

Combination therapy of basal-cell carcinoma in 31-year-old patient with nevoid basal cell carcinoma syndrome – Case study

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Abstract

Nevoid basal cell carcinoma syndrome (NBCCS) is a rare genetic disease that is manifested in a number of disorders concerning the skin, skeleton, cardiovascular and nervous systems. Various defects which can be observed at first contact with a patient visiting a dermatologist or dentist may help to diagnose this syndrome. Frequent problems with odontogenic cysts and metastatic basal-cell carcinomas result in patients being under the constant care of a specialist. This short study presents the case of 31-year-old patient with Gorlin-Goltz syndrome treated with combination therapy using CO₂ laser and photodynamic therapy.

Key words

Gorlin-Goltz syndrome, CO₂ laser, basal-cell carcinoma, photodynamic therapy

INTRODUCTION

Nevoid basal cell carcinoma syndrome (NBCCS) was described for the first time in 1960 by an American doctor, a clinical geneticist, as a set of defects with a prevalence of 1:57,000 [1]. This disease is inherited in an autosomal dominant pattern. The reason for the occurrence of numerous lesions of a basal-cell carcinoma character is the total loss of *PTCH-1* gene function [2, 3]. The protein encoded by the *PTCH-1* gene acts as a transmembrane receptor and contributes to the regulation of development and proliferation of stem cells in skin, skeleton and central nervous system [1]. Many symptoms of NBCCS have been described to-date and include, among others, defects in the central nervous system (*Meningioma*, *Astrocytoma*, cysts in the choroid plexus), optic tract (eyelid microcysts, hypertelorism, congenital cataracts), urogenital system (ovary cysts and fibromas in females, hypogonadism and gynecomastia in males, L- and U-shaped kidneys), skeleton (too high sinuses aeration within the face, increased skull diameter), as well as in the alimentary and cardiovascular tracts [4, 5, 6, 7]. Diagnostics criteria were established for rapid detection of this disease [8] in which the occurrence of two major criteria, or one major and two minor criteria are essential (Tab. 1.)

Patients suffering from NBCCS require multi-specialist neurological, gynecological, dermatological, and oral care, as well as a maxillofacial surgeon. The main dermatological problems are skin lesions of basal-cell carcinoma (BCC) nature. In order to obtain the best final esthetic effect, these patients require the combination of a few treatment methods during removal of BCC type changes. The methods used

Table 1. Diagnostic criteria in Gorlin-Goltz syndrome

Major diagnostic criteria	Minor diagnostic criteria
– basal-cell carcinoma: >2 or 1 under 20 years of age	– macrocephalia
– odontogenic cysts	– cleft palate, hypertelorism, domed frontal bones
– depressions in skin of hands: 3 or more	– high position of shoulder blades, pectusexcavatum, syndactyly
– calcification within cerebral falx	– sella turcica <i>multilamellar</i> calcification
– anomalies of the ribs: split, adherence	– ovarian fibroma, brainmedulloblastoma
– 1st degree kinship with the person with NBCCS syndrome	

in the therapy of this kind of skin lesions include surgical, CO₂ laser, photodynamic lamp with an application of 5-aminolevulinic acid, and cryotherapy using liquid nitrogen or carbon dioxide.

CASE STUDY

In February 2011, a male 31-year old patient attended Med-Laser for the first time due to numerous disseminated skin lesions within the whole body (Fig. 1). Dermoscopic examination gave rise to the suspicion of changes of a *basal cell carcinoma* nature, which qualified the patient for surgical therapy. Histopathological examination confirmed the preliminary diagnosis. Additionally, based on morphological features and an interview, the patient was diagnosed with nevoid basal cell carcinoma syndrome. Physical examination demonstrated such features as excessively domed frontal bones, low degree hypertelorism, moderate mental retardation, epidermal cysts of eyelids, as well as fine depressions in the skin of hands.

Because of anomalies within tooth apparatus, the patient was subjected to numerous procedures within the bones of jaw and mandible. In January 2012, extensive oral and

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Figure 1. Basal cell carcinoma

maxillofacial surgery was performed in Independent Public Clinical Hospital No. I (Samodzielny Publiczny Szpital Kliniczny Nr. I – SPSK I) in Lublin, Poland. This procedure involved removal of an abscessed cyst in the vicinity of tooth 37, extraction of teeth 36 and 37, multichamber cysts from the corner of the right mandible, extraction of gangrenous roots of teeth 16, 17 and 18, as well as removal of a large cyst in the right maxillary sinus. In addition, craniofacial radiographs taken before surgery revealed cerebral falx calcification.

Removal of the first changes using dermatosurgical procedures was not satisfactory and resulted in the formation of keloids (Fig. 2). Therefore, the decision was made to implement a combination therapy using CO₂ laser, photodynamic lamp and 5-aminolevulinic acid. In the first stage, the CO₂ laser and removal of the changes of a BCC nature by cutting and evaporation was carried out. The procedures were performed under local anesthesia with 1% lidocaine. The next stage involved the use of a photodynamic lamp and 5-aminolevulinic acid (ALA).



Figure 2. Keloid after surgical procedure

Three days after the laser therapy, 10% ALA preparation was applied to the lesion under occlusion for 3 hours. This region was then exposed to the photodynamic lamp at 630 nm wavelength for 20 minutes. The scab covering the wound after laser therapy, had to be removed prior to ALA application. The aim of this procedure was to facilitate the penetration of the preparation and polarized light to the cancer cells, which increased the therapy effectiveness. The patient was subjected to 3 PDT treatments using ALA, each at 3 week intervals.

The therapy carried out this way produced good therapeutic effects while maintaining a satisfactory esthetic result (Fig. 3). The patient remained under constant dermatological care, and attended regular dermoscopic examinations. The last visit was in June 2015.



Figure 3. Scar after combination therapy

DISCUSSION

Currently, there are numerous methods of basal-cell carcinoma therapy. The highest effectiveness is obtained for microsurgical therapy using the Mohs technique which seems to be indispensable in the treatment of lesions in difficult locations (*low-risk*BCC); in this therapy, less invasive methods do not guarantee successful prognosis [9, 10]. Unfortunately, the therapy is expensive and difficult to obtain due to the high cost of the equipment, as well as the small number of specialists who use this technique in Poland. Similar esthetic and therapeutic effects at much lower cost are guaranteed by methods such as classic surgery, CO₂ laser, cryosurgery, and photodynamic lamp. It is clear that the choice of therapy method depends on the skills of the dermatologist, available equipment, and the ability to predict the final result achieved by applying the above-mentioned techniques.

The experience of our centre shows that the combination of particular methods additionally contributes to improvement of therapy effectiveness [11]. This particularly relates to CO₂ laser and photodynamic lamp in combination with 10% 5-aminolevulinic acid. Moreover, the combination of these two techniques should be used only in the case of changes of a surface character, confirmed with clinical and histopathological examinations (*low-risk* BCC). Other forms of basal-cell carcinoma, i.e. nodular, cicatricial and ulcerative, among others, should be treated surgically.

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