Generalized External Cervical Resorption - A Possible Indicator of Aggressiveness in Lymphangiomatosis - Gorham-Stout Syndrome

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Abstract

Lymphangiomatosis with Gorham-Stout syndrome is a rare disease involving two pathological processes: Infiltration of nonmalignant, aberrant lymphatic vessels into soft and hard tissues and bone lysis. This report will describe a patient suffering from Lymphangiomatosis with external dental cervical resorption that mimics caries lesions. The rate of resorption was found to be related to disease progression. All the radiographic records of the patient were collected and compared, including three full mouth peri-apical series. Seven teeth with external cervical resorption were selected for comparison. Measurements of total tooth area and resorbed areas were performed and the ratio of resorption versus total tooth area was calculated. The rate of resorption in this patient correlated with the destructive activity of the disease. Since the clinical course of the disease followed the dental resorption rate we can assume that both bone lysis and root resorption are parts of the same process.

Keywords
Gorham-Stout Syndrome, Dental Caries, External Dental Cervical Resorption

1. Introduction

Lymphangiomatosis with Gorham-Stout disease (LGSD) is a disease of unknown etiology and pathophysiology that affects various bones of the body [1] with a predilection for: the long bones of the extremities, spine, pelvis and the jaws [2]. Bone at involved sites is replaced by a proliferation of new lymphatic vessels in LGSD and angiomatous changes are noted in Gorham-Stout Syndrome (GSS) [3]. LGSD is sometimes considered an uncommon, more aggressive subtype of GSS, with higher morbidity and mortality rates [4].

From the first description by Jackson [5] in 1838, until now, some 300 patients have been reported in the English literature as suffering from GSS. Of these, approximately 40 have been diagnosed with Lymphangiomatosis type of GSS (LGSD)[6]. Patients with the aggressive form of LGSD may have recurrent pleural effusions, pathological bone fractures, general edema and early death with survival rate of about 50% [7]. In general, the first signs of GSS are found on total body radiographic studies and are radioluencies with no definitive borders and no other clinical signs of malignancy.

Any bone can be affected, although there is a predilection for pelvis, mandible, humerus and axial skeleton.

The cause of the bone resorption is uncertain, either involving osteoclasts or the angiomatous tissue, or both. To the best of our knowledge this is the first article presenting dental involvement noticed on periapical radiographs.

2. Case Report

Patient

Figure 1. Complete full-mouth parallel radiographic series performed at admission.

A 56-year-old woman was referred to the Oral Medicine Clinic by her attending physician for evaluation of generalized external cervical root resorption. The extent of the phenomenon was noted on a full mouth periapical radiograph series as multi concentric symmetrical, well defined areas, resembling a "bitten apple" (Fig. 1). For five years prior to presentation, the patient had recurrent pleural effusions, and cytotoxic drugs, systemic steroids, plasmapheresis and surgical pleurodesis were employed to treat. A biopsy from an ill defined borders, radiolucent lesion of a lytic lesion on the left tibia presented abnormal lymphatic vascularisation compatible with the clinical and radiographic findings of GSS. Over the following 4 years the patient underwent regular dental checkups and treatments in the Oral Medicine clinic. A biopsy was also taken from a
resorbed lesion on the mesial of the lower right first molar. Histological findings revealed connective tissue, with young fibroblasts interspersed by thin-wall vascular spaces, and remnants of degraded dentin. The patient was stable for the first three years, and began to deteriorate thereafter. In the fourth year after initial presentation (9 years following start of the disease) the patient was treated several times for hypo-albuminemia and severe whole body edema. She died that year, nine years following her initial clinical complaint, in the fourth year of oral medicine/dental follow-up. Autopsy confirmed the diagnosis of combined LGSD, and histological examination revealed lymphangiomatosis involving serous membranes of the thorax, pericardium and peritoneum, as well as a lytic lesion on her left upper tibia, infiltrated by thin wall lymphatic vessels with focal bone resorption. Three complete full-mouth parallel technique of intraoral radiography series were compared. The first was from the initial visit in 1997, the second was from 1999 and the final series in 2000 [1]. Seven teeth with no dental treatments showing external cervical resorption were compared. Four central incisors teeth [teeth no. 8, 9, 24, 25] (Fig. 2) and three molars: upper right first [tooth no.3] and the lower second molars on both sides [teeth no. 18 & 31.

4. Results

The total tooth area calculated from the radiographs (baseline presentation) was 521459, 520065 and 533254 pixels, from the 1997, 1999 and 2000 respectively. The resorbed areas were: 45096, 82126 and 157160 pixels respectively. The ratio of resorption to total area was 0.08, 0.15, 0.29, respectively. (Table 1, Fig. 3)

![Figure 3. Extent of tooth resorption: tooth area to resorption areas ratio.](image)

Statistically significant differences were found between the first (0.08) year and fourth (final) year (0.29) as well as between the third (0.15) and fourth year (0.29). There were no significant differences between the first (0.08) and the third year (0.15) (Fig. 4).

Both the resorbed area and the ratio of resorbed area to tooth area showed increased significantly during follow-up (p=0.0004, p<0.0001 respectively).

Changes in resorbed area and the ratio of resorbed area to tooth area were insignificantly different in the 1997 and 1999 radiographs (p=0.1051, p=0.0752 respectively), whereas these measures were significantly different when comparing the 1999 and 2000 radiographs was (p=0.0015, p=0.0002).

5. Discussion

Bones affected by Gorham-Stout Syndrome lose normal structure, as the hard tissue is replaced by fibrotic connective tissue interspersed with newly formed blood or lymphatic vessels [2]. Bones of the extremities, spine, pelvis and face, i.e., all skeletal bones can be affected by this syndrome. Patients may experience pathological fractures of the involved bones. In those with concomitant lymphangiomatosis, pleural effusion, hypoalbuminemia, general edema, and early death may occur [3]. Oral involvement is included the predilection of the disease for the facial bones [4] most commonly the jaws [8]. To the best of our knowledge the patient reported here is the first to exhibit direct dental involvement with external cervical resorption as the presenting sign.

Dental resorption has been reported in GSS, and has also been overlooked in several reports. Internal resorption can be seen in radiographs from various reports, including the original report by Jackson [5], Bucher [9] and the
multi-dental, apical absorption reported by Paley et al. [10].

In addition to highlighting the resorption of dentin caused by GGS and LGSD, it is important to note that the rate of dentin resorption correlated with disease activity in the presented patient. The data and statistical workup are based on data relating to seven teeth of a single patient. Resorbed area was determined using ImageJ processing software; developed by the "National Institute of Health" in 1997 [11]. The software has the ability to calculate tooth area size and may be helpful in dental research for determining root resorption after orthodontic treatment [12] or bone resorption after dental implants [13]. When the changes in radiographic appearance were not statistically significant, the patient was clinically stable. In contrast, when the radiographic changes increased dramatically, the patient experienced a serious decline in her medical condition. During the patient’s final year of life, the absorption rate accelerated as her condition deteriorated, and the radiographic changes reflected this clinical course. In other words, the changes noted in the 1997-1999, showed that the progression of resorption front was relatively slow, whereas in the 2000 radiographs rapid loss of tooth structure was noted.

She suffered from multi-organ edema and anasarca requiring treatment in the emergency room followed by hospitalization in the intensive care unit until her death. During this period the accelerated resorption caused decoronation of several teeth.

Histological findings from the soft tissue from the dental resorption area were compatible with LGSD. There were remnants of dentin and cementum within this soft tissue. The resorption began apical to the cemento enamel junction (CEJ) and then invaded the crown under the enamel. Heithersay described aggressive cervical resorption and graded resorption severity according to the extent of invasion, into four classes: class 1 represents the least invasive and class 4 represents the most, with resorption beyond the coronal third of the root [14]. Heithersay's description of resorption is based on the radiographic “moth eaten” appearance, with no clear borders, and involves the crown as well as the root. The resorption presented here is different; we noted symmetrical, well defined areas, resembling a "bitten apple". Since the resorption described here is different from the "invasive cervical resorption" described by Heithersay we will use the term concentric symmetrical cervical resorption to describe our findings. This form of resorption has been previously described, but no etiology was mentioned [15].

These findings imply that the destructive lymphangiomatotic potential may affect all tissue types: skin, soft tissue of internal organs, bone and viable tooth material – dentin and cementum but not enamel. The rate of resorption in our patient served as a relatively simple aid monitoring the disease.

Conflicts of Interest

The authors deny any conflicts of interest. The authors declare self-funding of the research.

REFERENCES