International Journal of Pediatric Otorhinolaryngology 87 (2016) 67–70

Contents lists available at ScienceDirect

International Journal of Pediatric Otorhinolaryngology

journal homepage: http://www.ijporlonline.com/

Case report

Pediatric lymphedema caused by diffuse cervical lymphadenopathy: A case report and review of the literature☆

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ARTICLE INFO

Article history:
Received 9 March 2016
Received in revised form
12 May 2016
Accepted 15 May 2016
Available online 20 May 2016

Keywords:
Head and neck lymphedema
Lymphadenopathy
Kawasaki's disease
Internal jugular vein
Pumpkin head

ABSTRACT

Pediatric head and neck lymphedema is rare and there have not been any reported cases in children. Here we discuss severe, diffuse head and neck lymphedema in a child caused by compression of the internal jugular veins by lymphadenopathy from Kawasaki's disease. With steroid and intravenous immunoglobulin treatment, the lymphadenopathy improved and facial edema slowly resolved. In review of the literature, complications of head and neck lymphedema including airway obstruction and blindness are discussed. This case highlights the importance of the pediatric otolaryngologist considering lymphedema as a cause for facial swelling and monitoring for complications of lymphedema.

1. Introduction

Lymphadenopathy is a common chief complaint for the pediatrician and pediatric otolaryngologist. The differential diagnosis of bilateral cervical lymphadenopathy includes infection—such as in association with viral or streptococcal pharyngitis—Kawasaki's disease, lymphoma, collagen vascular disease or a medication reaction. It is most commonly self-limited and rarely requires further intervention. However, further workup and treatment are indicated in complicated cases of lymphadenopathy. Common complications of lymphadenitis include abscess formation, skin drainage and less commonly, fistula formation. Rarely does it lead to compression of adjacent structures then leading to head and neck lymphedema.

Pediatric cases of head and neck lymphedema are rare and cases of lymphedema due to internal jugular compression in children have not been previously reported. In adults, head and neck lymphedema as a result of obstruction of venous outflow has been described with various etiologies. Direct compression of the internal jugular vein can be caused by any adjacent structure. Compression has been reported by the omohyoid [1,2], thyroid enlargement [3], tumors [4] or hematoma formation [5]. Here we present a case of severe, diffuse head and neck lymphedema caused by compression of the internal jugular veins in a child with bilateral cervical lymphadenopathy from Kawasaki's disease. This case report was approved by the Baylor College of Medicine institutional review board and written consent was obtained from the family to obtain and publish photographs of the child.

2. Case report

A 4-year-old previously healthy Hispanic male presented to the emergency room with a five-day history of rapidly progressive face and neck swelling. The facial swelling was associated with daily subjective fevers and dysphagia. His family brought him to the hospital when the swelling progressed to the point where he was unable to open his eyes. He did not have any preceding viral illness, respiratory distress or cough. He did not have any significant medical or surgical history. His birth history was unremarkable and his immunizations were up to date. He did not have any exposures to ill contacts, tuberculosis or cats. He did not have any family history of similar symptoms, lymphoma or known genetic syndromes.

On exam, he was febrile to 104.6 F and had diffuse facial edema with associated ptosis (Fig. 1). He was unable to open his eyes without assistance. On eye exam, the sclera were clear, visual acuity

☆ Presented at the 2015 SENTAC annual meeting in San Antonio, Texas.

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http://dx.doi.org/10.1016/j.ijporl.2016.05.028
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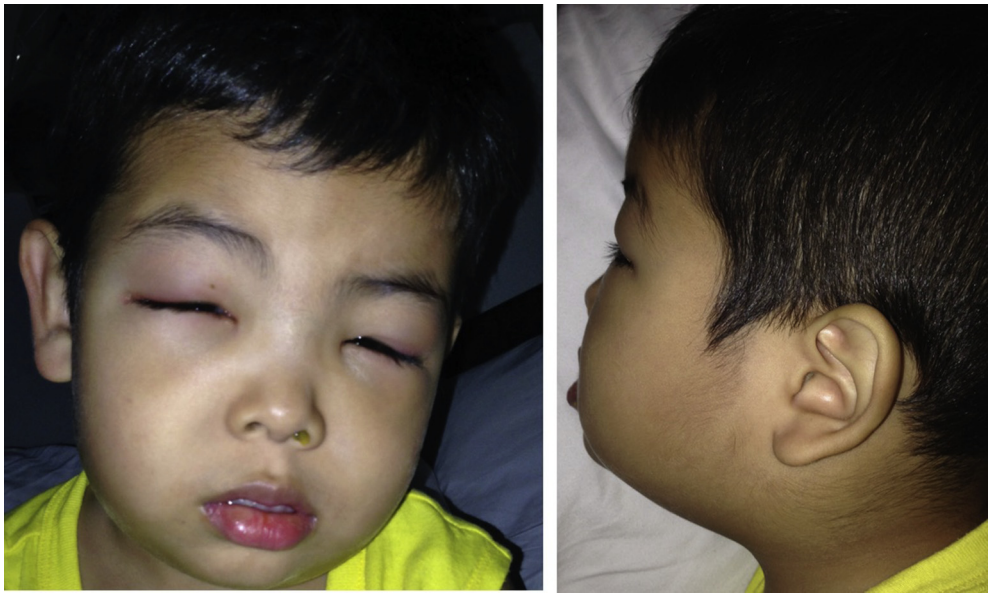

Fig. 1. The patient presented with severe, diffuse facial edema and ptosis.

was normal and intraocular pressure was normal. His oral cavity and oropharynx did not exhibit any edema and his voice was strong and clear. He had diffuse, bilateral, non-tender cervical lymphadenopathy with full neck range of motion. On flexible fiberoptic laryngoscopy, he had mild edema of the supraglottis, but his airway was widely patent and bilateral vocal folds were mobile. He had leukocytosis to 19.1 $\times$ 10^9 cells/L and initial rapid streptococcal antigen test, Monospot test and nasal wash adenovirus PCR were all negative. A computed tomography scan of the neck with intravenous contrast revealed diffuse bilateral lymphadenopathy causing direct compression of the internal jugular veins (Fig. 2). The carotid arteries were also medialized bilaterally by the massive lymphadenopathy.

The patient was started on empiric parenteral clindamycin and admitted for further work-up and treatment. Further infectious work up was negative for HIV, bartonella, EBV and tuberculosis. The patient was diagnosed with atypical Kawasaki's disease and started on high-dose intravenous immunoglobulin (IVIg) on hospital day four. Within two days of initiating treatment the patients' fevers and cervicofacial lymphedema resolved and the lymphadenopathy improved dramatically (Fig. 3). After treatment with high-dose IVIg and resolution of his symptoms, he was discharged home on hospital day six. On two-year follow up, he did not have any recurrence of lymphedema symptoms or long-term sequelae of Kawasaki's disease, such as coronary artery aneurysm.

3. Discussion

Severe, generalized head and neck edema, also called “pumpkin head lymphedema,” is caused by obstruction of lymphatic or venous outflow. Obstruction can occur at the level of the superior vena cava, as in superior vena cava syndrome, or the bilateral internal jugular veins, as seen in this case. There is extensive collateral blood flow in the head and neck, so there must be significant disruption of multiple drainage channels to cause generalized symptoms. Disruption of the lymphatic drainage itself can be a primary congenital problem, as seen in lymphatic malformations. This has only been previously reported in the extremities of

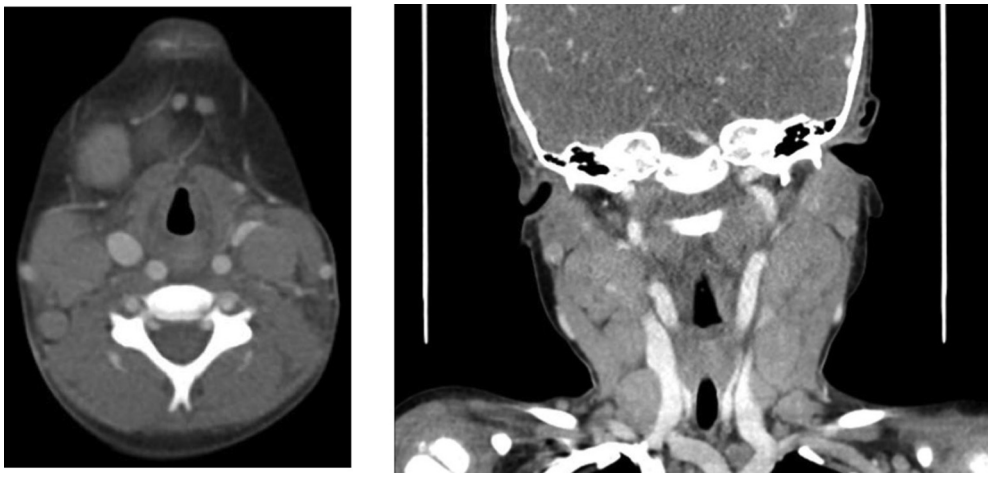

Fig. 2. Axial and coronal computed tomography images revealing bilateral cervical lymphadenopathy compressing the internal jugular veins.