Oral Manifestations of Hyperoxaluria

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Abstract: Primary hyperoxaluria is a rare, inherited autosomal recessive disease caused by defects in the metabolism of glyoxylate. Oral manifestations of hyperoxaluria are rare. However, bone and tooth resorption may be the result of chronic inflammation and the presence of osteoclastic cells surrounding the oxalate crystal deposit. A deposit of calcium oxalate in the periodontium was identified in a patient with end-stage renal disease. Dental radiographs indicated bone loss and external tooth resorption. Radiolucent image in the inferior incisor region was observed and removed. The tissue showed granulomatous inflammation with foreign body reaction and associated crystalline deposits. When viewed in polarized light, these deposits are green and presented a birefringent aspect, which were interpreted as calcium oxalate crystals compatible with oxaluria. Oral manifestations of hyperoxaluria are of particular interest because of the unusual location of the oxalate crystal deposition, resulting in aggressive tooth resorption and alveolar bone loss, which may be misdiagnosed.

Key Words: Primary hyperoxaluria, oral manifestations, tooth resorption, oxalate crystal

Primary hyperoxaluria type 1 (PH1, Mendelian Inheritance in Man [MIM] 259900) is a rare, inherited autosomal recessive disease caused by defects in the metabolism of glyoxylate. Primary hyperoxaluria type 1 is caused by low, absent, or mislocalized activity of liver-specific peroximal alanine-glyoxylate aminotransferase (*AGXT* gene in chromosome 2q37.3), which leads to elevated urinary excretion of both oxalate and glycolate. On the other hand, type 2 primary hyperoxaluria (HP2, MIM 260000) is caused by mutation in the glyoxylate reductase/hydroxybutyrate reductase gene (*MIM* 604296) in chromosome 9.

These urinary abnormalities in PH1 produce urolithiasis and medullary nephrocalcinosis. Recurrent renal stones and/or progressive medullary nephrocalcinosis lead to progressive kidney damage and declining glomerular filtration and thereby produce an elevation above regular levels of plasma oxalate concentration and plasma calcium oxalate saturation. The resulting oversaturation provokes crystal deposits in the parenchyma of most solid organs, as well as in the bones, joints, and in the retina, defining the entity as systemic oxalosis.1–3

The prevalence and incidence of PH among the general population are difficult to determine and are underestimated because of their rare availability late after the onset of end-stage renal failure or even after renal transplants. Transplantations have, to date, been carried out on patients with PH1 because of the occurrence of end-stage renal failure. These patients are now eligible for 3 different strategies: isolated kidney, preemptive liver, and combined liver-kidney transplants. In recent years, combined liver-kidney transplantation has become the elected procedure for most patients.2

CLINICAL REPORT

A 34-year-old Brazilian white male patient was referred by his nephrologists to the dental clinic of the University Hospital Because of dental infections and dental mobility in the inferior incisor region. Patient’s medical history shows that he was under hemodialysis treatment because of an end-stage renal disease.

FIGURE 1. Periapical radiograph of the patient with PH1 and oxalosis showing periodontal destruction, aggressive external root resorption in several teeth, and extensive radiolucent image in the inferior incisor region.

FIGURE 2. A, Surgery of the lesion in the lower incisor region revealed extensive bone loss. B, Granular deposits removed from the lesion.
He had received a renal transplant, which failed after 18 months because the new kidney became similarly affected by nephrolithiasis. The oral clinic and radiographic examinations revealed periodontal destruction, aggressive root resorption in several teeth, and extensive radiolucency image in the inferior incisor region (Fig. 1). An important aspect to note is the preservation of the viability of the dental pulp even in the presence of an apical lesion. At the time of the removal of this lesion, a green granular deposit was identified (Figs. 2A, B). A routine section of the tissue showed granulomatous inflammation with foreign body reaction, associated with crystalline deposits. The individual deposits were roughly circular, with needle-like crystals radiating from their centers like spokes on a wheel. When viewed in polarized light, the crystalline deposits are green and exhibited a birefringent aspect, which were interpreted as calcium oxalate crystals. Calcium oxalate crystal in the periodontal ligament adjacent to the external root resorption area was also observed (Figs. 3A–C). The biopsy in the bone loss area was helpful in establishing the diagnosis of oxalosis due to oxalate crystals. The patient is still under dental treatment to improve oral health quality and is awaiting for a combined liver-kidney transplant.

**DISCUSSION**

In 1973, Glass found the presence of birefringent, needle-like crystals in the dental pulp, alveolar bone, and salivary glands, indicating oxalosis as a consequence of primary hyperoxaluria. Since then, there have been few recorded cases of oral manifestations of oxalosis. The crystal deposits cause radiolucent bone lesions, extensive periodontal destruction, and tooth resorption. Microscopically, crystals appear as birefringent, needle-like structures surrounded by chronic inflammatory infiltrate and giant cells, featuring an inflammatory response to a foreign body. Oral manifestations of oxalosis are rare but seem to be more common in the alveolar bone and periodontal ligament. Most remarkable findings, such as bone and tooth resorption, may be the result of chronic inflammation and the presence of osteoclastic cells surrounding oxalate crystals. In Figure 3, the calcium oxalate crystals can be seen in the periodontium adjacent to the external root resorption area. Some authors mentioned the importance of alternative diagnosis and the relation between oral manifestations of oxalosis and hyperparathyroidism and osteomalacia lesions. In the present case, no relation with other diseases was found.

With the increased usage of dialysis to maintain life in patients experiencing chronic renal failure, more acquired forms of oxalosis are likely to be seen, and therefore, periodontal and dental aspects of this disease will become a more common finding. Recently, Rinksma et al have published a case of a young man experiencing PH1, and they did observe similar oral findings.

Our case reinforces the importance of some basic aspects concerning the dentist’s role for the diagnosis and treatment of this disease. The accessibility of the oral cavity is of clinical importance, which is a good examination area when one is trying to establish a diagnosis of oxalosis.

**CONCLUSIONS**

Oral manifestations of hyperoxaluria are of particular interest because of the unusual location of oxalate crystal deposition, resulting in aggressive tooth resorption and alveolar bone loss, which may be misdiagnosed as a periodontal disease. This case of primary hyperoxaluria suggests that oxalosis should be considered in the diagnosis of bone loss associated with external root resorption. We recommend a multidisciplinary-team approach in dealing with patients with chronic renal disease.

**REFERENCES**