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GORHAM STOUT DISEASE:A RARE CASE REPORT

Orthopaedics

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ABSTRACT

Background: Disappearing bone disease -Gorham Stout disease a unparallel case of Unknown Origin . Medical condition of Bones, which does not have any specific etiology. An extremely rare disorder, which causes the proliferation of endothelial channels and can lead to results and disappearance of bone. Here sudden bone osseous matrix is destroyed and also lymphatic vascular structures up proliferated and further leads to excessive regional osteolysis.With a variating clinical presentation and very often is diagnosed as benign disease which further spreads to an unclear prognosis.Mainly diagnosed by single and various combinations like clinical features, histological findings and radiological findings. Many therapeutic options have managed to provide certain efficacy, but most effective therapeutic option is still controversial without a certain standard treatment protocol

Case presentation: A case of disappearing bone disease affecting the proximal femur, pelvis, sacrum in a 45 year old male has been described here. Clinical features of the disease are pain , swelling of the affected region, functional impairment. In this case the diagnosis was made after the pathological fracture of femur. Based on radiological and clinical findings. Considering stabilisation as an important aspect internal fixation that was done followed by Bisphosphonates used for progession control and is now being currently followed up with no cessation of disease. But internal fixation has prevented complete severe disability.

Conclusion: Hereby given that confirmed ethiopathology of disappearing bone disease is not completely verified and further study is still needed to effectively treat and provide intervention. But the need for every physician to know of it's presence, promptly direct a patient towards the right direction and we are hence reporting this for it's rarity.

KEYWORDS

Disappearing bone disease, massive idiopathic osteolysis, Gorham stout syndrome,Phantom bone

BACKGROUND:

Excessive sudden osteolysis of bone first described by Gorham et al [1] in 1954 Gorham and stout[2] in 1955 where the first 24 cases of this disease were presented after which until lately many names have been given to this condition which are Phantom bone, vanishing bone disease, lymphangiomatosis hemangiomatosis and Gorham stout syndrome but commonly favored is Gorham's disease[3] .Currently approximately 300 cases of Gorham stout syndrome are reported. [4] Even after wide extensive Research and investigation of pathogenic mechanisms, the main cause still remains a mystery characterized mainly by sudden unnatural proliferative growth of endothelial lined thin walled channels of vascular origin or lymphatic origin which has an increased osteoclasts formation and leading to progressive bone resorption[2] .Here in most cases a history of trauma is said to be a trigger of osteolysis and hence said to be a blamed cause of GSD[1] .Major sites affected belong to the upper extremity involving a single bone. The lesion being non expansile and monocentric,maybe polyostotic but locally expanding aggressively[5-8]. A cause of non familial type trauma triggered Gorham's disease is presented here.

Case report

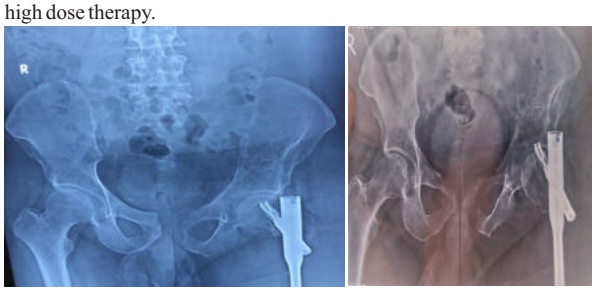
A previously healthy 45 year old male visited the hospital in 2014 for pain in the back of the pelvis and vague symptoms of pain and tingling sensation radiating to the left lower limb. Pain persisted despite being given conservative therapy like various analgesics and sitz bath therapy.

There were no constitutional symptoms such as fever, anorexia or weight loss on presentation. No significant family history. No history of addictions or significant habits. He follows a vegetarian diet and is a BSNL office worker. Patient revisited in February 2015 where x-rays were done and on physical examination range of motion was restricted at the hip joint. The X-rays had no abnormality followed by which he visited the local bone setter as there was no pain relief and got a massage in March 2015. He had temporary relief but pain again reoccurred in 15 days for which in August 2015 he got an MRI LS spine done followed by physiotherapy at Chandigarh. The findings of the MRI showed in favor of benign lesion of the vascular origin hemangioma. He was further given an Injection depomedrol in November 2015. Followed by open and closed biopsy at Left SI joint at Jalandhar in December 2015 which also proved that it was negative for malignancy/TB and is an inflammatory lesion. After which in January 2016 he was given steroids and brucellosis medicine course but no improvement was observed. Followed by which Anti tubercular treatment was given for one year. In 2017 x-ray were normal with no changes seen He also got a core needle biopsy fine which also showed no necrosis, granulomas or fungal profiles. After all this, an unfortunate event occurred and he had a fall on 19/7/2017 and was operated for CRIF with PFN nailing which was followed by gradual bone loss in 2018 where he was diagnosed with idiopathic massive osteolysis and was started on medicine GEM FOS 70mg every week for 5months. The disease was not controlled and the patient now also has severe teeth erosion. Patient has continuous pain and limping in left lower limb and recently on examination it has been observed of a swelling and collection over left buttock and multiple wound mark over left buttock along with muscle wasting. X-rays were repeated which showed loss of proximal femur, loss of sacral bone with pubic symphysis bone loss. Further to which he was continued on 2 months medications with analgesics and Bisphosphonates but patient showed no improvement and is now bed ridden. Recent x-rays show absence of pelvis and it also shows the implant in situ has moved proximally.

Background

Our Patient in this study was followed up for seven years with increased progression. Despite being self-limiting and benign it has fatal outcomes when at important anatomically benefiting locations specially the spine. Medical therapy includes Bisphosphonates, interferon Alpha 2b,vitamin D, calcium, Anti-VEGF-A antibody bevacizumab, steroids,LMWH here in which stop bone destruction and stop bone disease progression. According to literature Bisphosphonates had been more effective treatment in GSD then all others. Thou it does not help in bone formation. It stops the cell death of osteoclasts and reduces its activity. Radiotherapy thou reported as a

success in most cases. Results are still in controversy as there is no exact dose for GSD it further had complications of instigating secondary malignancies and growth restrictions in patient's who took high dose therapy.



CONCLUSION

Finally concluding that given that ethiopathology of disappearing bone disease is not completely verified. And as demonstrated in previous studies. That reconstruction biologically is not effective for disease control. A radiotherapeutic with histological and Clinical features and Bisphosphonates helps in cessation of progressive growth. But further study is still needed to effectively treat and provide intervention and to make known to every physician of its presence and promptly direct a patient towards the right approach. The case is being reported for its rarity.

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