Biomedical & Pharmacology Journal Vol. **8**(Spl. Edn.), 09-13 (Oct. 2015)

Massive Osteolysis of Oral Cavity: A Review with Emphysis on its Etio-Pathogenesis and Histological Appearance

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(Received: August 15, 2015; accepted: September 20, 2015)

DOI: http://dx.doi.org/10.13005/bpj/644

ABSTRACT

Massive osteolysis is a rare condition of unknown aetiology resulting in progressive destruction of bony structures. Literature reviews suggest less than 40 cases of massive osteolysis in the jaws has been reported.Clinically it is seen mostly between age groupof 18-40 years and depending on the site of involvement is shows various clinical presentation. Even after so many advances in the field of medicine still this disease is still considered as nightmare for the pathologist as the course of the diseaseand prognosis after treatment are unpredictable. As its name indicates its radiological features varies from slow resorption of affected bone finally resulting in complete loss of affected bone.No specific treatment is proven effective against this disease. Hence the purpose of this review is to provide comprehensive in depth idea about the disease to help in early diagnosis.

Key words: Massive osteolysis, Bone disease, Vanishing bone disease.

INTRODUCTION

Massive osteolysis is a rare destructive bone disease that is characterized by progressive destruction of one or more bones followed by proliferation of vascular channels and fibrous tissue. No bones is immune to this disease, however maxillofacial and upper extremity are most preferred region out of which mandible and scapula are common site respectively.[1,2,3] In the oral cavity, clinical presentation ranges from being completely asymptomatic in early stages to slight pain, tooth mobility, pathological fracture in advanced cases.[4] Mostly seen in male patients. Etiopathogenesis of this disease is still controversial as there is no evidence of malignant, infectious and neuropathic cause.[5]

It can be studied by various diagnostic aids namelyplain radiograph, computed to mography and magnetic resonance etc.[6]The treatment is the main setback for this disease as no treatment is found to be successful in treating the case.[7]Hence it is imperative to diagnose this disease in early stage to reduce its morbidity.Even thougha lot is spoken about this disease in literature, there is still lack of comprehensive review of this astonishing bone disease. Hence the purpose of the review is make you understand the pathogenesis, and have a clear idea about various clinical feature, radiological and histological features of this disease so that it to help in early diagnosis.

History

In 1872 J.B Jackson identified complete osteolysis of humerus in 12 year old boy and described as 'vanishing bone disease'.[8] However, the first case to be reported in jaws was by Romer in 1924.[9]Finally in 1955, namedas 'Gorham's disease' after Gorham and Stout, when they

presented first overview on this disease by reporting 24 cases.[10]

Etio-pathogenesis

The exact pathogenesis of massive osteolysis is still unknown.There is no evidence for genetic inheritance or associated metabolic or endocrine abnormalities.[11]It is hypothesized that the sluggish circulation of blood in wide capillary-like vessels produces local hypoxic effect and a decreased pH. Such an environment favours the activity of various hydrolytic enzymes like acid phosphatase resulting in bone resorption.[12]

Since most often it is preceded with trauma, it may arise due to aberrations in the normal healing process of bone i.e. neovascularization of the blood clot, after injury.[8]Trauma may even activate silent hamartoma inside the bone.[13]Some of other concept which are considered are endothelial dysplasia of blood and lymphantics,[14] defect in osteoclast activity.[15] Even thyroid cell, calcitonin are considered in pathogenesis.[16]

Clinical feature

Massive osteolysis has a predilection for children and young adults. About 50% of affected patients give a history of trauma of trivial nature.[5] maxillofacial region and shoulder are most commonly affected site.[1]It is usually unilateral.[18]Clinical manifestation includes dull pain, weakness, swelling and deformity of the affected bone.[3]Laboratory findings are completely within normal limits.[11]

Among the cases with maxillofacial involvement, the mandible was the most frequently affected bone.[11]Oral manifestation includes pain, mobile but vital teeth,[1] malocclusion, deviation of the mandible due to involvement of tempero-mandibular joint.[19] In the later stages, normal functions of jaw such as mastication, swallowing, speaking is also affected.[1]

Radiographic Feature

Radiographic feature plays a major role in the diagnosis of massive osteolysis.[20] In 1976, Patrick classified gradual changes in radiology of massive osteolysis into four stages. It usually starts as small radiolucent foci initially. This leads to tapering of bone ends.If it still progresses, it leads to complete resorption of bone. In fourth stage it may even spread to adjacent bone and joints.[21]

CT plays a major role in evaluating the involvement of facial bones and the skull, and can determine any extension into the base of the skull.[22]

Histopathologic features

Histopathology section of massive osteolysis shows features depending on the stage of the disease. During the early stages, vascular proliferation made up of thin - walled channels are present.Another peculiar feature is sparcity or absence of osteoclasts.[9]In the later stages, the area is more collagenized and paucicellular.[11]

Heffez *et al* suggested eight criteria for diagnosis of massive osteolysis, involving histopathologic and clinical for definitive diagnosis. It includes:

1. Positive biopsy in terms of angiomatous tissue presence
2. Absence of cellular atypia
3. Minimal or no osteoclastic response and absence of dystrophic calcification
4. Evidence of local bone progressive resorption
5. Non-expansive, non-ulcerative lesion
6. Absence of visceral involvement
7. Osteolytic radiographic pattern and
8. Negative hereditary, metabolic, neoplastic, immunologic and infectious etiology.[23]

Differential Diagnosis

Most often diagnosis of massive osteolysis is arrived by exclusion of other bone pathology resulting in osteolysis. So proper patient history, clinical examination, lab investigation, radiological examination and finally histological examinationare need of the hour.Some of the other common diseases considered are Acro-osteolysis of Hajdu and Cheney, Idiopathic multicentricosteolysis (carpal-tarsal osteolysis), Multicentricosteolysis with nephropathy, Hereditary multicentricosteolysis, Neurogenic osteolysis, Acro-osteolysis of Joseph, Acro-osteolysis of Shinz, Farber's disease and Osteolysis with detritic synovitis.[24]

Treatment

Review of literature showed that no single treatment plan is effective in stopping the disease process and restoring the aesthetic and function of the affected bone.Therapeuticregieme considered are fluoride, calcitonin, and chemotherapeutic agents).[11]Surgical intervention includes resection of affected bone and subsequent reconstruction using bone graft.[25] but most often it leads to resorption of bone graft itself.[26] Other treatment modalities tried are chemotherapy and radiation therapy. Anti-osteoclastic medications like bisphosphonate and alpha-2b-interferon are example of chemotherapeutic agents used with some successes.[27]

Radiation therapy is most accepted, especially in its early, as it stops the progression by fastening sclerosis of the proliferating vessels and prevents its regrowth.[11]The most effective dose is 30 to 45 Gy. However it also comes with some disadvantages like gastrointestinal irritation causing nausea and vomiting, osteoradionecrosis, damage to tooth eruption (in case of maxilla-facial region) etc. [28]

Prognosis

Because of controversy regarding the course of massive osteolysis, its prognosis also remains unpredictable. Some cases may get arrested in early stages itself, whereas some may lead to complete destruction of affected bone.

Complications

It can leads to chylothorax, hemothorax,[21] paraplegia (related to spinal cord), respiratory dysfunction and finally it can lead to death.[29]

DISCUSSION

The massive osteolysis is a fascinating bone disease characterized by gradual resorption of affected bone and replaced by fibrous connective.It is mostly seen in male patients usually less than 40 years of age.[20]Although no bone is immune to this disease, it mostly seen in skeletalstructures like thorax, hip bones, spine and mandible.[1] Its course can be divided into two phases, an active phase of bone destruction and a second latent phaseBut the duration of each phase varies from few months to several years.[18]Clinical presentation may vary fromasymptomatic, mild pain and in extreme cases leads to chylothorax depending on the extent of the disease.Important clue for the diagnosis is that all the laboratory findings will be normal.[31] Because it is primarily a bone disease, imaging tools like Conventional radiograph, CT,MRI are very useful in diagnosing the disease.[32]There is no proper treatment is effective in arresting the progression of this disease,even though radiotherapy sort with some success.

CONCLUSION

"Vanishing bone disease" among many names describe the disease in terms of clinical aspect, radiological aspect as well as histological aspect.Because of its devastating effect on the bone and shortcoming in its treatment plan, it is prudentfor us to be educated in depth about this disease. So that it can be diagnosed in its early, subsequently improving the prognosis of patient. As dentist,it is important to investigate the head and neck region when suspecting a case of massive osteolysis as without doubt mandible is also one among the risk prone.

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