

Exodontia in patient with Gaucher's disease

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Gaucher's disease, the most frequent lipid storage disease, is an inherited disorder caused by partial or total deficiency of glucocerebrosidase, an enzyme responsible for the breakdown of glycolipids.⁽¹⁾ Type 1, or the non-neuropathic form, accounts for 95% of all cases. Found in children and adults, it affects internal organs (enlarged spleen and liver), causes blood (anemia and low platelet counts) and bone disorders associated with pain, infarction, demineralization, osteopenia and osteosclerosis, but no neurological impairment.⁽²⁾

The involvement of the maxillo-mandibular complex in Gaucher's disease is often asymptomatic and signs are only detected during routine radiological examinations.⁽³⁾

The main findings reported in the literature are: generalized osteopenia,⁽⁴⁾ loss of trabecular structure,^(4,5) lamina dura thinning,⁽⁵⁾ pseudocystic radiolucent lesions^(4,5) and apical root resorption of teeth adjacent to the lesions.^(4,5)

Yellow pigmentation of the oral mucosa⁽⁴⁾ and delayed eruption of permanent teeth⁽⁵⁾ are additional signs found in the few studies published so far.

Reports of procedures conducted in patients with Gaucher's disease are relevant because of the scarcity of information in the literature, especially about approaches, management and specific dental protocols for this population.

A 24-year old woman reported a family history of anemia and hemorrhages in two brothers, a femur fracture at age 9 years, blood transfusion during delivery of her only child and much bleeding during extractions and menstruation. She had a diagnosis of type 1 Gaucher's disease, was not splenectomized, and had been receiving enzyme replacement therapy for over one year when a dental examination detected a large and deep carious lesion affecting the pulp, and extensive crown destruction of tooth #26. Extraction was indicated (Figure 1).

Four days before the intervention, prophylaxis was initiated with one capsule of 500 mg amoxicillin every 8 hours. One day before the intervention, an oral antifibrinolytic drug was administered (2 tablets of 250 mg tranexamic acid every 8 hours) to prevent hemorrhages similar to previous extractions.

The tooth was extracted and the wound was sutured. Antibiotic (1 vial of 1-g sodium ceftriaxone), analgesic (1 vial of 500 mg/ml tramadol hydrochloride) and an antifibrinolytic drugs (1 vial of 250-mg tranexamic acid) were

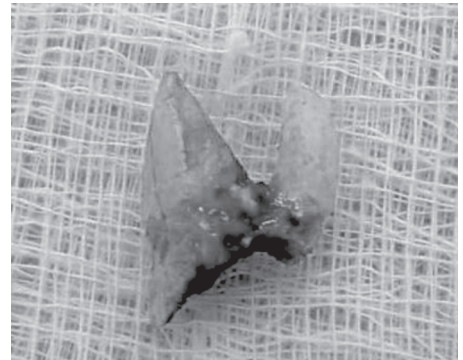


Figure 1 – Exodontia of tooth #26

diluted in saline solution and administered intravenously.

For home medication, oral antibiotics (1 tablet of 500-mg cephalexin every 6 hours) and an antifibrinolytic medication (2 tablets of 250-mg tranexamic acid every 8 hours) were prescribed.

After three days, the patient was examined again and a clot was found from tooth #25 to #28. The site was cleaned, a new suture was placed and the home medication previously prescribed was continued. After ten days, the wound was healing, but the suture could still not be removed. On the seventeenth day, healing was at an advanced stage, and the suture was removed. Two months after extraction, the area was completely healed and new bone formation was seen. Radiographic follow-up was continued for 3 years (Figure 2).

Thrombocytopenia, often found in individuals with Gaucher's disease, may cause spontaneous bleeding and hemorrhages. Non-splenectomized patients are more likely to have prolonged bleeding after surgical procedures,^(3,4) as in the case described here. Givol et al.⁽⁶⁾ evaluated the risk of hemorrhage during dental treatment of patients with Gaucher's disease. They used platelet transfusions and tranexamic acid as prophylaxis for patients with a high risk of hemorrhage. For average risk patients, desmopressin acetate (DDAVP) and tranexamic acid were administered and for low risk, only tranexamic acid. An antifibrinolytic drug was used as a prophylactic and therapeutic agent in this case.

The higher incidence of postoperative infection in patients with Gaucher's disease is explained by dysfunction of the reticulo-endothelial system.⁽⁷⁾ In our case, we chose to use amoxicillin for prophylaxis and cephalexin in the postoperative period to

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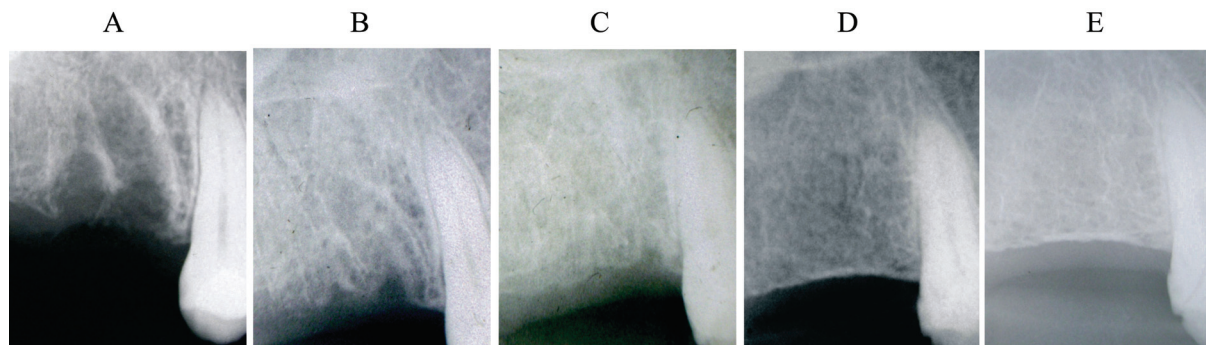


Figure 2 – Radiographic follow-up of region after extraction of tooth #26: A: 1 month; B: 2 months; C: 1 year; D: 2 years; E: 3 years

control infection and our results were good about two weeks after the extraction.

Bender and Bender and Regenye et al.^(8,9) found bone regeneration after extractions in patients with Gaucher's disease in one year and one year and two months. Those times were longer than in our case, as we identified new bone formation sixty days after extraction.

Healing in seventeen days, detection of new bone formation in only two months and the satisfactory alveolar closure confirmed during the three-year radiographic follow-up after extraction demonstrate the efficacy of the treatment protocol used in this case.

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