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Preparation and dental surgical procedure in patients with *osteogenesis imperfecta* on the example of a 15-year-old patient with difficult tooth 38 eruption

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Abstract

The article presents a case of surgical extraction of a completely impacted tooth 38 under general anesthesia in a 15-year-old female patient with *osteogenesis imperfecta*, type III A.

Key words: surgical procedure; patients; osteogenesis; 38 eruption

Introduction

Osteogenesis imperfecta (OI) is a genetically determined disorder of the connective tissue. It is most often inherited as autosomal dominant, less frequently recessive, with a frequency of 1 in 10,000 births [1]. Its characteristic feature are recurrent fractures of long bones with progressive deformation as well as disturbed bone mineralization. With respect to clinical symptoms OI's current classification follows the Sileance division (modified in 2011), into 5

types: non-deforming form - the mildest and the most common (classic type I), lethal form with multiple fractures in the prenatal period (type II), progressive-deforming with severe course (type III A and B), moderate (type IV) and type V with calcification of the interosseous membranes and/or hypertrophic bone tissue [2]. Over 90% of OI cases are caused by a dominant mutation within the COL1A1 and COL1A2 genes coding for type I collagen, or by a recessive mutation of genes responsible for post-translational processes of pro-collagen type I and bone formation [3]. Type III A and B are inherited heterogeneously, as autosomal dominant in 75% of cases and as recessive in 25% [1].Treatment of OI involves drugs from the bisphosphonate group and vitamin D [4]. Administration of pyrophosphonate analogues and their derivatives may result in osteonecrosis, which may lead to pathological fractures of the mandible, especially after tooth extraction [5].

The article describes the preparation and surgical treatment of a 15-year-old girl with *osteogenesis imperfecta* of the progressive-deforming type treated with sodium pamidronate, who underwent surgical extraction of a completely retained tooth 38.

Case report

The 15-year-old patient was referred to the Department of Dental Surgery of the Medical University of Lodz from the Department of Dentistry of the Developmental Age by a specialized physician for the purpose of surgical extraction of a completely retained left third molar of the mandible, due to severe pain.

On the basis of general interview and clinical examination, it appears that the patient suffers from *osteogenesis imperfecta* type III A, according to the modified Sileance classification. The patient is treated with intravenous infusions of sodium pamidronate. The last infusion took place in February 2021. As a result of the therapy, the bone mass has improved slightly, while bone density has remained slightly below normal level. The most recent fracture (of the left jaw bone) took place two years ago. The patient has also been treated endocrinologically for hypothyroidism and pulmonologically for asthma.

The clinical study revealed numerous features of *osteogenesis imperfecta* type IIIA in the form of: short stature that prevented walking, scoliosis, blue sclera, skeletal deformities and the condition after multiple fractures of the long bones and the left jaw bone. The study also revealed dental abnormalities in the form of persistent deciduous teeth and tooth crowding as well as occlusal jaw narrowing. The teeth showed no signs of *dentinogenesis imperfecta*. The patient showed no symptoms of deafness.

After the clinical and radiological examinations, and following specialist consultations with an endocrinologist and an orthopedist, the patient and her mother were presented with a treatment plan including surgical extraction of the completely retained left third molar under general anesthesia (Fig. 1). The patient and her mother were provided with comprehensive information including the potential consequences of the procedure. After obtaining the written consent of the mother, and of the supervising physician, an endocrinologist and an orthopedist a one day procedure was scheduled for the patient.

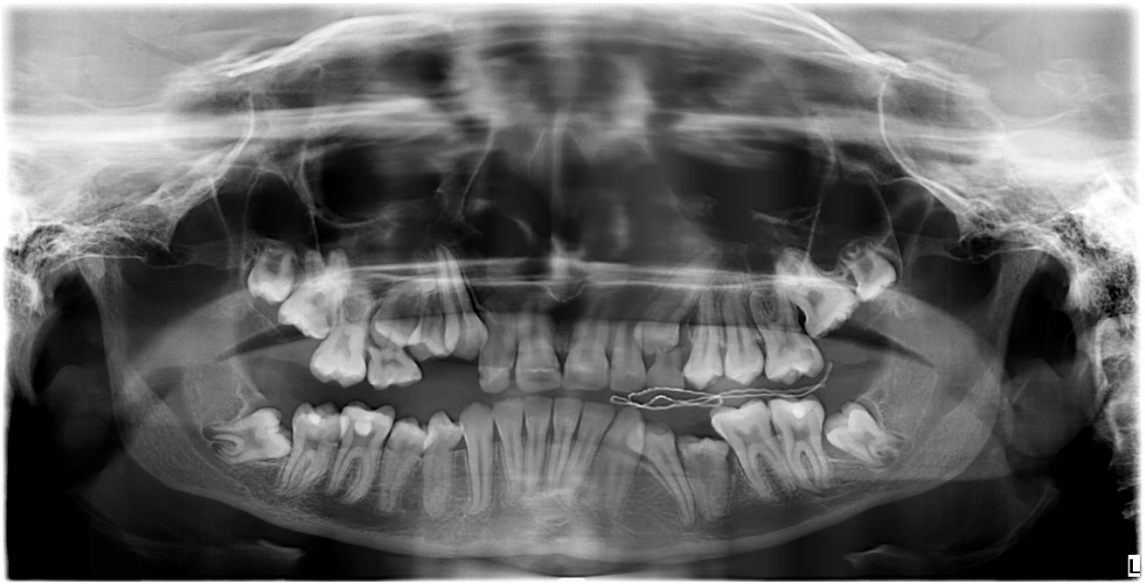

Fig. 1 Orthopantomogram

On the day of admission, the patient was in antibiotic cover (Augmentin 1.0 1 tablet every 12h). Under general intravenous anesthesia, the mucoperiosteal flap was detached around tooth 38 (Fig. 2), the vestibular bone plate was removed and the completely retained tooth 38 was surgically removed (Fig. 3). Sharp bone edges were smoothed with a cutter (Fig. 4). The intraoperative and postoperative course went uneventful (Fig. 5). The patient was discharged from hospital in good general condition under the care of her mother.

Fig. 2 The intraoral view of operative region.

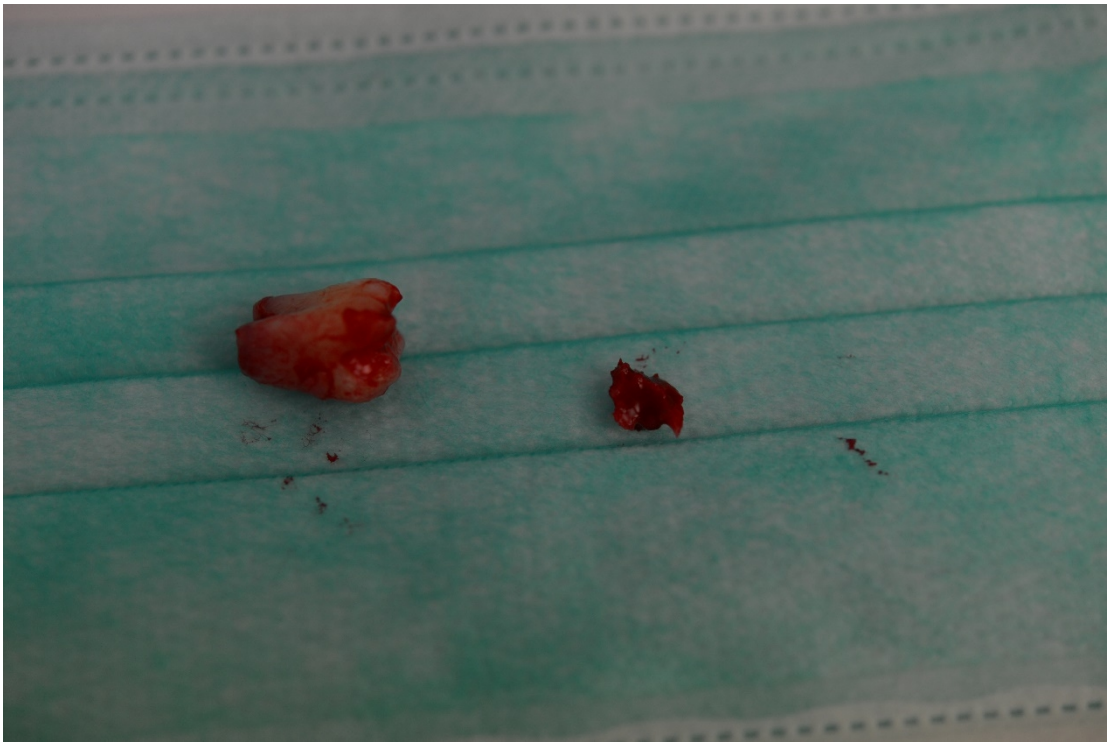

Fig. 3 The removed tooth 38.

Fig. 4 The flap was design and the bone was removed.

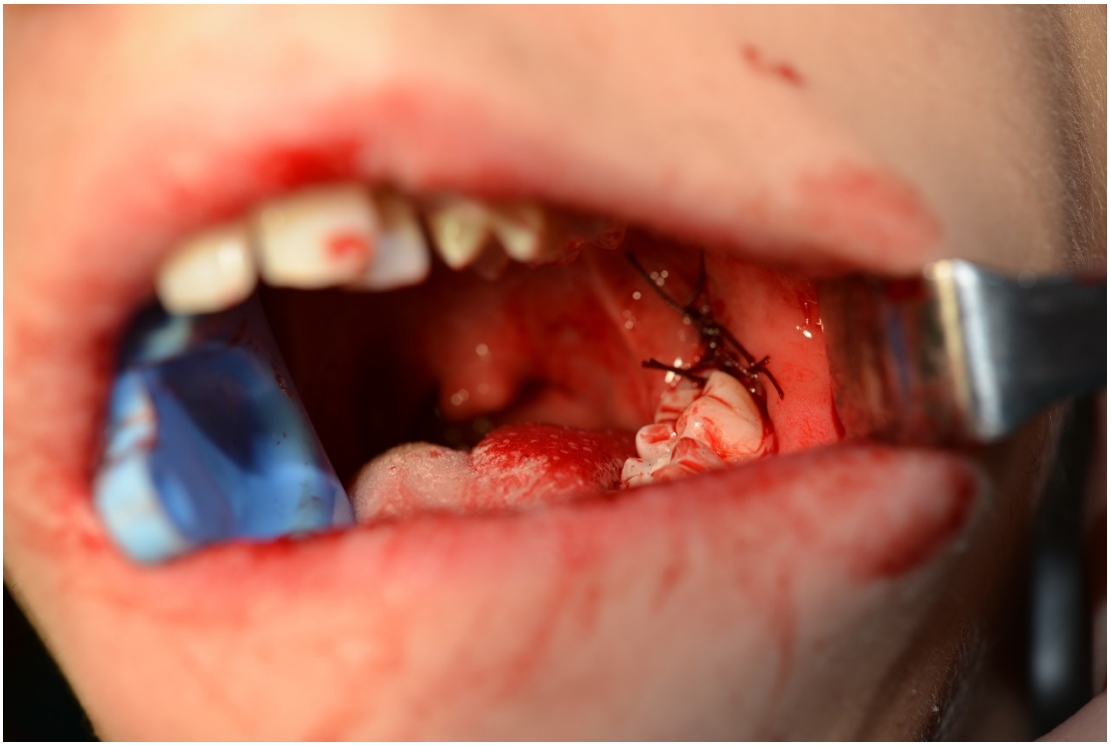

Fig. 5 The postoperative view.

On the first day after surgery, during the follow-up visit, a slight swelling of the operated area was visible, with no sensory disturbances from inferior alveolar and lingual nerves. The patient reported a slight pain in the operated area. Postoperative wound cleaning was performed, and the patient was prescribed compresses, a soft diet for 2 weeks and antibiotic therapy to continue for 10 days (Augmentin 1.0 1 tablet every 12 hours). On the 7th day after the surgery, the stitches were removed - the swelling and pain completely disappeared. The patient remains under continued care and control of the Department of Dental Surgery and the Department of Dentistry of the Developmental Age.

Discussion

Osteogenesis imperfecta is a rare genetic disease (1 in 10,000 live births). In 90% of cases it is a result of mutations in the COL1A1 or COL1A2 gene [1,3,6]. OI type III A is characterized by short stature of the patient which makes it impossible to walk, numerous deformities of the bones and body shape that increase with age, a tendency to scoliosis, increased bone fragility, laxity of joints, ligaments, congenital heart defects and asthma. Hearing impairment, blue sclera and *dentinogenesis imperfecta* occur in approximately 50% of cases [2,3]. Dental symptoms are more intense in primary dentition [6]. Bone fractures already occur in the prenatal period [2]. Radiological examinations show abnormalities in bone mineralization of the skeleton and bone fractures with excessive callus at the fracture site. The subtype A differs from B in that it shows no symptoms of *dentinogenesis imperfecta* [1]. Patients with OI type III A should remain under dental care, including conservative, periodontal, orthodontic and surgical treatment from an early age [6]

OI is currently treated with pyrophosphate analogues and their bisphosphonate derivatives. The most commonly used is sodium pamidronate - which has an anti-resorptive effect and an

anabolic effect on the bone tissue (increases bone mineral density) [6,7]. It is a strong osteoclast inhibitor [4]. It may also have an anti-angiogenic effect. Pamitronate reduces bone pain, lowers the number of fractures, and increases muscle growth and strength [8]. Unfortunately, it can disrupt proper bone remodeling and lead to microdamages [4]. Moreover, the mineralization of enamel and dentin is disturbed, tooth eruption is delayed and, as a consequence, leads to malocclusion [5,7,9,10]. The literature also reports a 4-fold increase in risk of osteonecrosis of the jaw associated with intravenous administration of bisphosphonates (pamidronate and zoledronic acid) and resulting from their anatomical structure [4,5,7]. After oral administration of bisphosphonates, the percentage of osteonecrosis of the jaws is lower (0.7 per 100,000 patients), however it leads to inflammation of the gastrointestinal mucosa, constipation or diarrhea [4,5]. Parenteral administration is associated with electrolyte disturbances, which is particularly dangerous in people with circulatory diseases [4]. Bisphosphonate therapy related jaw osteonecrosis (BON) has symptoms similar to osteonecrosis resulting from head and neck radiotherapy. Intraoral symptoms include pain or absence of pain, swelling, fistulas, loosening of the teeth, and exposure of the bones. It is most often associated with the surgical tooth extraction, but it can also occur spontaneously or due to diseases of the tooth tissues [5,7,11]. BON may also be asymptomatic and is detected accidentally in a control radiograph or only at the time of bone exposure [4]. According to the American Dental Association (ADA), 94% of patients with BON had been treated with intravenous infusions, and 6% took oral pamitronoate [12]. BON after i.v. may be caused by a higher dose of bisphosphonates and greater bioavailability. A post-i.v. course of BON is more severe [5]. The occurrence of BON is positively correlated with pamidronate therapy continued for over 2 years, with oral corticosteroid therapy, type 2 diabetes, periodontal disease, smoking, age of 60-65 years and female sex [5,7]. The etiology of BON is unclear, and the treatment is long-term and problematic [7, 10]. Proper hygiene and treatment of comorbidities significantly improves the prognosis after surgery [4].

Should surgical treatment be necessary, it is recommended to carry it out in the least traumatic manner (single extraction). The socket should be fitted with sutures, sharp bone edges should be smoothed and the patient should be recommended an extraordinary hygienic regimen until the wound is healed [5, 6].

Conclusions

Treatment of patients with OI type III A in dental surgery is difficult due to many factors: high susceptibility to bone fragility and its subsequent deformation, unfavorable occlusal conditions, impaired healing of wounds and bones, increased risk of bleeding and the risk of BON. In our case, there was a high risk of osteonecrosis associated with the 5-year intravenous pamidronate therapy, as well as below normal bone density / bone mass. Nevertheless, due to the atraumatic procedure, there were no complications and the healing was normal. Surgical treatment of the patient was necessary due to recurrent inflammation which caused pain and swelling. In view of such good treatment results we decided to follow up with surgical extractions of the remaining completely retained third molars using one-day surgical procedures. It should be noted that in the case of patients with OI type IIIA, close cooperation of dental surgeons with endocrinologists and orthopedists is required.

Conflicts of interest

There are no conflicts of interest.

Data availability

The references used to support the findings of this case report are listed in References.

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