USE OF SIROLIMUS AND SCLEROTHERAPY FOR LYMPHATIC MALFORMATIONS IN A DEVELOPING COUNTRY: A CASE SERIES

Resumen

Las malformaciones linfáticas y su manejo no han sido bien descritas en República Dominicana. Es por ello, que el objetivo de este artículo es la presentación de tres casos, con diferentes patrones y necesidades de tratamiento, de modo que sirva como referencia para trabajadores de la salud en países en vías de desarrollo.

Palabras clave: malformación linfática; sirolimus; bleomicina; doxiciclina; país en desarrollo.

Abstract

Lymphatic malformations and its management are not well described in the Dominican Republic. That is why this article's objective is to present 3 cases, with different patterns and treatment needs, so it will work as a reference for healthcare workers in developing countries.

Keywords: lymphatic malformation; sirolimus; bleomycin; doxycycline; developing country.

Introduction

A lymphatic malformation (LM) is an anomaly that results from an error in the embryonic development of the lymphatic system and can be classified as macrocystic, microcystic, and mixed. If acquired, lesions generally arise from obstruction of the lymphatic system due to trauma or infection. Most of the lesions are congenital and develop in infants and children under 2 years of age. The incidence of LM is estimated to be 1 per 6000-16,000 live births, with the majority of LMs (50-65%) evident at birth. Two-thirds of all the reported cases are located in the head and neck. Complications can include obstruction or distortion of vital structures, infection, bleeding, and pain.

Case presentations

Patient 1

A 10-year-old girl visited a new primary care physician in January 2015 with a previous diagnosis of facial hemangioma, treated with propranolol for 3 years and no major change observed. Since there was no improvement with the treatment given, her parents decided to look for a second opinion. The patient had already done a sagittal MRI (T1 sequence, with and without contrast), coronal (T2 sequence) and axial (FLAIR). A consultation, through telemedicine, with the Cincinnati Children’s Hospital Medical Center was done and the definitive diagnosis was a lymphatic malformation. The therapy with sirolimus started in April 2015 along with trimethoprim/sulfamethoxazole. The patient was clinically better in follow up visits but in December 2015, the mass was more inflamed, which was the reason why an injection with bleomycin was appropriate. In July 2016, due to bleomycin unavailability, an injection with doxycycline was another option. In September 2016, a second injection with bleomycin was required. In December 2016, a third injection was applied. She completed 2 years of sirolimus therapy. The clinical improvement was notable in subsequent visits [image 1].

Patient 2

A previously healthy 8-year-old boy visited a new pediatrician in February 2017, complaining about a mass in his gluteal region. His physical exam revealed what appeared to be a vascular malformation that was also infected. An axial, sagittal and coronal MRI of the gluteal area (T1, T2 and fat sat sequences with gadolinium) was performed and a complete blood count, liver function tests, cholesterol, blood urea nitrogen, serum creatinine and uric acid were ordered. After the MRI, the diagnosis of lymphatic malformation with inflammatory changes due to infection was confirmed. Laboratory results showed a microcytic hypochromic anemia and a slightly reduced uric acid. The treatment decided was sirolimus for 24 months, trimethoprim/sulfamethoxazole and a supplementation with iron, folate and vitamin B12.

In the follow up visits, there was clinical evidence of improvement of the patient’s lymphatic malformation until the treatment with sirolimus was over. Then, in March 2019, an increase in the lesion’s volume was seen. Sirolimus was continued for another 24 months. In subsequent visits, the patient showed improvement. Different MRIs are attached in this report to compare the lesion before and after the sirolimus [image 2].

Patient 3

An otherwise healthy 7-day-old term girl visited her primary care physician in January 2019, with a mass in the left side of her neck. An MRI of the neck was performed with and without intravenous contrast administration. The findings consisted in a multiloculated cystic lesion. The diagnosis made was macrocystic lymphatic malformation. An injection with bleomycin was applied in February and March the same year. A remarkable improvement was evident in posterior visits [image 3].
which is convenient for a developing country. 12. Lable, inexpensive and have well known effectiveness.


