

CLINICAL CASE

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Hereditary Gingival Hyperplasia – Case Report

Dziedziczny przerost dziąseł – opis przypadków

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Abstract

Hereditary gingival hyperplasia is a rare case in clinical practice. The study describes two cases of this pathology occurring in several members of one family. This disease is inherited in an autosomal dominant or recessive way. Patients affected by this disease require a thorough diagnosis. In addition to basic medical history and dental examination, outside and inside oral cavity, additional study may be needed. The first reported case concerns a young mother with enlargement of gum tissue, which also appeared in her children, mother and grandmother. In children, hyperplasia has caused the delay in the eruption of permanent teeth, diastema secondary, dental abnormalities, changes in facial appearance and problems with oral hygiene. Similar changes were observed in the second case described a father and son for whom gingival hyperplasia was a serious problem, with respect to functional and aesthetic disturbances, deteriorating quality of life. The aim of this study is to present the problems associated with enlargement of the gums and the difficulty in the treatment this disease which is not always successful (**Dent. Med. Probl. 2011, 48, 3, 443–449**).

Key words: gingival hyperplasia, inherited disease, gingivectomy.

Streszczenie

Dziedziczny rozrost dziąseł jest przypadkiem rzadko spotykanym w praktyce klinicznej. W pracy opisano 2 przypadki tej patologii, która wystąpiła u kilku członków jednej rodziny. Jest to choroba dziedziczona w sposób autosomalny dominujący lub recesywny. Pacjenci dotknięci chorobą wymagają szczegółowej diagnostyki. Oprócz podstawowego wywiadu oraz badania stomatologicznego zewnątrz- i wewnątrzustnego niezbędne mogą okazać się badania dodatkowe. Pierwszy z opisywanych przypadków dotyczy młodej matki z powiększeniem tkanki dziąseł, które pojawiło się również u jej dzieci, matki i babki. U dzieci rozrost ten spowodował opóźnienie w wyrzynaniu zębów stałych, diastemy wtórne, nieprawidłowości zębowe, zmianę w rysach twarzy oraz problemy z higieną jamy ustnej. Podobne zmiany zaobserwowano w drugim opisanym przypadku ojca i syna, dla których rozrost dziąseł był poważnym problemem z powodu zaburzeń funkcjonalnych oraz estetycznych pogarszających jakość życia chorych. Celem pracy jest przedstawienie problemów związanych z powiększeniem dziąseł oraz trudności w leczeniu tej jednostki chorobowej, które nie zawsze kończy się sukcesem (**Dent. Med. Probl. 2011, 48, 3, 443–449**).

Słowa kluczowe: przerost dziąseł, choroba dziedziczna, gingiwektomia.

Hereditary gingival fibromatosis is characterized by a mild hypertrophy of fibrous connective tissue of the gum. This disease is genetically determined, as the name itself indicates. It is passed on from generation to generation. It appears quite rarely with frequency of 1 per 175000 individuals. As a disease, it is also known as elephantiasis gingivae, fibromatosis diffusa gingivae, fibromatosis gingivae hereditaria, hypertrophica gingivae [1]. Hereditary gingival fibromatosis occurs as frequently in women as in men. The size of hyper-

trophy may differ within one family members [2]. Normally, it develops as isolated disease with different degree of intensity or as one of the symptoms of medical condition correlated with serious developmental disorders including mental retardation, such as syndromes: Zimmerman-Laband, Murray-Puretic-Drescher, Rutherford, Cross, Crowden, Goltz-Gorlin [3]. The medical condition almost always accompanied by gingival fibromatosis is Zimmerman-Laband Syndrome. There are 33 cases of this disease recorded in the world. This

heredity occurs extremely rarely and is autosomal dominant. Interesting fact seems to be the first change occurred was massive gingival hypertrophy noticed at a sick child by its mother. Other manifestations of the disease, apart from gums hypertrophy are cartilage disorders in ears and nose, hypoplasia or lack of finger nails or distal phalanges of hands and/or feet as well as mental retardation [4]. Hereditary gingival fibromatosis concerns permanent dentition mainly, rarely deciduous teeth, however it may interfere with deciduous teeth eruption. Some authors observed hyperplasia already at birth [5]. Gingival hypertrophy and deciduous teeth eruption appearing in parallel way can prove the participation of inflammatory cells in gingival hypertrophy pathogenesis [6]. Two forms of hypertrophy may be distinguished, localized and generalized which can appear in smooth and nodular form. Localized form is most often recorded on the protuberance of the jaw as well as the cheek side of the gum of lower jaw molars, however the generalized form is usually situated symmetrically on a cheek, tongue and palatal part of gums [7]. Abundant hypertrophies appear mainly in the nodular form, and localized in the smooth form. The mechanism of fibrosis pathology is not yet entirely known. Amongst the causes of hypertrophies there is transforming growth factor- $\beta 1$ (TGF- $\beta 1$), fibrinogenesis mediator, a cytokine participating in the processes of healing, regeneration, stimulates fibroblasts to proliferate, and later to fibrosis. Some authors, such as Martelli-Junior [8], undermine the participation of proliferation of fibroblasts in the pathogenesis of hereditary gingival fibromatosis. The increase of TGF β 1 stimulates metalloproteinase (MMP) inhibitors expression, which cause increased output of collagen. Others explain growth of fibroblasts proliferation with increased expression of synthase of fatty acids (FAS) [9]. In pathogenesis, there is also nuclear c-myc protooncogene that is taken into consideration [10]. Gums fibrosis is hereditary in autosomal dominant or recessive way. Usually, one parent carries the disease, a heterozygote, the other is a healthy homozygote. On average, the odds is 50% of chances of the ill as well as healthy offspring. Expression lability and vertical pattern of origin are observed. Genes responsible for gingival fibromatosis can be located on short arm of chromosome 2 – in its two loci in area 2p21-2p22 and 2p13-p16 [11]. With regards to histologic structure, in hereditary gingival fibromatosis the epithelium is hyperplastic and is of disturbed layered structure. Elongated system of epithelial appendages and connective tissue outgrowing it can also be observed [6]. Centers of small calcifications are also sometimes visible.

Hereditary gingival fibromatosis is characterized by increase of collagen, elastic, and oxytalan fibers of unparalleled course, as well as by small number of fibroblasts and vessels [12]. The histologic picture is non-specific therefore detailed clinical examination, family history and anamnesis are needed.

Case Reports

Case 1

The patient aged 42 reported to the Clinic of Oral and Maxillo-Facial Surgery in Lublin with children at which she noticed massive gums hypertrophy of the jaw and mandibles. The patient had been subjected to gingivectomy 10 years before also due to gingival hypertrophy. The result of the treatment was satisfactory, however it required few repetitions in a short period of time. At present, the patient is being prepared for prosthetic restorations. The gingival hypertrophy also appeared at her children: 12 year old son, and two daughters of 14 and 16. Confirming the occurrence of similar changes at other members of her family (mothers and grandmothers) was a relevant fact (Fig. 1). Children were subjected to interview and physical examination outside and inside oral cavity, as well as performing additional necessary examinations was recommended. At the interview, general diseases and taking any medicines were not confirmed. First child provided with examination was 14 year old daughter (Fig. 2). Apart from the aesthetic problem, the patient complained about discomfort in talking, taking foods and the problem in the correct dental hygiene. In the clinical research gums hypertrophy almost entirely covering all teeth crowns were stated, both in the jaw as well as the mandible. Gums were hard, with correct colouring, painless at touch. Deep alveolar

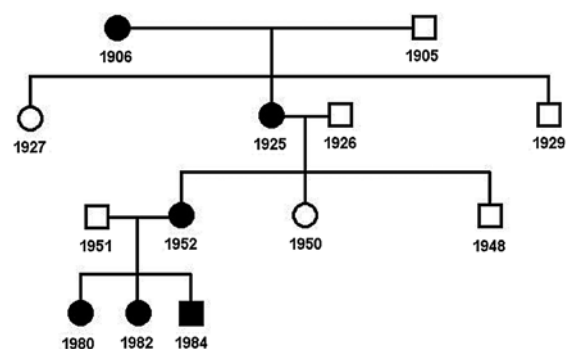


Fig. 1. Schematic of hereditary gingival hyperplasia in the family

Ryc. 1. Schemat występowania dziedzicznego przerostu dziąseł w rodzinie



Fig. 2. Gingival hyperplasia in a 14 year old daughter before the stage of oral hygienization

Ryc. 2. Przerost dziąseł u 14-letniej córki przed etapem higienizacji jamy ustnej



Fig. 3. Gingival hyperplasia in a 16 year old daughter before the stage of oral hygienization

Ryc. 3. Przerost dziąseł u 16-letniej córki przed etapem higienizacji jamy ustnej

pockets up to 6 mm accompanied the hypertrophy which much hampered dental hygiene, what was proven by high API 51% (Approximal Plaque Index) and PBI 1.7 (Papilla Bleeding Index). Numerous carious cavities, supra and subgingival tartar, dental gaps qualified the patient for the comprehensive treatment. 16 year old daughter was also provided with the examination (Fig. 3), as changes of a similar nature were stated. In this case changed appearance of face and occlusion abnormalities which required the orthodontic treatment were the most considerable problem. The patient reported problems in the dental hygiene, what was confirmed by high API 69% and PBI 2.1. At the 12 year old son (Fig. 4) a massive gingival enlargement disturbing correct carving teeth and negatively influencing aesthetics of the face was stated. Functional disorders hampering correct chewing foods and removing dental plaque off teeth were a serious problem. In the described case, the level of hygiene was the worst of all three children examined API 80% and PBI 2.3. Gums were red, painful, bleeding at the touch. At every-



Fig. 4. Gingival hyperplasia in a 12 year old son before the stage of oral hygienization

Ryc. 4. Przerost dziąseł u 12-letniego syna przed etapem higienizacji jamy ustnej

one examined, changes were not observed on mucous membranes of the oral cavity. Patients were also sent for blood count which did not reveal any abnormalities. Similarly, pantomography did not reveal the loss of the bone of the alveolar process. Because of poor condition of the dental hygiene at everyone examined the treatment began with prevention, motivation and correct briefing of dental hygiene. Also professional cleaning of teeth was conducted, scaling above and subgingival. After about 2 weeks the value of API and PBI indicators decreased at everyone examined what was tantamount to the improvement in dental hygiene however it was still not ideal. Patients were categorized for treatment of removing the outgrown gingival tissue (gingivectomy) connected with reshaping and improving the course of the gingival garland (gingivoplasty). In every case, the treatment was conducted in general anesthesia. In the course of surgeries fragments were taken for the histological examination which confirmed the fibrous gingival hypertrophy. After the treatment, a rinsing of the oral cavity was recommended to patients using chlorhexidine solution 2 times a day for 2 weeks. Control visits were scheduled for in 2 weeks, 1 and 3 months after the treatment. Result of the examination and of treatment was satisfactory at each of the siblings (Fig. 5b, 6b). An attempt to delay a relapse was also made using cryotherapy, however, since it did not bring expected results, it was aborted. Patients did not demonstrate the conscientiousness and the regularity in check investigations. Next visits confirmed the need for further treatment. In the clinical examination of three siblings unsatisfactory dental hygiene was stated, together with high API and PBI indicators. Also orthodontic treatment, which would lower the risk of the another gingival enlargement, was not commenced. Within the next 2 years, the siblings were subjected to gingivectomy 2 times in



a)



b)



c)

Fig. 5. Condition of the gums in a 14-year-old daughter: a) before the first gingivectomy, b) 7 days after the first gingivectomy, c) 7 years after the first gingivectomy and 6 months after the last treatment

Ryc. 5. Stan dziąseł u 14-letniej córki: a) przed pierwszym zabiegiem gingiwektomii, b) 7 dni po pierwszym zabiegu gingiwektomii, c) 7 lat po pierwszej gingiwektomii i 6 miesięcy po ostatnim zabiegu

the general anesthesia. The result, as before, was not satisfactory. Gingivectomy treatment was repeated two times in 3 years. At two patients, treatments of removing the excessively outgrown gum were performed at single teeth in clinical conditions. Youngest of siblings after a few gingivectomy treatments in general anesthesia did not give

consent to further treatment. Figs. 5c, 6c, 7b present the results after 7 years from the first gingivectomy and 6 months after last treatment. The best result was achieved at younger from sisters who demonstrated the greatest discipline in keeping dental hygiene and in accommodating to recommendations of the attending physician.



a)



b)



c)

Fig. 6. Condition of the gums in a 16-year-old daughter: a) before the first gingivectomy, b) 7 days after the first gingivectomy, c) 7 years after the first gingivectomy and 6 months after the last treatment

Ryc. 6. Stan dziąseł u 16-letniej córki: a) przed pierwszym zabiegiem gingiwektomii, b) 7 dni po pierwszym zabiegu gingiwektomii, c) 7 lat po pierwszej gingiwektomii i 6 miesięcy po ostatnim zabiegu

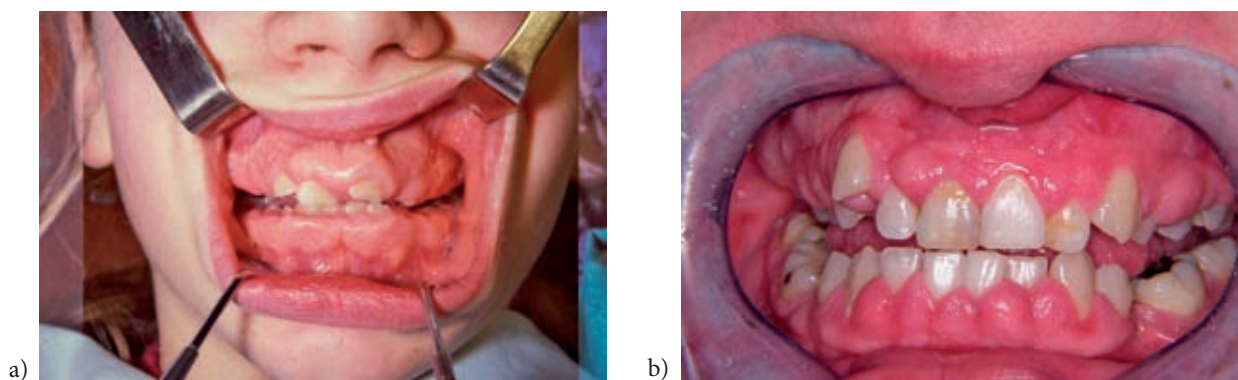


Fig. 7. Condition of the gums in 12-year-old son: a) before the first gingivectomy, b) 7 years after the first gingivectomy and 6 months after the last treatment

Ryc. 7. Stan dziąseł u 12-letniego syna: a) przed pierwszym zabiegiem gingiwektomii, b) 7 lat po pierwszej gingiwektomii i 6 miesięcy po ostatnim zabiegu

Case 2

The patient aged 42 years reported to the Clinic of Oral and Maxillo-Facial Surgery because of massive gingival hypertrophy (Fig. 8). The patient was referred to the Clinic in order to pre-prosthetic preparation. He did not report general diseases as well as took no medicines. Clinical examination revealed enormous gingival hypertrophy covering some teeth entirely both in the jaw as well as the mandible. The patient required surgical treatment. Firstly, he was suggested to have the excess of the gingival tissue removed (gingivectomy), followed by the extraction of remaining teeth not classified for further non-invasive treatment. Similar changes were also observed at his 9 year old son, whose main problem was hampered, disturbed carving of permanent teeth (Fig. 9). He also required gingivectomy treatment. However, patients did not give consent to further treatment and did not accept presented plan of the treatment.

Discussion

From a clinical point of view, the hereditary gingival fibromatosis stands out with relation to hypertrophies of different background. This ill-

ness is characterized by a mild hypertrophy of the gum. The tissue can be hard and in the correct colour or reddened-complicated with inflammatory condition. Excesses are usually massive, as in cases described, leading to forming of pseudopockets. Correctly performed pantomography does not reveal the loss of the bone of the alveolar process [6]. Because of big pseudopockets, the dental hygiene is hampered. All of described cases of gingival enlargements, the level of hygiene was unsatisfactory [3, 13]. The dental plaque lying in deep pockets caused an inflammatory condition of gums, and hence scarifying, bleeding and pain. The appropriate dental hygiene at onset could lead to reducing the excess, and can certainly delay a new hypertrophy after the treatment of removing the excess of the gingival tissue [2]. Problems with hypertrophy usually end after the extraction of the last tooth. Gingival enlargements can determine functional and aesthetic disorders. The majority of suffering people reports problems in talking, chewing foods, not to mention closing mouth [14, 15]. Large gums change the appearance of the face, leading to protrusion the upper lip, dental irregularities, secondary diastemata, open occlusion



Fig. 8. Gingival hyperplasia in 42-year-old man

Ryc. 8. Przerost dziąseł u 42-letniego mężczyzny

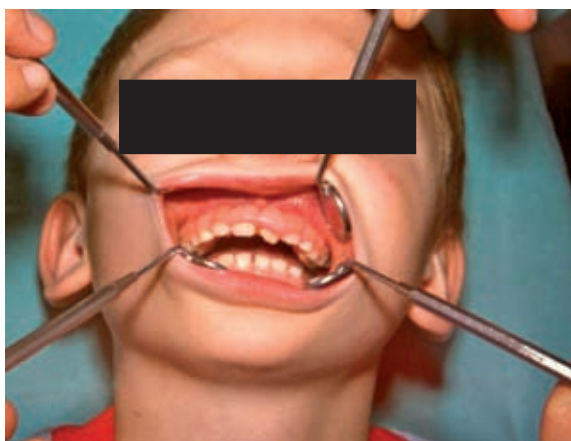


Fig. 9. Gingival hyperplasia in 9-year-old boy

Ryc. 9. Przerost dziąseł u 9-letniego chłopca

and or cross occlusion, which require orthodontic treatment later. In many cases, perhaps associated periodontal and orthodontic treatment may be necessary. Correct placing teeth in the dental arch allows reduction of the hypertrophy as well as delays subsequent hypertrophies after the surgical treatment [1]. Clocheret et al. [16] point at the need of the co-operation between the orthodontist and periodontologist in treating patients with the gingival hypertrophy. Other authors confirm it by obtaining satisfying effects of connecting treatment [1, 17]. Hereditary gingival fibromatosis overlaps with carving one's permanent teeth, rarely deciduous ones, what can make permanent teeth eruption difficult and prolong the retention of deciduous teeth in the oral cavity. As mentioned before, it can manifest as one of the symptoms of medical condition accompanied by serious mental and developmental disturbances. Katz et al. [18] describe the case of a boy with characteristic manifestations of the gingival hypertrophy, with delayed carving of permanent teeth and other deformations that look like hereditary Zimmerman-Laband Syndrome. In diversifying the isolated form of hereditary gingival fibromatosis, one should take into consideration hypertrophies triggered by inflammatory process, such as inflammatory hypertrophy of gingival papillae, where dental plaque is a main cause, hypertrophic inflammation of gums usually accompanying untreated diabetes, hypertrophies triggered

by permanent irritation, hormonal changes (puberty, pregnancy, diseases of systems, especially circulatory blood system). Borakowska et al. [19] describe a case of gingival enlargement at a patient with chronic myelomonocytic leukaemia. In clinical practice, many gingival hypertrophies are often triggered by the effect of medicines. There are three groups of medicines giving the side effect in the form of hypertrophy: anticonvulsant medicines (phenytoin, valproic acid), immunosuppressive medicines (cyclosporin) and blocking agents of the calcium channel (nifedipine) [20]. Gingival hypertrophies most often appear in the 3rd month of the administration. Based on the researched writing, it is possible however to claim, that average frequency of hypertrophy occurrences after the phenytoin, nifedipine, cyclosporin A amount to appropriately: 50%, 20%, 30% [21, 22]. Surgical removing of the excess of gingival tissue (gingivectomy) is one of the methods of hypertrophy treatment. It is possible to perform it in the classic way with scalpel or laser. One should remember about the due periodontal treatment which should be conducted at the very beginning, still before the surgical treatment [2]. Correctly conducted briefing of hygiene, appropriate motivation of the patient and implementing the effective treatment can lead to improvement of general condition of paradontium, not to mention the reduction of gingival hypertrophy [23].

Conclusions

Patients with gingival hypertrophy require the interdisciplinary care. Conducting the detailed anamnesis is necessary, as well as family background interview, clinical examination and additional necessary physical examinations which can clarify the reason for gingival hypertrophy. Curing hypertrophies often requires the cooperation of specialists of a few fields of medicine. One should remember that even correctly performed gingivectomy may not prevent from recurrent illnesses, what requires the need to repeat the treatment. However, eliminating irritants, the correct dental hygiene, regular professional cleaning of teeth and frequent control visits can lower the risk of hypertrophy recurrence.

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