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Vanishing Bone Disease of Hand- Case Report and Review of Literature

Orthopaedics

KEYWORDS: Vanishing Bone Disease (VBD), Micro vascular Proliferation, Bone Resorption, Carpal and Metacarpal Bones

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ABSTRACT

INTRODUCTION Vanishing bone disease (Gorham-Stout syndrome) is an extremely rare disease characterised by gradual resorption and disappearance of osseous structures caused by micro vascular proliferation. Exact aetiology is unknown so far with few cases of idiopathic osteolysis have been reported. Cases have been reported in almost all age groups but mostly among children and adolescents. Clinical presentation depends upon the site of skeletal involvement. The disease has highly variable prognosis. It can cause minimum disability and death due to possibility of critical structure involvement. **CASE REPORT** A 22 year old male presented with complaints of weakness in left hand, gradual shortening and hyperextension of middle and index fingers without any history of trivial trauma, fever or pain. **CONCLUSION** VBD is characterized by spontaneous and progressive disappearance of bone and is associated with angiomatosis. Radiography is best tool for diagnosing this disease. Hereditary osteolysis, Sudeck's atrophy, endocrine or autoimmune diseases, infection and tumors should be excluded

Introduction

Vanishing bone disease is characterized by vasculogenesis and resorption of osseous matrix with poorly understood aetiology (15).Typical presentation is that of gradual resorption of a single bone or multiple bones sharing common joint. Most commonly involves; shoulder, skull and pelvic girdle, with extremely rare involvement of bones of hand (11). Asymptomatic to severe disability and life threatening cardiothoracic complications has been seen with this disease. Lack of bone healing following fracture is hallmark of the disease (11). We reported a case of vanishing bone disease in a 22 year male.

CASE

A 22 year old male presented with complaints of weakness in left hand, gradual shortening and hyperextension of middle and index fingers without any history of trivial trauma, fever or pain. There was no family history of similar complaints. On plain radiograph thinning & sclerosis of 3rd and 4th metacarpals with lytic areas in head of 3rd metacarpal. Initially diagnosis of chronic osteomyelitis was made and patient was put on antibiotic treatment Patient again came after 1 month with complaints of pain in affected parts. Plain radiograph showed fracture near base of 3rd metacarpal. Plaster cast was applied after reduction. On serial follow up for 6 month, gradual thinning and resorption of 3rd & 4th and metacarpals were noted with appearance of lytic areas in capitate and hamate . A clinical suspicion of essential osteolysis was raised and CT Angiography was advised to look for vascular proliferation. Meanwhile complete blood counts, biopsy and culture & sensitivity test were made which were negative for osteomyelitis and other bone tumors. On ct angiography , no macro vascular proliferation was seen in affected site . 3rd metacarpal was replaced by soft tissue attenuation band without any evidence of surrounding fluid collection .lytic areas was noted in capitates and hamate with thinning and sclerosis of 4th metacarpal .Diagnosis of vanishing bone disease was made retrospectively which was supported by increase osteoclast and micro vascular proliferation in histological analysis .

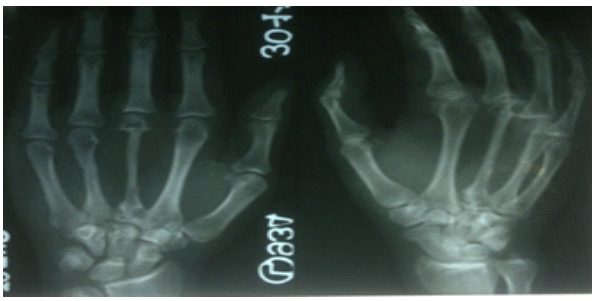

Figure 1- concentric thinning of 3rd & 4th metacarpal with lytic lesions in epiphysis

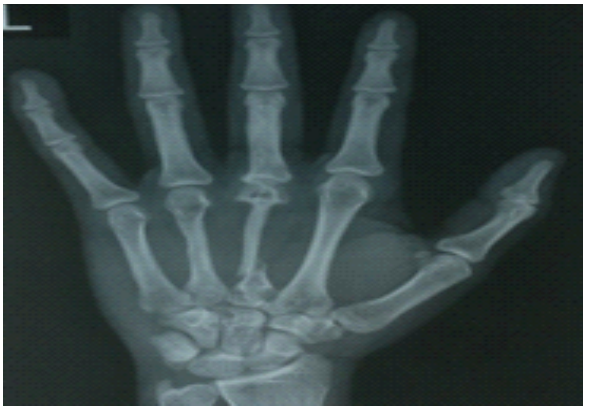

Figure 2- pathological fracture at base of 3rd metacarpal

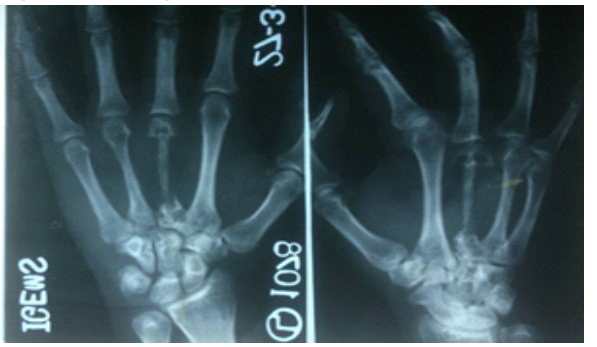

Figure3 – gradual resorption of 3rd metacarpal

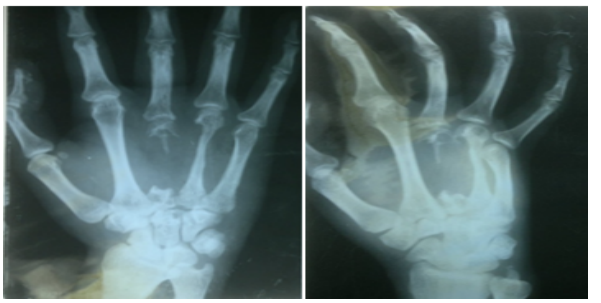

Figure 4- near complete resorption of diaphysis of 3rd metacarpal

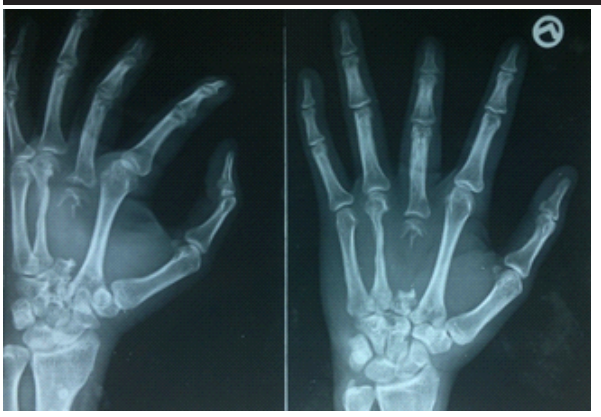

Figure 5- osteolysis increased towards epiphysis of 3rd metacarpal, thinning of shaft of 4th metacarpal compared to fig. 1 and apperance of lytic lesions in capitate and hamate

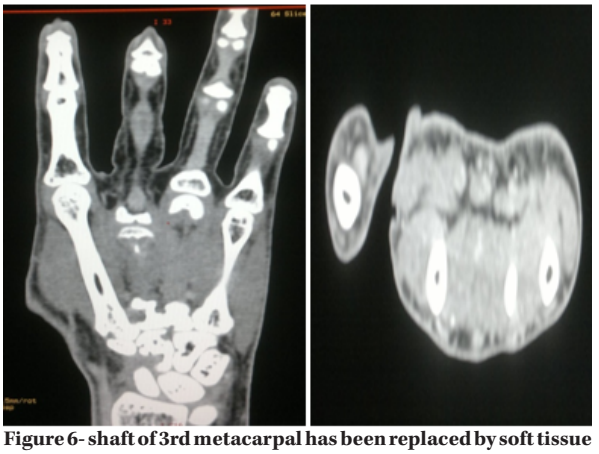

Figure 6- shaft of 3rd metacarpal has been replaced by soft tissue attenuating cord . No macrovascular proliferation, enhancement of scar tissue or phleboliths are seen in arterial phase .

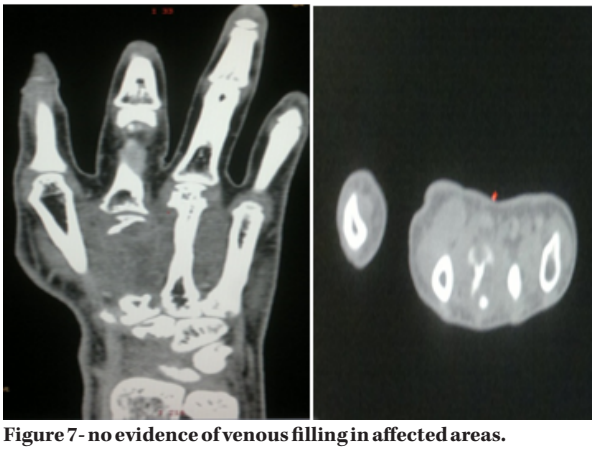

Figure 7- no evidence of venous filling in affected areas.

Discussion

VANISHING BONE DISEASE is characterized by spontaneous and progressive resorption of bone. The disease was first described in 1838 (6). Many names are used for this disease like phantom bone, massive osteolysis, essential osteolysis and disappearing or VBD, acute spontaneous absorption of bone, Gorham's disease. Shoulder, pelvic girdle and the maxillofacial region are the common locations for the disease. Male predominance has been seen (6). The disease can be monotonic or polyostotic, but multicentricity is unusual (14). The disease may often involve adjacent bones and soft tissues. No association with endocrine or metabolic abnormalities are seen (8). Etiopathological mechanism of this disease is still unknown without clear evidence of malignant, neuropathic and infectious factors (9). Incidence may be related to history of trauma, but half of the patients are without history of trauma (14). Prognosis of this disease is highly variable ranging from minimum disability to death caused by involvement of vital structures (7). Insidious course is noted due to painless nature of resorption process allowing normal activity while osteolysis occurs which may later present with pathological fractures (7). The diagnosis of the disease is based on high clinical suspicion, radiologic finding, and histopathology examination. Mild pain, decrease strength in the affected site and deformities are common presentations (11). Laboratory findings are nonspecific and variable. Alkaline phosphatase level may be elevated due to pathological fracture. Role of increased interleukin-6 has been described in increased osteoclast activity (2). Many imaging techniques may be used likes plain radiographs, radioisotope scans, computed tomography, scintigraphy and MRI (10). Extent of bony and soft tissue involvement can be seen by Computed tomography and may help in biopsy of the affected bone (1). Conventional angiography, scintigraphy, and MRI are not diagnostic imaging modalities in VBD. A radiograph is best tools for detecting the disease (13). The radiographic findings of unilateral partial or total disappearance of contiguous bones, tapering of bony remnants, and absence of a sclerosing or osteoblastic reaction are present. Two phases of the histopathological changes are proposed in the literature (5). Initially increased micro vascular proliferation occurs in the bone-displacing fibrous tissue part and in the second phase only fibrous tissue remains. No definite treatment is known so far. Surgical intervention may be a method of choice (3) which involves local resection of the affected bone. Radiation therapy has shown apparent arrest in some cases (4). Radiotherapy causes sclerosis of the proliferating blood vessels and prevents regrowth. Effective total dose has been proposed to be 30 to 45 Gy (4). Chemotherapy also has been used with success in few patients. Treatment of VBD should include combined or alone use of surgery, radiotherapy, and various medications. Involvement of the pelvis, thorax, and cervical spine can cause severe morbidity. However, the disease usually remains localized and undergoes eventual spontaneous arrest.

Conclusion

VBD is characterized by spontaneous and progressive disappearance of bone and is associated with angiomatosis. Radiography is best tool for diagnosing this disease. Hereditary osteolysis, Sudeck's atrophy, endocrine or autoimmune diseases, infection and tumors should be excluded. Epidemiology still has not been established because of fewer case reports. Anti-osteoclastic agents, radiotherapy and surgical interventions according to severity of disease with variable success have been proposed.

Clinical message

Recognition of the disease requires a high index of suspicion and an extensive workup. Because of its serious morbidity, Gorham's must always be considered in the differential diagnosis of osteolytic lesions

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