Journal of Pediatric Surgery Case Reports 23 (2017) 9–14

Contents lists available at ScienceDirect

Journal of Pediatric Surgery Case Reports

journal homepage: www.jpscasereports.com

Peri-orbital lymphangioma treated by lymphatic-venous anastomosis with indocyanine green lymphography analysis

Motoi Kato, M.D. [a, *], Shoji Watanabe, M.D. PhD. [a], Takuya Iida, M.D. PhD. [b], Azusa Watanabe, M.D. PhD. [a], Furuya Megumi, M.D. [c]

[a] Saitama Children's Medical Center, Plastic and Reconstructive Surgery, Saitama, Japan
[b] University of Tokyo, Plastic and Reconstructive Surgery, Tokyo, Japan
[c] Mie University, Plastic and Reconstructive Surgery, Mie, Japan

ARTICLE INFO

Article history:
Received 18 April 2017
Received in revised form
12 May 2017
Accepted 15 May 2017
Available online 17 May 2017

Keywords:
Indocyanine green lymphography
Lymphatic-venous anastomosis
Micro-cystic
Lymphatic malformation
Periorbit

ABSTRACT

Lymphatic malformation (LM), also known as lymphangioma, is a rare condition that can occur anywhere in the body in adults and children. LM, especially micro-cystic type, consists of multiple clustering skin/subcutaneous cysts that may involve deeper functional tissue. Although LM is generally benign, it may cause serious location-dependent problems. Periorbital LM, for example, often causes facial disfigurement and visual impairment. Especially, micro-cystic type of lymphatic malformation (mLM), is a challenging condition to treat. Currently available therapies for mLM have limited effectiveness and carry a high risk of complications. Thus, it is necessary to develop a novel treatment for mLM that is less invasive and highly effective.

There has been a question of the exact location and direction of lymphatic flow into, within, and out of LM. There were some reports investigating the intra-cystic flow of LM; however, only a few focused on the afferent and efferent lymphatic flow of LM. With our detailed study using lymphography, we were able to determine the lymphatic flow pattern in periorbital LM, which was large enough to perform lymphatic venous anastomosis (LVA). LVA is a surgically-placed bypass between afferent collecting lymphatic vessels and veins, which in our study resulted in decreasing both the influx and the total volume of lymph in the LM. To our knowledge, this is the first study of Indocyanine green (ICG) lymphography on mLM, and LVA for its treatment. Here we present our case of periorbital mLM with LVA and discuss its less invasive surgical strategy.

1. Introduction

Lymphatic malformation (LM), also known as lymphangioma, is a rare condition that can occur anywhere in the body in adults and children. LM, especially micro-cystic type, consists of multiple clustering skin/subcutaneous cysts that may involve deeper functional tissue. Although LM is generally benign, it may cause serious location-dependent problems. Periorbital LM, for example, often causes facial disfigurement and visual impairment. Periorbital LM, including micro-cystic type of lymphatic malformation (mLM), is a challenging condition to treat. Currently available therapies for mLM have limited effectiveness and carry a high risk of complications. For instance, the efficacy of sclerotherapy is limited as some cysts may be too small for injection. Furthermore, surgical resection of periorbital mLM can cause structural and functional damage of levator and frontal muscles. Also, other typical complications following surgical resection, such as wound dehiscence or lymphorrhea, may prolong the duration of medical care, which is unfavorable especially on infantile cases. Thus, it is necessary to develop a novel treatment for mLM that is less invasive and highly effective.

There has been a question of the exact location and direction of lymphatic flow into, within, and out of LM. There were some reports investigating the intra-cystic flow of LM [1–5]; however, only a few focused on the afferent and efferent lymphatic flow of LM [6]. With our detailed study using lymphography, we were able to determine the lymphatic flow pattern in periorbital LM. Additionally, our analyses revealed that afferent flow of LM was large enough to perform lymphatic venous anastomosis (LVA). The LVA is a surgically-placed bypass between the afferent collecting

* Corresponding author. 1-2, Shintoshin, Saitama, 330-8777, Japan.
E-mail address: motoikato25@gmail.com (M. Kato).

http://dx.doi.org/10.1016/j.epsc.2017.05.008

lymphatic vessels and the veins, which resulted in decreasing both the influx and the total volume of lymph in the LM. To our knowledge, this is the first study of Indocyanine green (ICG) lymphography on mLM, and LVA for its treatment.

We utilized ICG lymphography, as it allowed us to analyze the dynamics of lymphatic flow in detail. With this pre-operative examination, we were able to identify the exact location of afferent flow of mLM (collecting lymph vessels), which was used for the following LVA procedure. Here we present our case of LVA on periorbital mLM and discuss its less invasive surgical strategy.

2. Case report

The patient was an 11 month-old Japanese female diagnosed with large lymphatic malformation spreading on her left upper eyelid, forehead, and scalp. No surgeries or treatment were applied previously. Ultrasound and MRI revealed that the lesion was mLM, which widely involved orbicularis and forehead muscle. The deepest part of LM was connected to intra orbital and extraconal lesion. Since her first evaluation at an outpatient clinic, regular ophthalmology visits were continued.

Under general anesthesia, we marked twelve spots on her scalp, forehead, and face after shaving scalp hair as pointed #1–12 (Fig. 1); Six points were along the midline (#1–6), two were on the contralateral side (#7–8), two were on the surface of the lesion (#9–10), and two more lateral to the tumor (#11–12) (Fig. 1).

The fluorescent dye, ICG (Diagnogreen® 0.25 mg/ml; Daiichi Pharmaceutical, Tokyo, Japan) was injected 0.02 ml at each marked locus subcutaneously in numbered order (Fig. 1). All procedures were carefully performed under original infrared camera system and infrared mode of microscope (OPMI Pentero 900, INFRARED 900; Carl Zeiss, Tokyo, Japan).

The points were marked as (Fig. 1)

#1. Midline/5 cm dorsal to vertex
#2. Midline/vertex
#3. Midline/anterior edge of anterior fontanel
#4. Midline/hair edge
#5. Midline/midpoint between #4 and #6
#6. Midline/midpoint between eyebrows
#7. 1 cm horizontally right from #5
#8. 3 cm horizontally right from midpoint between #4 and #5
#9. Surface of the mass/Midpoint of left eyebrow
#10. Surface of the mass/hair edge and center of the mass width
#11. Lateral edge of the mass/horizontal to #10
#12. Midpoint between lateral edge of left eyebrow and the base of ear lobe

Fig. 1. ICG was injected subcutaneously, at marked #1–12 as ordered number.

Upon completion of all injections and observations, we harvested 3 × 3 × 3 mm biopsy of the lesion from small skin incision for pathological diagnosis. The biopsy specimen was formalin-fixed, paraffin-embedded, stained with hematoxylin and eosin (HE stain), and immunostained (CD31, D2-40) by the department of pathology in our institute.

As described in the result section below, we determined the afferent collecting lymph vessels of mLM. Two months after the first general anesthesia, LVA was performed to place a bypass between afferent lymph vessels and superficial vein in order to reduce the lymphatic in-flow of mLM. LVA was performed as described in previous report [7] with modifications, as the bypass contained a valve. A 7 mm skin incision on the scalp was placed and carefully dissected correcting lymph vessel and superficial vein. The main trunk of peripheral vein was large compared to the lymph vessel, thus a small branch of the vein was selected for the anastomosis to avoid a size discrepancy (Each vessel was almost same size with an external diameter of 0.2 mm). The anastomosis was performed in an end-to-end manner, with 12-0 nylon suture under microscope magnification (OPMI Pentero 900, Carl Zeiss, Tokyo, Japan).

3. Result

3.1. ICG lymphography findings

Surprisingly, the lymphatic flow of the affected side was stronger and faster than the contralateral side especially at the sites #2–6 (Fig. 1). No flow from the midline to the contralateral side was observed and all flowed into affected side. There were two intact linear afferent lymphatic vessels flowing into the LM. Unfortunately, the efferent flow was not clearly observed, likely due to its flow into intra orbital LM lesion.

No allergic reaction was observed during and after the procedure. On all spots, patterns were visible and clearly observable within 10 min of injection.

Detailed findings of lymphatic flow pattern from each injected point follows (Figs. 2 and 3):

#1. Major fast linear pattern was seen bilaterally from the injection spot
#2. Major fast linear pattern stronger and faster on the ipsilateral (left) side, no major linear pattern flowed into the contralateral (right) side.
#3. Two major fast linear patterns ran to the lesion, faded at 5 mm medial to the tumor margin (especially frontal linear was stronger). Not seen to the right side.
#4. Major fast linear pattern was found to connect with the frontal branch of the linear from #3. Faded at the terminal portion of the frontal branch from the #3. Not seen to the right side.
#5. Major fast linear pattern ran to her nose vertically along the edge of the mass. Not seen to the right side.
#6. Fast linear pattern was seen to the upper and lower eyelid of the right side. Not seen to the left side.
#7 and #8. Linear pattern was seen to the left side only.