DOI: http://dx.doi.org/10.3315/jdcr.2013.1145

Journal of Dermatological Case Reports

Life-threatening pharyngeal edema after sclerotherapy of oral venous malformations in a patient with blue rubber bleb nevus syndrome

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Key words:

Bean syndrome, blue rubber bleb nevus syndrome, oral mucous membranes, oral vascular malformation, sclerotherapy, venous malformation

Abstract

Background: Blue rubber bleb nevus syndrome (BRBNS) is a rare vascular disorder characterized by multiple venous malformations of the skin and internal organs. Oral lesions are very common and occur in over half of the patients with this condition. Sclerotherapy is currently the first-line treatment modality of symptomatic cases due to its high efficacy and low rate of complications.

Main observations: We report the case of a 68-year-old male with BRBNS who presented with dysphagia and difficulty with speech due to prominent oral venous malformations. After the use of sclerotherapy with ethanolamine oleate to control his symptoms, the patient exhibited severe edema of the tongue and posterior pharyngeal wall which caused constriction of his airway. The patient was intubated, and remained so for 72 hours until his edema resolved. In addition to his oral lesions, the patient also exhibited other features of BRBNS including cutaneous, soft-tissue, gastrointestinal, and neurological manifestations of disease.

Conclusion: Physicians should be aware of the potentially life-threatening complication of severe tongue and pharyngeal edema when using sclerotherapy for the treatment of oral vascular malformations. Additionally, dermatologists should be familiar with the many systemic manifestations which can be present in patients with BRBNS, as illustrated in this case. (*J Dermatol Case Rep*. 2013; 7(3): 74-76)

Introduction

Blue rubber bleb nevus syndrome (BRBNS) is a rare disease characterized by multiple venous malformations in the skin and gastrointestinal tract.[1] These malformations occur frequently in the oral cavity and have been noted in 59-64% of cases.[1] We present the case of a patient with prominent oral lesions who demonstrated severe edema of the tongue and posterior pharyngeal wall following treatment with sclerotherapy which required intubation. Significantly, sclerotherapy currently represents the first-line treatment option for symptomatic oral lesions from BRBNS.[2,3] It has been described as "safe and effective" with minimal risk of serious complications.[2,3] To our knowledge, this patient's complication from the treatment of lingual venous malformations has not been reported in the literature. In addition to his oral findings, our patient also demonstrated many other systemic symptoms that can be seen in BRBNS including cutaneous, soft-tissue, gastrointestinal, and neurological manifestations of disease.

Case Report

A 68-year-old male presented with dysphagia and difficulty with speech due to an enlarged tongue. His tongue had been oversized since childhood and had been getting progressively more swollen during the past two years. On physical exam, his tongue was distended (Fig. 1) with dramatically engorged vascular structures in the inferior and lateral portions (Fig. 2). The patient's lower lip had a purple hue with perioral, small, discrete purple papules (Fig. 2). His past

medical history was notable for coronary artery disease and BRBNS diagnosed in 1994, when he presented with headaches and focal neurologic defects due to an aneurysm of the left posterior communicating (PCOM) artery, attributed to a vascular malformation. He underwent a left temporal craniotomy and had a PCOM clip inserted. At that time, the radiologist also noted other venous malformations throughout the white matter and cerebellum. The patient was also found to have chronic anemia from bleeding venous malformations of the GI tract visualized by both endoscopy and colonoscopy.

The patient underwent an elective embolization of the lingual artery using sclerotherapy in an effort to decrease his macroglossia. Under general anesthesia, two large superficial venous malformations on the left anterior and right posterior side of the tongue were cannulated with a 21 g needle. Their contents were aspirated to decrease their size and a small amount of contrast was injected under fluoroscopy to confirm the appropriate location. Three cubic centimeters (cc) of 5% ethanolamine oleate mixed with 1 cc of contrast was injected under negative roadmapping into the vascular malformation on the left anterior side of the tongue. This was repeated in the right posterior location with 2 cc of ethanolamine oleate. Pressure was held for 5 minutes until hemostasis was achieved. The procedure was uncomplicated and the patient was extubated shortly after surgery.

After approximately two hours, the patient began to exhibit severe edema of the tongue and posterior pharyngeal wall, and demonstrated breathing difficulty. To preserve the airway, he was re-intubated and remained so for 72 hours until the edema resolved. At this point, a fiberoptic examination revealed multiple venous malformations of the lateral and posterior pharyngeal wall, the supraglottic region, and the esophagus. They were judged to be non-obstructive and no treatment was deemed necessary.

Figure 1

The patient's tongue was found to grossly distended and slightly malformed. Large veins can be visualized on the superior aspect of the tongue.

Discussion

BRBNS most commonly presents in the skin and gastrointestinal tract.[1] Cutaneous lesions are usually asymptomatic and present as small, cyanotic, blue papules with a nipple-like center.[4] Involvement of the GI tract is very common in patients with BRBNS, and these patients often present with iron-deficiency anemia from bleeding lesions of the small intestine.[1] Involvement of other internal organs is uncommon, but has been described in the kidney, heart, lungs, penis, spleen, thyroid, adrenal gland, bladder, tongue, and CNS.[5] Oral lesions are very common and occur in 59-64% of cases, mostly in the buccal mucosa, ventrolateral tongue, and retromolar trigone.[2]

There are a wide variety of treatment strategies for symptomatic oral lesions. Smaller malformations can be treated with sclerotherapy, surgical excision, lasers, radiation therapy, electrodessication and curettage, and cryotherapy.[1-3] Large lesions have been treated with systemic corticosteroids, sclerotherapy, and interferon alpha.[3] There are recent reports that low-dose sirolimus can be used as an anti-angiogenic agent to drastically reduce the size of the venous malformations in BRBNS.[6]

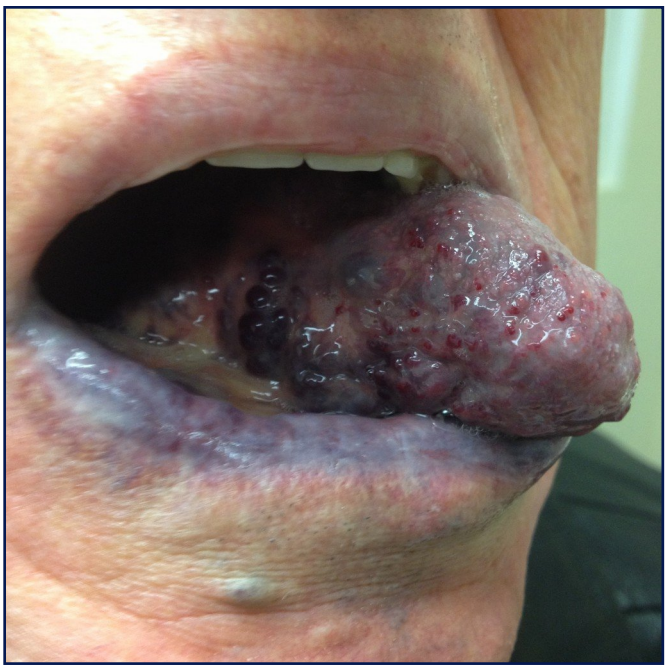

Figure 2

The patient's macroglossia and lingual venous malformation can be visualized. Additionally, a purple hue of the lower lip and perioral small purple papules can be seen.

Surgical excision of vascular malformations often gives good results but is relatively invasive and presents a risk of hemorrhage.[3] Embolization using sclerosing agents is highly efficacious and poses no risk of hemorrhage, and for this reason is considered by most physicians to be the first-line therapy.[2,3] Sclerosing agents commonly used include 5% ethanolamine oleate, 5% sodium morrhuate, 1% polidocanol, sodium tetradecil sufate, and hypertonic saline.[2,3] In a study performed on 30 patients with oral vascular lesions, the authors found that ethanolamine oleate was 100% effective in relieving symptoms and there were no serious adverse reactions.[3] The most common side effects of ethanolamine oleate are thrombophlebitis, localized inflammation, pain, and skin pigmentation.[7] Serious complications are very rare, but include anaphylaxis, renal toxicity, deep and superficial vein thrombosis, pulmonary embolism, transient bradycardia, stroke, tissue necrosis/ulceration, and nerve damage.[5,7-9]

We believe that the complication of pharyngeal edema seen in our patient was due to a local inflammatory response and vascular congestion, and not an allergic reaction. In our review of the literature, allergic responses to ethanolamine oleate are very rare and when they occur, include systemic symptoms such as fever, diffuse erythema, pruritus, wheezing, peripheral hypoperfusion, and hypotension.[8] On the other hand, local inflammatory responses and venous congestion are common and occur, at least to a small extent, in many patients. The difference is that in our patient, this inflammation occurred in a location that caused airway occlusion. For this reason, we believe the laryngeal edema seen in our patient can correctly be classified as a local inflammatory response, and not an anaphylactic reaction. In our review of the literature, this occurrence has not been previously described.

Conclusion

BRBNS is a rare disorder characterized by venous malformations found predominantly in the skin and GI tract. Oral lesions are very common and occur in over half of these patients. For the treatment of symptomatic oral lesions, sclerotherapy is currently the first-line treatment modality due to its efficacy and safety. We wish to present this case to illustrate the potential risk of severe pharyngeal edema leading to airway obstruction after the use of sclerotherapy to treat oral venous malformations in patients with BRBNS. We believe this is especially pertinent given the high prevalence of oral lesions in this population. This case also highlights many of the systemic findings found in patients with BRBNS of which dermatologists should be aware.

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