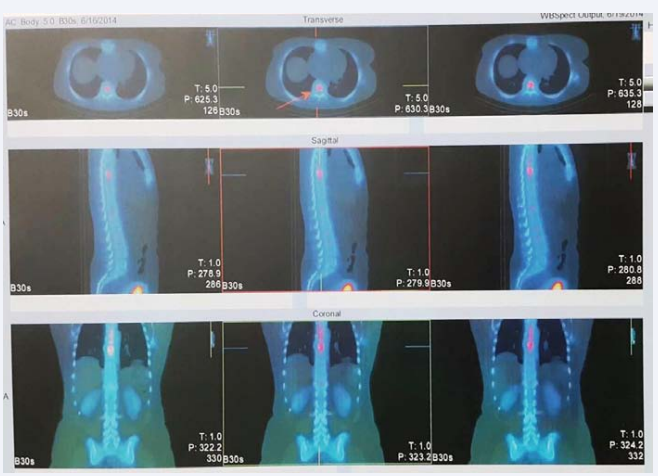

Figure 4 SPECT CT: increased uptake at spinal thoracic level.

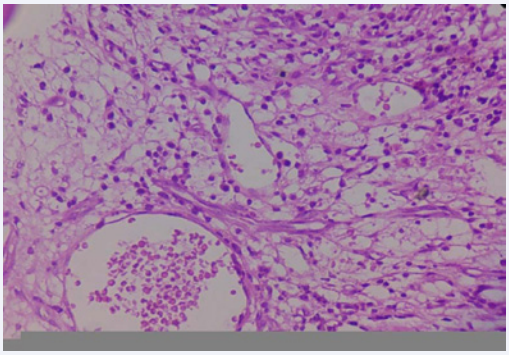

Figure 5 Histopathologic finding: Fragments of bone tissue replaced by proliferated angiomatous vascular space.

The patient underwent anterior thoracotomy, mass resection & spinal reconstruction. The histopathological results of the mass revealed fragments of bone tissue replaced by proliferated angiomatous vascular space in some areas, intervening hematopoietic cells mixed with prominent plasma cells and without presence of cellular atypia (Figure 5). The immunohistochemistry (IHC) staining was positive for CD3, CD20, CD31, BCL2, LCA markers and Ki67 in less than 5% of tumor cells was positive, suggested vascular origin of the lesion, presence of B and T lymphocytes and low cellular proliferative activity. These findings confirmed osteolysis with secondary angiomatosis "Gorham Disease".

After surgery, medical treatment with zoledronic acid and interferon was initiated &the patient was able to walk after 3 weeks.

DISCUSSION

Gorham disease is a rare mysterious bone disease of unknown etiology. Most of affected patients are before 40 years old. Less than 200 cases have been reported worldwide [4]. As noted Spinal involvement is extremely rare as an initial presentation of

Gorham disease which was revealed in our patient.

Diagnosis is often delayed as laboratory studies are usually normal; in our patient almost all laboratory findings were normal. A high clinical suspicion with radiologic & histopathologic findings can help for early diagnosis. Gorham disease is not a primary diagnosis [8]. The diagnosis should be made only after excluding other causes of osteolysis such as Paget disease at destructive stage, eosinophilic granuloma, Brown tumor, osteolytic metastasis &infections [1,4]. Gorham disease can also mimic various osteolytic syndromes like: Acro-osteolysis syndrome, Farber's disease and Winchester's syndrome.

The radiologic finding is variable. Intra medullary or subcortical lesions are seen during the initial stage, followed by progressive atrophy & fracture. The disease can extend to contagious bone & soft tissue [1]; as was seen in our patient that extended into epidural caused neurologic signs.

Radioisotope bone scan shows increased vascularity then decreased uptake at the site of diminished osseous tissue [1]. Tc scan & SPECT CT of our patient also revealed increased vascularity.

The natural history of Gorham disease is unforeseeable& spontaneous regression is also reported [1]. In some of reported cases after a variable time of evolution, the osteolysis would undergo spontaneous arrest [12]. Different treatment strategies propose for Gorham disease including: medical treatment with biphoshonates, a-2b interferon, calcium, vitamin D and androgens and also radiotherapy and even surgery in large lesion with patient disability [9,13].

CONCLUSION

Three weeks after surgery, our patient was able to walk & her disease has not been progressed during the next 6 months.

REFERENCES

1. Patel DV. Gorham's disease or massive osteolysis. Clin Med Res. 2005; 3: 65-74.
2. Jackson JBS. A boneless arm. Boston Med Surg. 1838; 10: 368-389.
3. Gorham LW, Stout AP. Massive osteolysis (acute spontaneous absorption of bone, phantom bone, disappearing bone); its relation to hemangiomatosis. J Bone Joint Surg Am. 1955; 37: 985-1004.
4. Parihar V, Yadav YR, Sharma D. Gorham's disease involving the left parietal bone: a case report. Cases J. 2008; 1: 258.
5. Scheller K, Herrmann-Trost P, Diesel L, Busse C, Heinzelmann C. Unspecific, idiopathic isolated osteolysis (Gorham-Stout syndrome) of the mandibular condylar process with its radiological, histological and clinical features: a case report and review of literature. Oral Maxillofac Surg. 2014; 18: 75-79.
6. Schumann E, Wild A, Seller K. [Gorham-stout disease]. Z Orthop Unfall. 2008; 146: 655-659.
7. Bode-Lesniewska B, von HochstetterA, Exner G, Hodler J. Gorham-Stout disease of the shoulder girdle &cervico-thoracic spine: fatal course in a 65 year old woman. Skeletal Radiol. 2002; 31: 724-729.
8. Tavakoli Darestani R, Sharifzadeh A, Bagherian Lemraski M, Farhang Zanganeh R. A Rare Case of Gorham's Disease: Primary Ulnar Involvement with Secondary Spread to the Radius and Elbow. Trauma Mon. 2013; 18: 41-45.
9. Nikolaou VS, Chytas D, Korres D, Efstathopoulos N. Vanishing bone disease (Gorham-Stout syndrome): A review of a rare entity. World J Orthop. 2014; 5: 694-698.
10. Barman A, Bhide R, Viswanathan A, George J, Thomas R, Tharion G. Gorham's disease of the spine. NeuroRehabilitation. 2013; 33: 121-126.
11. Kakuta Y, Iizuka H, Kobayashi R, Iizuka Y, Takahashi T, Mohara J, et al. Gorham disease of the lumbar spine with an abdominal aortic aneurysm: a case report. Spine J. 2014; 14: e5-9.
12. Boyer P, Bourgeois P, Boyer O, Catonné Y, Saillant G. Massive Gorham-Stout syndrome of the pelvis. Clin Rheumatol. 2005; 24: 551-555.
13. Hu P, Yuan XG, Hu XY, Shen FR, Wang JA. Gorham-Stout syndrome in mainland China: a case series of 67 patients and review of the literature. J Zhejiang Univ Sci B. 2013; 14: 729-735.

Cite this article

Esfandbod M, Khajavirad N, Galamkari M, Khatuni M (2017) Gorham Disease Involving Thoracic Spine: A Case Report. Ann Clin Cytol Pathol 3(7): 1080.