Lymphatic Malformation

Aim of the Study:

Lymphatic malformation (LM) is a rare slow flow of vascular malformation. Majority of LM present in the head and neck (H&N) region, causing potential risk of airway obstruction. Image-guided sclerotherapy is one of the most commonly adopted minimally invasive treatment of LMs. Herein we aim to review the outcome of doxycycline sclerotherapy on H&N LM.

Introduction:

Vascular anomalies encompass all vascular malformation and tumours; while vascular malformation includes components with arterial, capillary, venous, lymphatic, or a combination of the above types.

Lymphatic malformation is a type of slow flow vascular malformation that can be detected antenatally if lesions are large. It occurs one in every 2000 to 4000 live births. The aetiology is not well known, but it is proposed that lesions can be related to failure in communication between the immature lymphatic tissues with the lymphatic or venous system. Treatment for LM includes surgical resection, symptomatic control with medication, or sclerotherapy.

Despite having no evidence for a treatment of choice, studies have demonstrated that sclerotherapy has been a safe, effective, and inexpensive treatment. Efficacy and safety of using doxycycline for head and neck lymphatic malformation in children will be demonstrated with the results from our centre.

Methods:

Target population: (period of 2010-2019) patients under 18 years, with a diagnosis of macrocystic or mixed type LM in the head and neck region. All patients had pre-operative ultrasonography with Doppler and magnetic resonance imaging for diagnosis and assessment of the extent of the lesion.

Intervention: Sclerotherapy was performed in the endovascular operation theatre with bi-planar digital subtraction angiography guidance. Under general anesthesia, doxycycline sclerotherapy was performed under ultrasonic and fluoroscopic guidance in collaboration with radiologists dedicated to the study of vascular anomalies.

According to our periprojective protocol for sclerotherapy to head and neck LMs, post-operatively patients are not exubated and electively admitted to the paediatric ICU. Airway protection with mechanical ventilation via endotracheal tube in paediatric ICU were implemented if the LM was in close proximity to the airway. Short term steroid (0.1 mg/kg every 8 hours) with proton pump inhibitor cover was prescribed to decrease the post-sclerotherapy swelling. Patients were exubated and transferred to the general ward one clinical swelling has been controlled and deemed subsiding.

Outcome: assessed by the symptom and size change of the LM

Results:

10 patients with median age 6.5 (1.4-12) years were included in the series. Five patients (50%) had antenatal diagnosis, the rest presented with head and neck swelling up to 14 months of age. Lesions were predominantly found in the neck and submandibular region in 4 patients, parotid region in 2 patients. Other LM were found in the submental region, floor of mouth, and temporal region. The principal type of LM was mixed type LM in 4 patients; and macrocystic LM for 6 patients. The median pre-sclerotherapy volume was 23 ml (0.7-36 ml).

All 10 patients received close monitoring in PICU with short course of steroid. All of them had endotracheal tube kept in situ post-op with mechanical ventilation support shortly after the operation. Only one patient required repeated aspiration of sclerosant and cystic content due to reaccumulation of fluid. They were all able to extubate unexpectedly before being transferred to the general ward from PICU. No postoperative complications of airway obstruction and bleeding were reported.

Outcome: During follow in the outpatient clinic, 8 patients had clinically undetectable swelling or size reduction ranging from 46%-93%. Two patients with residual LM had staged sclerotherapy or required subsegent sirolimus treatment.

Conclusion:

Image-guided sclerotherapy by doxycycline in head and neck macrocystic and mixed LM is safe and effective. Post-sclerotherapy airway monitoring and protection can avoid dire consequence secondary to tissue swelling. Continuous follow up is necessary to monitor the long term outcome.

Reference: