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Surgical management of a child suffering from osteogenesis imperfecta, treated with bisphosphonates - a case report

Małgorzata Lipczyńska-Lewandowska Dr, Piotr Osica PhD, Anna Janas-Naze PhD Assoc. Prof.

Małgorzata Lipczyńska-Lewandowska ORCID ID: 0000-0002-4985-7107

Piotr Osica ORCID ID:0000-0002-6436-5442

Anna Janas- Naze ORCID ID:0000-0001-6885-4457

Department of the Oral Surgery, Central Clinical Hospital, Medical University of Lodz

Head of the Department: Anna Janas- Naze, PhD, Assoc. Prof.

Corresponding author: Małgorzata Lipczyńska-Lewandowska

Department of Oral Surgery, Medical University of Lodz

92-213 Łódź, ul. Pomorska 251, tel. 42 675 75 29

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Abstract

The article presents a case of surgical extraction of impacted third molars in a 17 y. o. boy with osteogenesis imperfecta.

Key words: osteogenesis imperfecta, genetic disorders, surgical treatment

Introduction

Osteogenesis imperfecta (OI) also known, as Brittle Bone Disease, is a complicated, variable and rare genetic disorder of increased bone fragility, low bone mass and other connective tissue manifestations, inherited in dominant manner. A faulty gene that reduces either the amount or the quality of type 1 collagen in the body causes more than 90% of cases. The incidence of OI is 6/7 cases per 100 000 births [1]. The most characteristic features are recurring bone fractures and low bone density.

OI is currently classified according to modified Sileance's classification, which bases on the clinical symptoms, to 5 types: the mildest and most common, with no bone deformity (type I), the most severe and lethal form, where infants are born with multiple fractures (type II), progressive and deforming with very severe course (type III), moderate (type IV) and type V distinguished by a characteristic mineralization defect [2]. More than 90% of OI cases are determined by dominant mutation of the COL1A1 and COL1A2 genes, which reduce either the amount of the quality of type I collagen. And also recessive mutation of genes responsible for post translation processes of type I collagen and bone formation [3].

To prevent and treat osteoporosis and increase bone density, bisphosphonate drugs are used [4]. The side effect of such therapy can be bone necrosis, and as a consequence pathological mandible fractures, especially after teeth extractions [5].

The following article presents a case of a surgical extraction of impacted molars in general anaesthesia in a 17 y. o. boy with osteogenesis imperfecta treated with bisphosphonates.

Case report

A 17 y. o. patient has been referred to the Department of Oral Surgery of Medical University of Lodz by an orthodontist for extraction of impacted third molars, which was supposed to be the first step of a comprehensive surgical and orthodontic treatment. The orthodontist diagnosed the patient with crowding of teeth in the anterior region and posterior crossbite in the region of tooth 15 and 45.

Medical interview revealed that the patient suffered from osteogenesis imperfecta type I A (basing on the symptoms), and was treated for 4 years with intravenous administration of bisphosphonates. As a result of this therapy, the bone mass significantly improved, the bone density is just below normal and there were no fractures in the last few years. After consultation with the Head of Paediatrics and Metabolic Bone Diseases Department, the bisphosphonate therapy was postponed for the time of surgical treatment.

Extra oral examination showed no abnormalities. The intra oral examination also showed no signs of dentinogenesis imperfecta. In regard to the comprehensive treatment, the OPG and cephalometric x ray have been performed (Fig. 1), which revealed the impacted third molars.

Basing on the clinical and radiological examination, the patient and his parents were presented with a treatment plan including the surgical extraction of third molars in general anaesthesia. After obtaining signed consent from both the patients and his parents, the surgery was scheduled. All of the impacted molars were removed in general anaesthesia. The performed procedure, same in every case, is discussed basing on the extraction of the tooth 38. In the general anaesthesia, after cutting and preparing the periosteal flap, the bone was removed and the impacted third molar extracted. (Fig. 2, 3). The alveolus was rinsed with 0,9% solution of NaCl and sutured (Fig. 4). The intra and postoperative course was uneventful. Patient was discharged from the clinic in general good condition. Follow-up examination was performed the day after operation, which revealed the oedema of operated area. Patient reported mild pain, good healing was observed. In the 7^{th} day after the surgery, the sutures were removed, and the patient reported no pain.

Discussion

Osteogenesis imperfecta is a rare genetic disorder, the both mild types I A and B are the most often and are the result of mutation of genes COL1A1 or COL1A2 [1, 6]. The type I A is the mildest and most common form, there are only few fractures and there may be only few obvious signs of the disorder. There usually is little or no bone deformity. Height is less affected than in other types of OI. People with Type I are often similar in height to other family members. Muscle weakness, joint laxity and flat feet are common. Dislocations and sprains may occur as well as fractures. Hearing impairment and blue sclera may be present in 50% cases [6]. Radiological examination reveals significant mineralization deficiencies of the skeleton and bone fractures with excessive ossification in the points of fracture. The subtype A and B are differentiated by the lack of dentinogenesis imperfect [1]. Patients with OI should be subjected to the dental treatment as soon as possible [6].

Nowadays the OI is treated with the use of vitamin D and bisphosphonates, especially pamidronates, which increase the bone density [6, 7]. They may also have the antiangiogenic effect. Pamidronates decrease the bone pain and number of fractures; increase the growth and strength of muscles [8]. Chronic use of bisphosphonates can result in osteomalacia, the inflammation of the esophagus' mucosa, fever and neurotoxicity. Above all there is the possibility of disturbances in enamel and dentin mineralization, delayed eruption of teeth, and in consequence malocclusion [5, 7, 9, 10]. The literature indicates the intravenous administration of bisphosphonates with the mandibular and maxillary necrosis, due to their anatomic structure [5, 7]. After oral administration, the necrosis percentage is much lower (0,7 per 100 thousand patients). Osteonecrosis of the jaws is strictly associated with the bisphosphonate treatment (BON) and resembles the osteoradionecrosis. It manifests with pain or its absence, oedema, tooth mobility and bone exposition. It is usually associated with the surgical extraction of the tooth, but can also occur spontaneously due to dental diseases [5, 7, 11]. The results of BON can also be asymptomatic and found by accident on a follow up x ray. According to ADA, 94% of the patients with BON were treated with intravenous infusion, and in 6% it was administered orally [12]. BON after intravenous administration can be caused by higher dose of bisphosphonates and bioavailability. Also its course is much more severe [5]. One of the factors favoring the occurrence of BON is the pamidronate therapy lasting more than 2 years, corticosteroids therapy, type 2 diabetes, periodontal diseases, age above 60-65 and female gender [5, 7]. In case of the necessity of surgical treatment, it is recommended to proceed in the gentlest manner possible and the alveolus should be sutured. The patient should also be instructed to follow extreme hygiene regime

until the healing of the wound [5, 6].

Surgical treatment of the patients with OI is difficult because of many factors: tendency to bone fractures, malocclusion, poor wound and bone healing and of course the possibility of BON. In the described case there was a high risk of osteonecrosis due to the 4 year intravenous pamidronate therapy, and also the bone density below the normal level. Thanks to the conducting of the surgery in the least traumatic way possible, it resulted in no complications and the healing was uneventful. Surgical treatment was necessary for starting the orthodontic treatment, which aimed at eliminating the overcrowding and crossbite.

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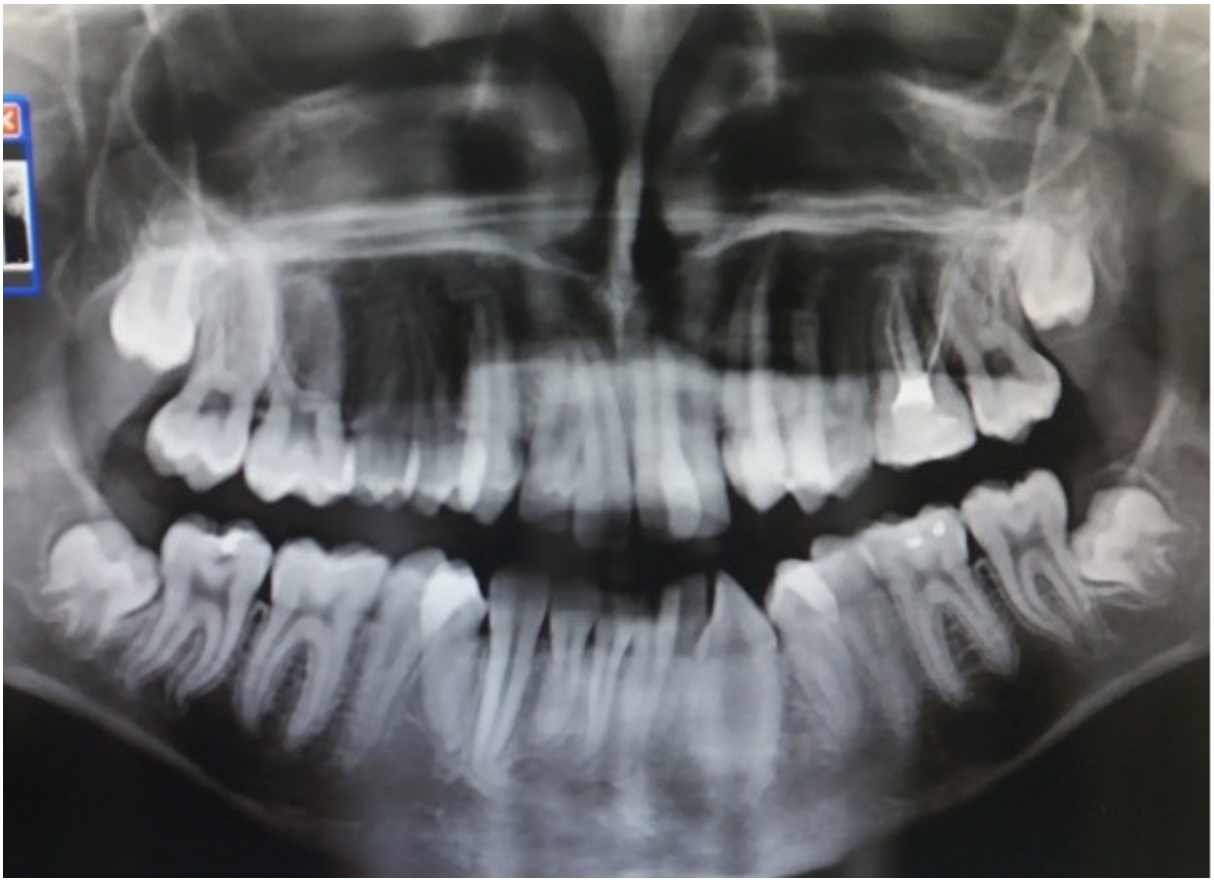

Fig. 1 Patients' OPG

Fig. 2. Totally impacted 38.

Fig. 3. Extraction of 38

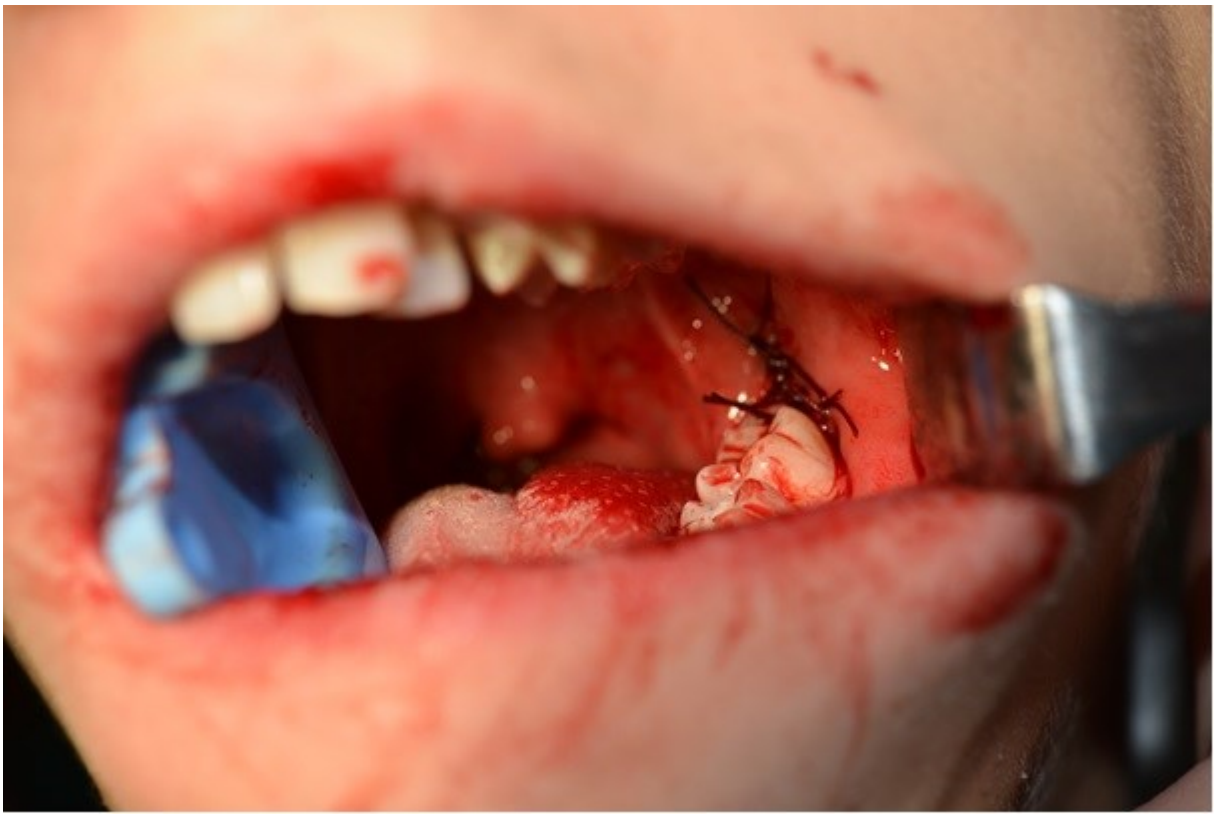

Fig. 4. Sutured wound.