CASE REPORT – OPEN ACCESS

International Journal of Surgery Case Reports 11 (2015) 124–128

Contents lists available at ScienceDirect

International Journal of Surgery Case Reports

journal homepage: www.casereports.com

PHACES syndrome: Diode laser photocoagulation of intraoral hemangiomas in six young patients

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ARTICLE INFO

Article history:
Received 12 December 2014
Received in revised form 23 March 2015
Accepted 24 March 2015
Available online 27 March 2015

Keywords:
Laser treatment
Rare disease
Vascular tumor

ABSTRACT

INTRODUCTION: The acronym PHACES describes the association of posterior fossa malformations, facial hemangiomas, arterial anomalies (cardiovascular or cerebrovascular), coarctation of the aorta and cardiac defects, eye abnormalities, and sternal or ventral defects. In this study we report on 6 patients affected by the PHACES syndrome and showing 34 intraoral hemangiomas (IH), treated by diode laser photocoagulation (DLP).

CASE PRESENTATION: IH appeared as red-bluish soft masses, smooth or lobulated, from a few millimetre to several centimetres in size, covered by intact mucosa and blanching on pressure. IHs were treated by DLP with 320 μm fibres at a wavelength of 800 ± 10 nm. The diode laser techniques applied were: Transmucosal DLP (DLTP), a no-contact technique in which laser energy is delivered by a flexible optic quartz fiber, which is kept 2–3 mm apart from the lesion, and Intralesional DLP (DLIP), in which the fibre is introduced into the lesion through a transmucosal access. DLTP was used for 20 flat, superficial IHs and, after a variable number of laser sessions (average = 3) depending on the size of the lesion, 65% completely regressed, while in the remaining 35% shrinkage of the lesion was achieved with minor and few complications.

The remaining 14 deep/multi-lobulated IHs were treated by DLIP, resulting in complete regression of 79% of them.

CONCLUSIONS: DLP techniques are an effective and minimally invasive procedure for IH in patients with PHACES, in consideration of the multiple lesions to treat, of the necessity of multiple interventions and the higher compliance of the patients.

1. Introduction

PHACES is an acronym which refers to a rare syndrome characterized by Posterior fossa defects, facial hemangiomas, arterial lesions, cardiac abnormalities, eye anomalies and sternal cleft [1,2]. Firstly described by Frieden in 1996 [1], this syndrome shows predilection for the female gender (M:F = 9:1) [3,4]; there is no evidence of familial tendency [5].

Single/multiple facial hemangiomas are the most important clinical manifestation of PHACES. Hemangiomas manifest during the first weeks of life as teleangiectasias or erythematous plaques; subsequently, they increase in size (>5 cm), and converge together, occupying one or more distinct segments: fronto-temporal, maxillary, mandibular or fronto-nasal [6]. In 80% of the cases, hemangiomas in the mandibular region are associated with sternal clefts [7] and cardiovascular abnormalities [2,7,8]. Hemangiomas could completely or partially regress during the first decade, thus acquiring a port-wine like aspect.

The most common extracutaneous features are: cerebrovascular, structural brain and cardiovascular abnormalities [9].

Cerebrovascular anomalies consist in abnormalities of major cerebral arteries and are divided into four categories: dysplasia, narrowing, aberrant course/origin and persistence of embryonic anastomoses [8].

A spectrum of congenital structural brain abnormalities have been described in the PHACES syndrome, the most common of which involving the cranial posterior fossa [8].

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http://dx.doi.org/10.1016/j.ijscr.2015.03.045

Table 1
Diagnostic criteria for PHACES syndrome.

Organ system	Major criteria	Minor criteria
Cerebrovascular	Anomalies of major cerebral arteries: Dysplasia[a] of the large cerebral arteries[b] Arterial stenosis or occlusion with or without moyamoya collaterals Absence or moderate to severe hypoplasia of the large cerebral arteries Aberrant origin or course of the large cerebral arteries Persistent trigeminal artery Saccular aneurysms of any cerebral arteries	Persistent embryonic artery other than trigeminal artery: Proatlantal intersegmental artery (types 1 and 2) Primitive hypoglossal artery Primitive otic artery
Structural brain	Posterior fossa anomaly Dandy-Walker complex Unilateral/bilateral cerebellar hypoplasia/dysplasia	Enhancing extra-axial lesion with features consistent with intracranial hemangioma Midline anomaly[c] Neuronal migration disorder[d]
Cardiovascular	Aortic arch anomalies: Coarctation of aorta Dysplasia[a] Aneurysm Aberrant origin of the subclavian artery with or without a vascular ring	Ventricular septal defect Right aortic arch (double aortic arch)
Ocular	Posterior segment abnormalities: Persistent fetal vasculature (persistent hyperplastic primary vitreous) Retinal vascular anomalies Morning Glory disc anomaly Optic nerve hypoplasia Peripapillary staphyloma Coloboma	Anterior segment abnormalities: Sclerocornea Cataract Coloboma Microphtalmia
Ventral or midline	Sternal defect Sternal cleft Supraumbilicam raphe Sterna defects	Hypopituitarism Ectopic thyroid

[a] Includes kinking, looping, tortuosity, and/or dolichoectasia.
[b] Internal carotid artery, middle cerebral artery, posterior cerebral artery, or vertebrobasial system.
[c] Callosal agenesis or dysgenesis, septum pellucidum agenesis, pituitary malformation, or pituitary ectopia.
[d] Polymicrogyria, cortical dysplasia, or gray matter hetrotopia.

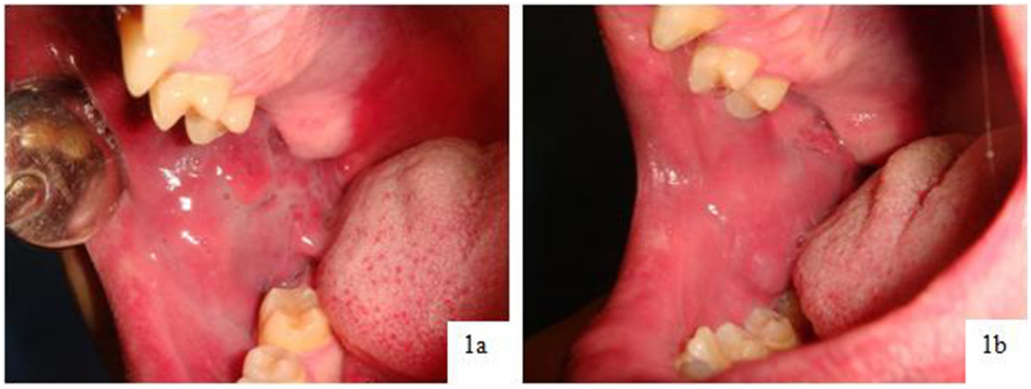

Fig. 1. IH on the right cheek before (1a) and after Diode Laser Transmucosal Photocoagulation (1b).

Fig. 2. A gingival lesion in region 2.1–1.8 before laser treatment (2a), after 2 Diode Laser Intralesional Photocoagulation sessions (2b) and after complete healing (2c).