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CASE REPORT

Exfoliation of non-resorbed primary incisors in a 4-year-old child – case report and literature review

Eksfoliacja niezresorbowanych siekaczy mlecznych u 4-latka – opis przypadku i przegląd piśmiennictwa

Ilona Wieczkowska[1], Anna Jarząbek[1], Magdalena Gońda-Domin[1], Karolina Węsierska[1], Rafał Rojek[1], Joanna Mikołajczyk[1], Hanna Bielawska-Victorini[2]

[1] Zakład Stomatologii Dziecięcej, Pomorski Uniwersytet Medyczny w Szczecinie, Polska
Department of Pediatric Dentistry, Pomeranian Medical University in Szczecin, Poland
Head: dr n. med. *J. Manowiec*

[2] Katedra i Zakład Ortodoncji, Pomorski Uniwersytet Medyczny w Szczecinie, Polska
Chair and Department of Orthodontics, Pomeranian Medical University in Szczecin, Poland
Head: prof. *K. Woźniak*

Abstract

Introduction. *Early exfoliation of non-resorbed tooth can be a symptom of severe systemic disease, even a fatal one.* ***Case description.*** *A 4-year-old boy presented with increased mobility of the lower incisors which resulted in missing 81; the tooth exfoliated with non-resorbed root six months earlier. Intra-oral examination revealed that mobility of 71 was more pronounced than that of the lateral incisors (it exfoliated non-resorbed within the next three months), while the remaining teeth were of normal mobility. Additionally, teeth 51 and 61 had pinkish crowns with horizontal gingival recessions. The boy was caries-free and had not experienced any dental trauma (DMF=0). His oral mucosa was sound, but he presented poor oral hygiene. The pantomogram showed a horizontal reduction of the maxillary front alveolar bone height. The patient was mouth breathing due to recurrent infections of the upper respiratory tract with adenoid hypertrophy. The skin of the hands and feet was normal. There was a case of inherited skeletal disorder in the patient's family. The patient was consulted by the paediatric haematologist, orthopaedic surgeon and laryngologist (adenoidectomy). The laboratory tests showed proper*

Streszczenie

Wprowadzenie. *Przedwczesna eksfoliacja zębów mlecznych bez resorpcji korzeni może być oznaką poważnej choroby ogólnej, nawet śmiertelnej.* ***Opis przypadku.*** *W pracy opisano przypadek czteroletniego chłopca, który zgłosił się do leczenia z powodu zwiększonej ruchomości mlecznych dolnych siekaczy, w wyniku której stracił on 6 miesięcy wcześniej ząb 81 (korzeń niezresorbowany). Badanie wewnątrzustne wykazało, że najbardziej zwiększona spośród dolnych siekaczy jest ruchomość zęba 71 (ząb wypadł w ciągu następnych 3 miesięcy), zaś ruchomość pozostałych zębów jest w normie. Ponadto korony zębów 51 i 61 miały różowawy odcień oraz współwystępującą poziomą recesję dziąseł. Wykluczono tło próchnicowe i urazowe przedwczesnej utraty zębów (PUW=0). Błona śluzowa była prawidłowa, jednak higiena jamy ustnej chłopca była niezadowalająca. Na pantomogramie zaobserwowano poziomą redukcję wysokości wyrostka zębodołowego szczęki w odcinku przednim. Chłopiec oddychał przez usta z powodu nawracających infekcji górnych dróg oddechowych oraz przerostu migdałków. Skóra dłoni i stóp była prawidłowa. W rodzinie pacjenta wystąpił jeden*

KEYWORDS:
tooth exfoliation, deciduous tooth, child (2-5 years)

HASŁA INDEKSOWE:
eksfoliacja zęba, zęby mleczne, dziecko (2-5 lat)

blood cell count and no signs of calcium-phosphate metabolism disorders. The boy stayed under dental observation for six years without any further abnormal exfoliations. ***Discussion.*** *Careful medical and dental diagnostics was required to exclude possible reasons for the tooth loss such as quantitative or qualitative neutrophil defects, metabolic or connective tissue disorders or neoplasia.* ***Conclusion.*** *This was a case of idiopathic early teeth exfoliation, which self-limited. Nevertheless, thorough diagnostics, adenoidectomy and oral hygiene improvement were fundamental for the diagnostic and treatment process.*

przypadek zaburzeń szkieletowych. W trakcie procesu diagnostycznego pacjent był konsultowany przez hematologa dziecięcego, ortopedę i laryngologa (zabieg adenoidektomii). Badania laboratoryjne nie wykazywały zaburzeń w zakresie morfologii pacjenta czy gospodarki wapniowo-fosforanowej. Podczas 6-letniej obserwacji stomatologicznej chłopca nie doszło do dalszych idiopatycznych eksfoliacji zębów. ***Dyskusja.*** *W opisanym przypadku niezbędna była dokładna diagnostyka stomatologiczna i medyczna, aby wykluczyć potencjalne ogólnoustrojowe przyczyny problemu, takie jak: niedobory ilościowe i jakościowe neutrofilów, zaburzenia metaboliczne lub dotyczące tkanki łącznej, czy nowotwory.* ***Wnioski.*** *Opisany przypadek idiopatycznej eksfoliacji zębów uległ samoograniczeniu. Jednak staranna diagnostyka, adenoidektomia oraz poprawa higieny jamy ustnej były ważnymi elementami procesu diagnostyczno-leczniczego.*

Introduction

In physiological conditions primary incisors should undergo root resorption at the age of 4-5 years, and at the age of 6-7 years the lower incisors should start to shed to be replaced by their permanent successors.[1] Exfoliation of primary teeth before the age of 4-5 years is regarded as premature.[2,3] Premature loss of anterior teeth usually affects eating, aesthetics and speech. It can also lead to tongue dysfunction and in consequence to serious orthodontic problems. Early loss of primary posterior teeth leads to space loss, also with orthodontic implications.

The most common causes of premature tooth loss are complications of early childhood caries or traumatic injuries. Rarely, this may be also due to self-inflicted trauma like in hereditary sensory neuropathies. Unfortunately, early tooth loss can be a symptom of aggressive periodontitis or periodontitis as a manifestation of systemic disease, e.g. neutropenia, Papillon-Lefèvre syndrome, Chédiak-Higashi syndrome, hypophosphatasia, Langerhans cell histiocytosis.[4] This fact accentuates the role of the dentist who should recognize premature tooth loss, and in idiopathic cases should start the careful multidisciplinary diagnostic process.

Case description

A 4-year-old Caucasian boy was referred to the Department of Paediatric Dentistry by a general dental practitioner because of the increased mobility of the lower incisors and missing tooth 81, which exfoliated six months earlier without any signs of root resorption (Fig. 1). This situation was not caused by any dental traumatic injury or pulp disease.

The medical history revealed that the patient suffered from recurrent infections of the upper respiratory tract with adenoid hypertrophy and enlargement of the palatine tonsils, but was otherwise healthy. The patient was on 50th percentile in stature-and-weight-for-age percentile chart. His diet was well balanced. It was noted

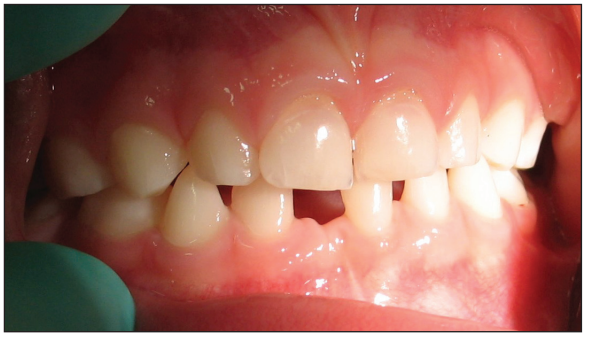

Fig. 1. *Clinical view of the patient's occlusion at the age of 3 years and 11 months – missing tooth 81.*

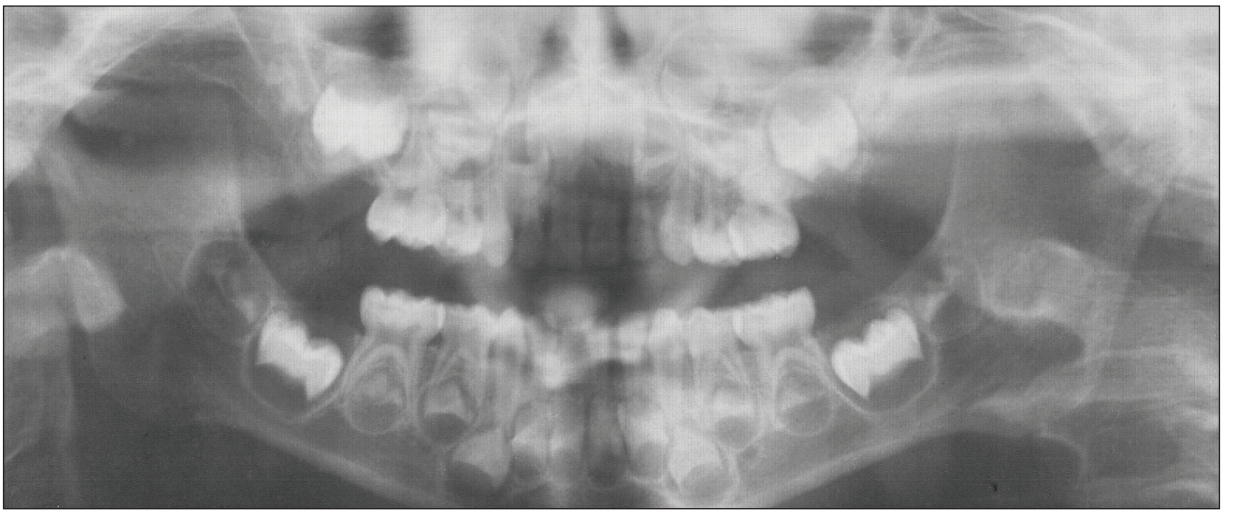

Fig. 2. *Initial pantomogram at the age of 4 years.*

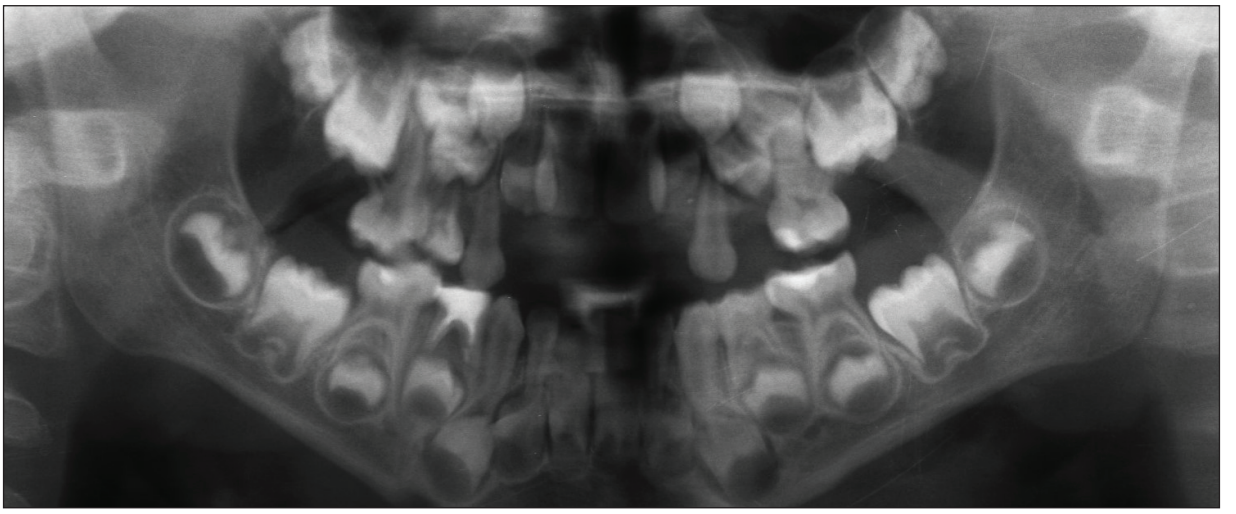

Fig. 3. *Pantomogram at the age of 5 years and 3 months.*

that the mother's brother suffered from inherited skeletal disorder – acromesomelic dysplasia.

The patient's dental history revealed that his first primary tooth erupted around the age of 6-7 months. There were no cases of premature tooth loss in the child's family.

Clinical examination showed that the skin had physiological elasticity without any lesions on the hands or feet. The joints had normal mobility.

Extra-oral examination revealed that the child had adenoid face and was mouth-breathing. Intra-oral examination revealed complete primary dentition without tooth 81. Tooth 71 mobility was confirmed to be more pronounced than the adjacent lateral incisors (it exfoliated within the next three months), while the remaining teeth were of normal mobility. Additionally, teeth 51 and 61 had horizontal gingival recessions accompanied by the pinkish colour of the crowns, but without increased mobility. The boy was caries-free, but presented poor oral hygiene. The colour and texture of gingiva was proper. The frenula were attached in non-irritating position. The depth of gingival sulci measured around nineteen teeth was within the physiological norm (0.5-3 mm) and probing did not provoke any bleeding. There were no pathological lesions on the oral mucosa.

The pantomogram showed a mild horizontal

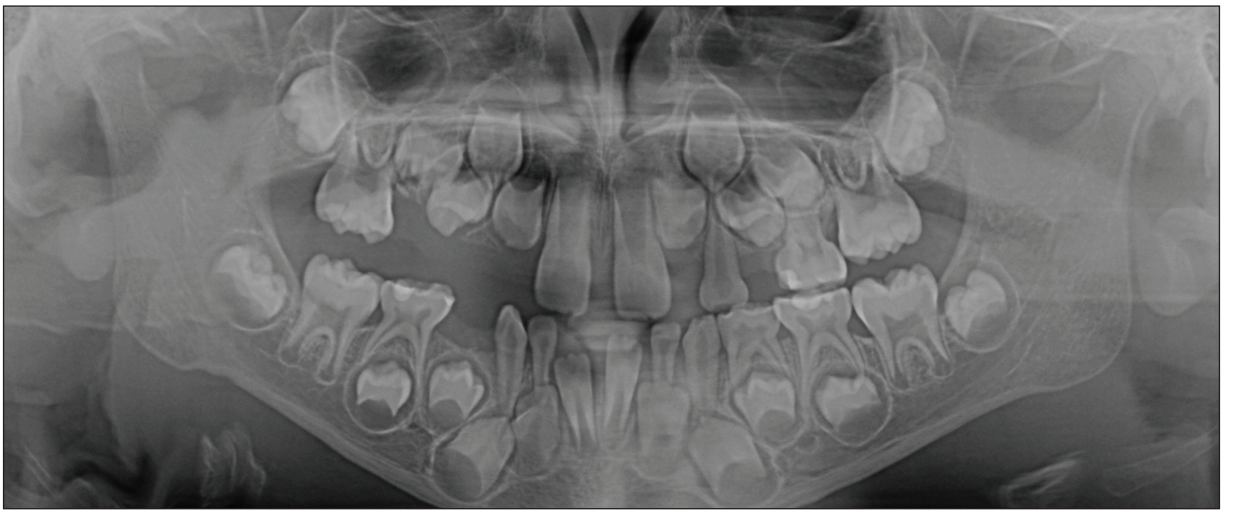

Fig. 4. *Pantomogram at the age of 6 years and 6 months.*

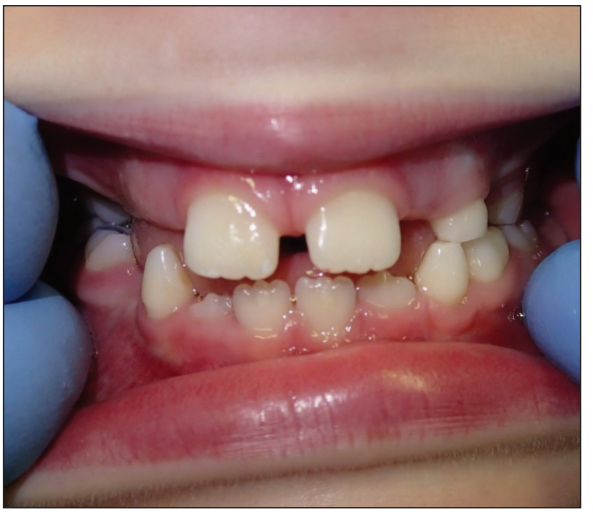

Fig. 5. *Clinical view of the patient's front teeth at the age of 7 years and 9 months.*

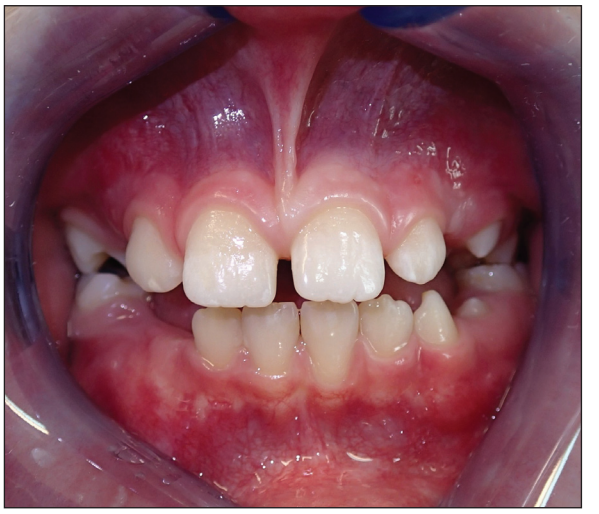

Fig. 6. *Clinical view of the patient's front teeth at the age of 9 years and 9 months.*

reduction of the maxillary front alveolar bone height. The stage of development of primary and permanent teeth corresponded to the patient's age. The size and shape of pulp chambers were undisturbed (Fig. 2).

The patient was monitored by a paediatrician and laryngologist due to recurrent infections of the upper respiratory tract and adenoid hypertrophy. In the first year of observation the boy underwent adenoidectomy. The child was also sent for consultation to the paediatric haematologist and to the orthopaedic surgeon. Blood investigation showed proper blood cell count and did not reveal any signs of calcium-phosphate metabolism disorders (proper levels of calcium, phosphate, alkaline phosphatase, thyrotropin, parathormone, 25-hydroxy vitamin D). The results of urine tests were also within the reference range. However, there was no possibility to check the phosphoethanolamine levels as no laboratory in the city had it on offer. The medical consultants recommended further dental observation as the medical examination and laboratory investigations did not reveal any systemic disease.

The patient had been under careful dental observation for six years. Three months after the first visit tooth 71 exfoliated without preceding root resorption. During the first year of observation the

boy had lost teeth 52, 51, 62, 61 due to a traumatic injury sustained on the playground. Tooth 53 was lost as a late consequence of this accident (Fig. 3). The patient was referred to an orthodontist for an application of a removable space maintainer. The boy also lost teeth 64 and 84 due to pathological resorption after complications of caries treatment undertaken by the general dental practitioner (Fig. 4). The paediatric dentist supported the general dental practitioner with further dental treatment and also focused on the improvement of the patient's oral hygiene.

To sum up, during these six years no other tooth was lost due to idiopathic causes (except the previously described exfoliation of tooth 71). The primary teeth were shedding in proper order with the roots physiologically resorbed. The condition of the successor permanent teeth was satisfactory (Fig. 5, 6). Oral hygiene improved, but was still not ideal. Unfortunately, the compliance in using the orthodontic appliance was not satisfactory and that resulted in loss of space. Now, the boy presents regularly for routine dental and orthodontic check-up visits. The re-education of breathing continues.

Discussion

The patient presented in this case report was generally healthy and fit. As the carious and traumatic background of the premature exfoliation of the lower incisors (even self-injury) were excluded, the dental team focused on exploring the periodontal condition of the patient and on studying the results of the medical consultations to confirm or exclude aggressive early onset periodontitis, and to look for systemic diseases that may lead to periodontitis and premature tooth loss.

The differential diagnosis of the possible causes of early tooth loss is presented below.

Aggressive periodontitis occurs in localized or generalized form with marked familial aggregation. Localized aggressive periodontitis (LAP) usually has a circumpubertal onset and the periodontal destruction is limited to the permanent incisors and first molars. Meanwhile, generalized aggressive periodontitis (GAP) occurs in people under 30 years of age and affects many teeth in addition to permanent incisors and first molars. In both forms, the amount of microbial deposits are inconsistent with the severity of the periodontal destruction. Nevertheless, elevated proportions of *Aggregatibacter actinomycetemcomitans* and, in some populations, *Porphyromonas gingivalis* are remarkable, as well as phagocyte abnormalities and elevated levels of prostaglandins (PGE_2, IL-1β).[4] However, cases of both LAP and GAP have also been reported in children with primary dentition.[5-9]

The systemic diseases that may manifest themselves with periodontitis and early tooth loss can be categorized as: quantitative or qualitative neutrophil defects, antibody deficiency disorders, metabolic disorders, connective tissues disorders, neoplasia and others. The review of systemic diseases causing early tooth exfoliation is modelled on one by *Cameron* et al.[10] with the modifications done by the present article's authors on the basis of contemporary medical publications.

Quantitative neutrophil disorders

Neutropenia – a significant reduction of the number of circulating granulocytes, which results in increased susceptibility to infections. Absolute neutrophil count (ANC) is less than 1500/mm^3. Severe and life-threatening form of neutropenia is when ANC<500/ mm^3 and this form is called agranulocytosis. In Kostmann syndrome ANC is less than 200/mm^3.

Neutropenia is in most cases acquired (e.g. drug-induced, due to viral infection, secondary to ionizing radiation or systemic lupus erythematosus), but it can also be inborn. Most cases of congenital neutropenia are part of genetic syndromes (e.g. Schwachman-Diamond syndrome, agamma- or dysgammaglobulinemia).

The congenital types may also appear in isolated forms: benign familial neutropenia, severe infantile genetic agranulocytosis (Kostmann syndrome) and cyclic neutropenia. In cyclic neutropenia the ANC is within the norms between the neutropenic episodes, which appear at regular intervals (14-36 days).

Oral manifestation:

Painful ulcerative necrotizing lesions of the oral

mucosa that infiltrate deeply and are hard to heal. The lesions can be accompanied by fever and enlargement of the mandibular lymph nodes.

Aggressive periodontitis (with typical erythematous gingivitis and bone destruction) that leads to premature tooth loss in both dentitions.

Oral mycosis (especially candidiasis).[4,11-14]

Qualitative neutrophil defects

Lazy leucocyte syndrome – neutropenia and defective neutrophil chemotactic response with abnormal inflammatory response.

Oral manifestation:

Aggressive periodontitis with bone and early tooth loss.[15]

Leucocyte adhesion defect – is a rare, genetic and often fatal disorder caused by inability to produce, or failure to express, integrin CD11/CD18, which is vital to adhere the leucocyte to the vessel wall at the infection site. Neutrophil defects include impaired migration and phagocytosis.

Oral manifestation:

Aggressive periodontitis starts during or just after eruption of primary teeth. Both primary and permanent dentition are affected, which leads to early teeth loss.[4,15,16]

Papillon-Lefèvre syndrome – an inherited, genetic disorder (mutation in cathepsin C-gene). Cathepsin C is a cysteine protease normally expressed in high levels in various cells (e.g. epithelium, polymorphonuclear leukocytes) that plays a role in degrading proteins and activating proenzymes in immune and inflammatory cells. Its dysfunction results in multiple functional neutrophil defects (myeloperoxidase deficiency, defective chemotaxis). This causes high susceptibility to infection with *Aggregatibacter actinomycetemcomitans* resulting in aggressive periodontitis with further sequela. Palmoplantar hyperkeratosis is a characteristic feature of this syndrome (also knees and elbows are affected).

Oral manifestation:

Severe aggressive periodontitis with pocketing and bone loss usually affects the primary and permanent teeth shortly after eruption resulting in early teeth loss.[2,14-18]

Chédiak-Higashi syndrome

Inherited rare autosomal recessive disorder that affects the production of organelles found in almost every cell. It affects mostly melanocytes, platelets and phagocytes. Neutrophils contain abnormal, giant lysosomes with invalid degranulation. The neutrophil chemotaxis is also decreased resulting in poor intracellular killing. Neutrophil granules fuse and form "megabodies". Patients develop severe neutropenia due to ineffective granulopoiesis.

General symptoms: hypopigmentation resulting in oculocutaneus albinism, strabismus, nystagmus, thrombocytopenia and infections of the respiratory system and the skin (result of neutropenia and decreased chemotaxis). Mental retardation and progressive neurologic abnormalities are also observed. Children often die before adolescence due to sepsis.

Oral manifestation:

Aggressive periodontitis with premature exfoliation of both dentitions and rapid alveolar bone loss.[2,14-16,19]

Down syndrome (trisomy 21, mongolism) – is a congenital disease caused by a chromosomal abnormality.

The characteristic physical features include: brachycephalic skull with a flat occiput and prominent forehead, hypotelorism and upslanting palpebral fissures with epicanthic fold. The profile is flat with flat nasal bridge and protruding tongue (mandibular prognathism). Other symptoms include: hypotonia, simian crease and gap between 1st and 2nd toes. Congenital heart disease and abnormalities of the alimentary tract may also be present, as well as retardation and mental deficiency of various degrees. Poor neutrophil chemotaxis and phagocytosis is reported and affects the condition of the periodontium.

Oral manifestations:

Characteristic orofacial appearance with large, protruding tongue affecting occlusion and high frenum attachments. High prevalence of periodontal disease, often generalized and rapid, with periodontal pockets and premature tooth loss is observed. The presence of *Aggregatibacter actinomycetemcomitans* and of *Prevotella*

intermedia is reported. Achieving proper oral hygiene may be difficult in mentally retarded patients.[2,14,15]

Acatalasia (acatalasemia). Autosomal recessive disorder resulting from deficiency of catalase enzyme, which is normally found in leukocytes, erythrocytes, hepatic and renal cells. The disease is endemic for Japan, but cases were also reported in Switzerland, Israel, Peru and Austria.

Oral manifestation:

Aggressive periodontitis with premature tooth loss.[2,10]

Metabolic disorders

Hypophosphatasia – a rare autosomal dominant or recessive disorder caused by mutation in tissue non-specific alkaline phosphatase gene. This leads to deficiency in alkaline phosphatase activity that disturbs osteogenesis (abnormal bone mineralization, skeletal anomalies) and cementogenesis (cementum hypoplasia or aplasia). This compromises periodontal ligaments attachment to cementum and results in premature tooth loss. In some patients an increased susceptibility to periodontal infections is also observed due to the presence of compromised periodontium.

Patients have low levels of serum alkaline phosphatase and high levels of: pyridoxal-5'-phosphate in serum and phosphoethanolamine in urine. There are six clinical forms of hypophosphatasia of various severity affecting the patient's lifespan, but the benign one limited only to the teeth is called odontohypophosphatasia.

Oral manifestation:

Premature tooth loss (sometimes limited only to the primary dentition) without clinical signs of inflammation and root resorption, and reduced alveolar bone height.

Enlargement of the pulp chambers and canals was also reported, as well as enamel hypoplasia, delayed dentine formation and delayed eruption.[2,3,15,20-23]

Connective tissue disorders

Ehlers–Danlos syndrome (type I, III, IV, VII, VIII) – an uncommon inherited, autosomal dominant disorder in collagen metabolism. It has been classified into eight major different types. General clinical manifestations of the disease are: joint hypermobility, skin hyperextensibility, and its fragility, and poor healing tendencies with "cigarette paper scars". Sometimes the syndrome is accompanied by cardiovascular and gastrointestinal complications, and that causes the poorest prognosis for the patient's lifespan (type IV=vascular).

Possible oral manifestation:

Periodontitis has been reported for Ehlers-Danlos syndrome type I, III, IV, VII and VIII, but only the last one is described in the medical literature as the "periodontal" type. Characteristic features of type VIII are: aggressive periodontitis with early loss of permanent teeth, fragility of the alveolar mucosa and gingival bleeding. This is the result of defective collagen which affects the quality of the periodontal junction as well as the oral mucosa.

Other possible orofacial features (type VIII): deep anatomic grooves and distinctive cusps, calcifications and pulp stones, irregularities of dentinal tubules, enamel hypoplasia. Fragility of gingiva and oral mucosa, higher frequency of TMJ disorders and marked tongue extensibility have also been noted.[2,14,24-27]

Marfan syndrome. A mutation in gene causes alternation in a glycoprotein forming part of the connective tissue matrix. This leads to disturbances including defects in: ocular lens suspensor ligament, blood vessel walls and periodontal ligament.

Orofacial manifestation:

Periodontitis with alveolar bone loss and presence of bacterial plaque. However, dental mobility is a result of periodontitis, not the syndrome itself.[14]

Erythromelalgia – a very rare episodic disorder characterized by continuous or episodic redness, elevated skin temperature, and burning fluctuating skin pain of the distal extremities (all symptoms together). The attacks can last from several minutes to a few hours with intensification during summer. Other risk factors for the attack may include: exercise, putting on shoes or gloves, placing the extremities close to heating appliances. The symptoms appear in early childhood.

Oral manifestation:

Alveolar necrosis and premature tooth loss,

probably due to the fact that the periodontal ligament is more vascular and less fibrous. The disease itself may also decrease the immune and inflammatory defences of the patient.[28]

Acrodynia (mercury toxicity). This intoxication, which is extremely rare now, is characterized by headache, insomnia, cardiovascular symptoms, pronounced salivation (ptyalism) and metallic taste.

Oral manifestation:

This toxicity causes linear pigmentation of the gingiva and exacerbates pre-existing inflammation that often leads to gingival/mucosal ulceration, alveolar destruction and even sequestration. Premature tooth loss can occur in extreme cases.[2,15]

Scurvy – is a deficiency in vitamin C, nowadays rare. Tooth loss is due to failure of proline hydroxylation and consequent reduction of collagen synthesis.[10]

Neoplasia

Langerhans cell histiocytosis (former: histiocytosis X) – it is a rare condition of unknown aetiology. The disease is caused by uncontrolled accumulation or primary proliferation of bone-marrow histiocytes (Langerhans cells) along with leukocytes, eosinophils, neutrophils, lymphocytes, plasma cells and giant multinucleated cells causing tissue destruction (via infiltration and replacement). It is more frequently located in skin and bones (also in other organs). Histiocytes are monocytes, dendritic cells and macrophages. Under electron microscope, Birbeck granules or "racket" bodies can be observed in these cells.

The clinical picture depends on the fact that different tissues and organs can be compromised and in disseminated form many of them are affected. Symptoms like fever, anorexia, weight loss, anaemia, haemorrhagic manifestations (petechiae on the trunk), asthenia and irritability may be reported.

Children and young adults are most commonly affected, although the disease can manifest itself at any age.

Oral manifestation:

Oral lesions (alveolar resorption accompanied by tooth loss) appear in all four quadrants with predilection to molars, and spreading mesially. Incisors involvement is infrequent and indicative of a negative prognosis. Pain and gum/bone swelling is characteristic and tumefaction is found under palpation, corresponding to the Langerhans cells accumulation. Other oral findings are: teeth mobility, erythema, spontaneous bleeding, halitosis and gingival lesions.

Radiographic picture – teeth "floating in the air" due to bone lesions that can also displace the teeth buds. The intraosseous lesions can be well defined with periosteal new bone formation or poorly marginated and badly defined.

Oral lesions can be the first symptom of the disease.[2,29-32]

Acute myeloid leukaemia

Leukaemia is malignant neoplasia of white blood cells (WBC) precursors. The main symptoms are: diffuse replacement of the bone marrow with proliferating leukemic cells and abnormal numbers and forms of immature WBCs in the circulating blood that also infiltrate tissues and organs. Two main types of the disease are: lymphocytic and myelocytic, while monocytic leukaemia is a subgroup. Leukaemia can be: acute (rapidly fatal, more "blast" cells in the blood), subacute and chronic (the abnormal cells in circulating blood are more mature). The disease causes the disturbance in the bone marrow components leading to anaemia (poorer oxygenation of the tissues resulting in their friability), leukopenia (reduction in non-malignant WBCs leading to increased susceptibility to infections) and thrombocytopenia (bleeding tendency).

Oral manifestation:

Oral and periodontal manifestations can be: leukemic infiltration of gingiva and less frequently alveolar bone, bleeding and oral infections (ulcerations). The infiltrated and enlarged gingiva forms gingival pockets, where bacteria can accumulate and that initiates secondary inflammation. The periodontal ligament (PDL) and alveolar bone may be also involved in acute and subacute leukaemia. In mice, this can lead to osteoporosis with destruction of the alveolar bone and PDL.

Increased tooth mobility may be an infrequent finding in children with acute lymphocytic leukaemia.

It is worth noting the long-term consequences of radiotherapy and chemotherapy in childhood like shortened and conical roots; that affects the crown-to-root-ratio and teeth's retention in the dental arch.[2,10,15]

Antibody deficiency disorders

Agammaglobulinemia or hypogammaglobulinemia – congenital (X-linked) or acquired immune deficiency resulting from inadequate antibody production due to deficiency in B-cells.

Oral manifestation:

Aggressive periodontitis is common in children diagnosed with agammaglobulinemia.[15]

Acquired immunodeficiency syndrome (AIDS) – destruction of lymphocytes causing susceptibility to opportunistic infections (also destructive periodontal lesions) and malignancies.

Oral manifestation:

Typical symptoms are oral infections: oral candidiasis, oral hairy leukoplakia, Kaposi's sarcoma, salivary gland diseases, oral warts, other oral viral infections, linear gingival erythema and necrotizing gingival and periodontal diseases.[15,33]

Apart from the described systemic diseases there are other conditions that may lead to early tooth loss. Among them, defects of dental hard tissues (isolated or accompanying systemic diseases).

Defects of dental hard tissues

Radicular dentinal dysplasia – the teeth have normal crowns, but with very short roots or they are rootless. This is accompanied by progressive obliteration of the dental pulp. Disturbed crown-to-root ratio and inflammation of the periapical tissues may result in early tooth loss.[2,10,34,35]

Singleton-Merten syndrome – a rare autosomal dominant disorder with significant variability in phenotype. The core manifestations are marked calcification of aortic arch and aortic valves, osteopenia and acro-osteolysis with muscle weakness and joint laxity. Dental anomalies are characteristic of this syndrome (delayed eruption and immature root formation, early loss of permanent teeth due to short roots, acute root resorption and aggressive bone loss).[2,36]

X-linked vitamin D-resistant rickets – uncommon metabolic disorder with low concentration of serum phosphate levels, elevated alkaline phosphatase, normal/elevated serum calcium and inappropriate levels of 1,25-dihydrohy-vitamin D. Apart from the general symptoms that lead to dental problems like delayed eruption, enamel hypoplasia, taurodontism and extremely large pulp chambers with structural defects of the enamel there may occur abscesses and early tooth loss.[2,10,37]

Coffin-Lowry syndrome – a rare x-linked disorder with psychomotor retardation, short stature, skeletal deformations, digit abnormalities (tapered fingers), large and soft hands. Oral and dental anomalies include: thick prominent lips, high palate, midline lingual furrow, hypodontia, microdontia, delayed eruption and early tooth loss (due to hypoplastic root cementum).[2,38]

Other diseases

The disturbance in Notch signalling pathway affects bone development and homeostasis in **Hajdu-Cheney syndrome**. Main characteristics of this syndrome include: acro-osteolysis, fibular deformities, osteoporosis with fractures and joint laxity combined with short stature, developmental delay and congenital heart/vessel defect. In orofacial region the disease manifests itself as facial dysmorphism, micrognathism, open sutures and Wormian bones, and periodontitis with tooth abnormalities and their loss.[2,39]

Cherubism is an autosomal dominant disease, which manifests itself early in childhood (2-5 years) and progresses until puberty when it begins to stabilize or regress. It is characterized by progressive, painless, bilateral enlargement of the mandible and/or maxilla. It is due to replacement of bone with multiocular cysts (fibrotic stromal cells, osteoclast-like cells). It also causes various consequences including ophthalmologic (upturned tilting of eyeballs, vision problems), respiratory, swallowing and speech implications.

Dental abnormalities include: displaced, unerupted or absent teeth. The teeth may appear

to be floating in cyst-like spaces. Malocclusion, premature exfoliation of the deciduous teeth and root resorption have also been reported.[2,40]

Early tooth loss and mobility may also accompany endocrine disorders like: **hyperthyroidism**, **hyperpituitarism** or **diabetes mellitus**.[2,41]

Finally, according to the classification of Periodontal Diseases from year 1999[4] there are systemic diseases of genetic origin that also manifest themselves with periodontitis (with its probable further consequence like tooth loss) for example: glycogen storage disease, Crohn's disease and Cohen's syndrome. **Glycogen storage disease (GSD)** is a metabolic defect that leads to glycogen accumulation in the body to disturb the glucose homeostasis in the blood. Delayed tooth eruption, aggressive periodontitis, oral ulcerations as a manifestation of neutropenia, and defect of neutrophil chemotaxis are observed in hepatic type of GSD.[42-45]

Crohn's disease is a chronic, inflammatory, intestinal process affecting the gastrointestinal tract from the mouth to the anus. The chief oral problems are aphthous-like ulcerations, diffuse lip and buccal mucosal swelling, oral cobblestoning and periodontitis (with the presence of *Aggregatibacter actinomycetemcomitans*).[46-48]

The diagnosis of **Cohen's disease** is based on the presence of following typical clinical features: mental retardation, microcephaly, facial dysmorphism (downward slanting and wave-shaped palpebral fissures, prominent nose, short and upturned philtrum with an open-mouthed expression and patient's face grimace during the act of smiling), slim, tapering extremities with relative truncal obesity, hypotonia, joint laxity and ophthalmic abnormalities. Neutropenia accompanies this disease affecting the patient's immune response and also leading to periodontitis (with the presence of *Aggregatibacter actinomycetemcomitans, Porphyromonas gingivalis* and *Prevotella intermedia*).[49,50]

According to the results of the medical consultations in the presented case of a 4-year-old boy, the condition of the skin, bones and the mobility of the joints were physiological and the patient was generally healthy and fit. The patient's medical problems were recurrent infections of the upper respiratory tract with adenoid hypertrophy and enlargement of the palatine tonsils, which are quite common in preschool children. The results of laboratory investigations confirmed that there were no haematological disorders and disturbances in the calcium and phosphate metabolism. On the basis of this information it was possible to exclude qualitative and quantitative neutrophil defects, metabolic disorders, connective tissues disorders, neoplasia and other systemic diseases causing early tooth loss. The periodontal examination did not reveal any other signs of aggressive periodontitis except for the premature exfoliation of teeth 71 and 81. Radiographic investigation showed only mild horizontal reduction of the anterior height of maxillary arch which was self-limited. As the symptoms resolved the idea of the microbiological investigation of the oral flora was abandoned. The patient was under dental observation for six years.

To summarise, this case and any case of idiopathic exfoliation of the teeth should be an alert to the dentist and should encourage him/her to initiate careful dental and medical diagnostics, because the underlying cause may be a severe systemic disease that could potentially be fatal.

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Address: 70-111 Szczecin, Al. Powstańców Wielkopolskich 72
Tel.: +4891 4661730
e-mail: zstdz@pum.edu.pl

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