Introduction

Lymphatic malformations (LM) are benign lesions that are the result of abnormal embryogenesis of the lymphatic system. Over fifty percent occur in the head and neck. Histologically; they are composed of dilated endothelium-lined cysts and are commonly grouped into three categories based on the size of the cysts: microcystic (cysts are individually less than 2 cm in volume), macrocystic (cysts are greater than 2 cm in volume), and mixed. This structural classification system has implications for treatment and prognosis. The treatment of lymphatic malformations (LM) has included a wide variety of techniques with no superior treatment identified. Treatment options include sclerosing agents such as picibanil (OK-432) and doxycycline. We wanted to explore the efficacy of doxycycline sclerotherapy in patients who have failed previous therapy with OK-432.

Case Series Cont.

Case 2:
8 year old F with a right parotid mixed LM (Fig 5). She was previously diagnosed with a macrocystic LM and treated with OK-432 sclerotherapy at 5 years of age with initial improvement; however, it quickly returned to previous size. Parents elected to observe, and the LM remained stable. At 8 years of age, additional treatment was pursued and pre-procedure ultrasound evaluation revealed a complex mixed LM with thick fibrous septations. This was treated with doxycycline sclerotherapy. (Fig 6). She tolerated the procedure well, however, at 4 month follow up had no clinically apparent change in mass. Ultrasound evaluation demonstrated resolution of dominant macrocysts with innumerable microcysts and residual fibrous tissue. They elected for continued observation with consideration for surgical excision with parotidectomy in the future.

Case 3:
5 year old M with a left neck and supraclavicular fossa lesion approximately 7 x 7 cm (Fig 7). He first presented to our clinic at 2 years of age. MRI performed during initial evaluation characterized the lesion as predominantly macrocystic. He was then treated with OK-432 sclerotherapy three times over a two year period. There was some temporary fluctuation in size with each treatment; however, the lesion continued to increase in size. When evaluated for sclerotherapy with image-guided doxycycline, a large, complex, cystic lesion was clinically apparent. Ultrasound performed demonstrated a mixed LM with complex morphology. He was treated with image guided sclerotherapy with doxycycline. He tolerated the procedure well and had no peri-operative complications. At 4-month follow up he demonstrated a decrease in macrocystic disease and an approximately fifty percent decrease in size (Fig 8). At 7 months follow up ultrasound demonstrated a mixed LM that was similar to pre-treatment size. Due to persistent disease, the patient underwent resection of the residual mass (Fig 9). He has done well since surgery with no evidence of recurrence (Fig 10).

Case Series

Methods

Institutional database of patients with head and neck lymphatic malformations from 2008 to 2012 was reviewed to identify patients who had persistent disease after treatment with OK-432 and subsequently underwent sclerotherapy with doxycycline. We identified three patients.

References