


Research Article

Clinical Efficacy of 755 nm Laser Treatment of Lip Mucosal Pigmentation in Children with Peutz–Jeghers Syndrome

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Peutz–Jeghers syndrome (PJS) is a rare genetic disorder characterized by multiple gastrointestinal polyps and mucocutaneous pigmentation. Abnormal pigmentation typically develops in infancy or childhood. As PJS-related facial pigmentation can lead to psychological burden due to its effects on esthetics, treatment is required. Herein, we report on the efficacy and safety of treatment of lip mucosal pigmentation using a Q-switched 755 nm Alexandrite Laser in children with PJS, aged 2–12 years of age. A topical anesthetic was used prior to the application of laser therapy. A treatment efficacy of 100% was achieved with one to three treatments, with an excellent outcome achieved in five cases (62.5%) and a good outcome in three (37.5%). Recurrent pigmentation was observed in one case over the 6-month follow-up period. There were no adverse effects, such as scarring or hyperpigmentation or hypopigmentation. The treatment did cause pain, apprehension, and crying in some children, requiring special attention. Although our sample size is small, our findings do provide support of the high efficacy and safety of the Q-switched 755 nm Alexandrite Laser for the treatment of lip mucosal pigmentation in children with PJS. Further studies are required to confirm these findings.

1. Introduction

Peutz–Jeghers syndrome (PJS) is a rare genetic disorder characterized by multiple polyps in the gastrointestinal tract and abnormal mucocutaneous pigmentation [1]. Although serine/threonine kinase 11 (STK11) is the only identified causative gene for PJS, with autosomal dominant inheritance [2, 3], mutation at other loci has been reported [4]. Therefore, the genetics of PJS need further investigation. Clinically, however, treatment has focused on the gastrointestinal tract polyps. Yet, the mucocutaneous pigmentation, which characteristically appears in infancy and childhood, although not having a negative impact on health, can cause a significant mental burden due to its esthetic effects, particularly when present on the face and, thus, requires treatment.

The pigmentation is generally distributed around the mouth, nose, eyes, anus, and distal ends of the limbs. While pigmentation on the hands and feet tends to fade over time without treatment, pigmentation of the buccal mucosa persists

[5]. Herein, we report on the efficacy of the Q-switched 755 nm Alexandrite Laser for the treatment of PJS-related pigmentation on the lips and our experience with the treatment.

2. Methods

2.1. Study Sample. Our study sample included eight patients, 1 male and 7 females, ranging in age from 2 to 12 years of age (mean, 7.75 years), treated for PJS-related pigmentation of the lips on an outpatient basis at our hospital between February 2017 and February 2022. The onset of PJS occurred prior to the age of two years in all patients. Five patients had undergone endoscopic or surgical treatment for gastrointestinal polyps. Before treatment, the parents of the children signed the informed consent form.

2.2. Treatment Description. The Q-switched Alexandrite Laser (Cynosure, USA) was selected. Topical anesthetic was applied 30 min before the laser treatment, except for the

youngest patient who, at 2 years of age, was unable to cooperate with the process. The laser treatment consisted of 6.0–6.6 J/cm² of energy, delivered to a 3-mm area, at a pulse frequency of 2.0 Hz. Eyes were shielded during laser application and the laser beam was directed vertically to the target spots on the lip, with no overlap in the treatment area. The immediate appearance of a white or off-white reaction was monitored to avoid overtreatment. Antibiotic ointment, such as fusidic acid, was applied once daily after treatment until scabs peeled off naturally. Avoiding sunlight exposure and irritating foods required posttreatment. The number of treatments was based on the extent of pigmentation fading achieved, with a 4-month interval dependent between treatments.

2.3. Evaluation of Treatment Efficacy. Overall treatment efficacy was evaluated by comparison of before-and-after photographs by the treating physician and the parents, using the criteria described by Chang et al. [6] for the area of pigmentation decrease achieved: excellent, >75%; good, 50–75%; fair, 25–49%; and poor <25% or no change. The total efficacy rate was calculated as follows: (excellent cases + good cases)/total number of cases × 100%.

3. Results

3.1. Treatment Outcomes. An excellent outcome was achieved in five (62.5%) cases and a good outcome in three (37.5%), for a total treatment efficacy rate of 100%, with 2.12 treatments on average (range, 1–3), with two treatments used in 62.5% of cases (Table 1). Typical before-and-after photographs are shown in Figure 1. Over a 6-month follow-up, pigmentation recurred in one case. This child had undergone previous endoscopic intestinal polypectomy and pigmentation recurrence coincided with new intestinal polyps.

3.2. Adverse Effects. Most children reacted to pain by crying during treatment. Local erythema and mild swelling observed posttreatment recovered spontaneously, without complications. There was no scar formation or hyperpigmentation or hypopigmentation.

4. Discussion

In patients with Peutz–Jeghers syndrome, skin mucosal pigmentation is usually the earliest clinical manifestation. Histopathologically, increased pigmentation is localized to the basal and spiny layers of the epidermis, with no increase in the number of melanocytes in the basal layer [7]. Usually, the area of pigmentation is small (1–5 mm), with clear borders and black or dark brown coloring [8]. However, the lesions located on the face are aesthetically displeasing, which adds to the psychological burden on the patients and therefore are supposed to be taken seriously by dermatologists.

Previous treatments for PJS-related pigmentation, including surgical excision, electrocautery, liquid nitrogen

TABLE 1: Number of treatments and outcomes for the eight cases in the study sample.

Number of treatments	Number of cases	Excellent	Good
1	1	0	1
2	5	3	2
3	2	2	0

freezing, and dermabrasion are invasive and often cause adverse effects, such as scarring, pigment changes, or infection [4]. With advances in laser therapy, effective treatment of PJS-related mucosal pigmentation was reported with a high safety profile [9] although evidence for children is lacking. We selected the Q-switched 755 nm laser for treatment of pediatric cases of PJS as it is noninvasive and provides a selective destruction of melanosomes in skin melanocytes with minimal damage to other cells and tissues and results in particle fragments removed by phagocytosis [10].

In our series of eight cases, the total treatment efficacy rate of 100% included excellent outcomes in five cases and good outcomes in three, with no adverse effects. Considering the youngest child was 2 years old, we deem that the Q-switched Alexandrite Laser to be safe and effective for children, with the highest treatment effectiveness achieved with three treatments, a short treatment course. Moreover, over the 6-month follow-up, recurrence was observed in one case, which may have been associated with the development of new intestinal polyps. Considering the small sample size of our study and the short follow-up period, further studies are needed to determine the recurrence rate and the possible association with intestinal manifestations of PJS.

A few studies have evaluated that the efficacy of Q-switched Alexandrite Laser for the treatment of PJS-related lip mucosal pigmentation is study samples of adults or including a large age range. Xi et al. [10] reported disappearance of lip pigmentation in 14 adult patients, with no recurrence or complications over a two-year median follow-up. Li et al. [11] observed 43 patients aged 5–43; they reported excellent outcome in 23 cases (55.8%) and a good outcome in 20 cases (44.2%) after three treatments, for a total efficacy rate of 100%, with no adverse effects over a one-year follow-up. This result conformed to that of our study. By comparing children with patients within other age groups, we found that no serious complications were available during the observation of the two groups of patients after treatment, indicating the safety of Q-switched Alexandrite Laser for all ages. And in previous studies and this study, all the patients achieved significant efficacy of 100% within 3 treatments, which indicates that a definite effect on patients of all ages is available in Q-switched Alexandrite Laser and it takes effect quickly. However, further studies with a larger sample size and a longer follow-up period are required to confirm the efficacy of the Q-switched Alexandrite Laser for the treatment of PJS-related pigmentation in children.

Although limited, we provide a summary of our experience treating mucosal pigmentation in children with PJS using a Q-switched Alexandrite Laser. First, children tended



FIGURE 1: (a) Labial lesions of patient 1 before laser treatment. (b) Labial lesions of patient 1 after one laser treatment. (c) Labial lesions of patient 2 before laser treatment. (d) Labial lesions of patient 2 after 2 laser treatment.

to be apprehensive of the treatment, struggling, or crying, which required the use of protective eye shields to ensure eye protection during laser application. Second, the application of topical anesthetic partially reduced pain; however, in most cases, reassurance and safe restraint were required. Third, the mucous membrane of the lips is more vulnerable to damage during laser treatment, compared to the surrounding skin. As the skin is thinner in children than adults, laser energy should be increased gradually, based on the treatment response. Fourth, using the white or off-white reaction of the local skin ensures that areas of lighter pigmentation are not overtreated, avoiding local tissue damage. Fifth, topical antibiotic ointment should be applied after treatment, with parents ensuring that scabs peeled off naturally to prevent scarring, as well as protection from the sun and avoidance of irritating foods during the recovery period.

In conclusion, the Q-switched Alexandrite Laser can provide a high-efficacy and safe treatment for buccal pigmentation in children with PJS, with moderate-to-excellent outcomes obtained over a short treatment course, with no adverse effects. However, pain during treatment must be considered. Future studies are needed to confirm the long-term recurrence rate and the relationship between the recurrence of abnormal pigmentation and gastrointestinal polyyps.

Data Availability

Relevant clinical data are included within this article.

Conflicts of Interest

The authors declare that they have no conflicts of interest.

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