GMJ.2015;4(2):126-28
www.gmj.ir

CASE REPORT

Received: 2014-07-06
Revised: 2014-07-10
Accepted: 2014-08-12

Idiopathic Subglottic Stenosis in a Patient with Generalized Port-Wine Stain: A Case Report and Review of Literature

Elham Behrangi[1], Laya Yousefian[2], Aliakbar Moodi[3], Zahra Azizian[1] ✉

[1] Dermatology Department, Rasoul Akram Hospital, Iran University of Medical Sciences, Tehran, Iran.
[2] Otolaryngology Department, Rasoul Akram Hospital, Iran University of Medical Sciences, Tehran, Iran.
[3] Ziaiyan Hospital, Tehran University of Medical Sciences, Tehran, Iran

Abstract

Background: Subglottic stenosis is defined as inflammatory narrowing of the airway. It can be related to trauma, intubation, autoimmune disorders, vascular malformations, infection and congenital abnormalities. Pot-Wine Stain (PWS) is a small dermis venous malformation which is present at birth. PWS is found on face and neck, but can occur in throat and can cause subglottic stenosis. **Case Report:** Authors describe an 18-year-old girl with progressive dyspnea misdiagnosed with asthma having no response to medication. She has no histories of trauma, intubation, gastro-esophageal reflux disease (GERD) or allergy symptoms. **Conclusion:** Evaluations revealed subglottic stenosis in association with generalized PWS. To the best of our knowledge, this finding has not been reported in English medical literature. **[GMJ.2015;4(2):126-28]**

Keywords: Port-Wine Stain (PWS); Subglottic Stenosis; Vascular Malformation

Introduction

Subglottic stenosis is an inflammatory narrowing of the airway, usually at the level of cricoid cartilage. It can be related to trauma during airway intubation, surgery, radiation, autoimmune disorders such as Wegener's granulomatosis, vascular malformations such as hemangioma, infections such as tuberculosis or histoplasmosis and congenital abnormalities [1]. Roughly, 5% of cases will remain idiopathic after all known causes have been ruled out [2].

Pot-wine stain (PWS) or Nevus flammeus is defined as a macular telangiectatic patch which is present at birth and remains throughout life [3]. It is an uncommon problem encountered in 0.3% of newborns. At birth, PWSs are well-defined pink, purple or red macules and patches. These vascular malformations tend to grow with child, darken, and become more irregularly surfaced in adult life [4]. These lesions are often isolated findings, but also may be associated with other congenital abnormalities, such as Sturge-Weber or Klippel–Trénaunay syndrome, in which patients also have seizures, glaucoma, abnormal cerebral vasculature and mental retardation [5]. They are routinely found unilaterally on face and neck, but can occur anywhere on the body [6]. Lesions close to or inside mouth, are correlated with concomitant lesions in throat and can cause subglottic stenosis.

We describe the first case of idiopathic subglottic stenosis in association with generalized PWS that has not been reported in English language literature. Informed consent was taken prior to publication from the patient.

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Fax: +98 731 2227091
PO Box 7461686688
Email:info@gmj.ir

✉ **Correspondence to:**
Zahra Azizian, Resident of Dermatology, Dermatology Department, Iran Medical Science, Tehran, Iran
Tel (Fax): (+98) 21-66515000
Email Address: azizian_z@yahoo.com

Case Presentation

An 18-year-old girl was admitted to our clinic (Rasoul-e-Akram) with chief complication of progressive shortness of breath and dyspnea for 3 months. Symptoms were more severe during two past weeks. She was previously misdiagnosed with asthma and received Fluticasone for 4 years. Nevertheless, she had no response to therapy. There were no histories of trauma, intubations, gastro-esophageal reflux disease (GERD), or allergy symptoms. On examination, she had multiple, well-defined, 2-mm to 3.5-cm, erythematous to violaceous, blanching macules and patches located on both upper and lower limbs (Figure 1, 2). She had no systemic complaints and was otherwise healthy. Her parents reported that the lesions were present at birth. According to historical and clinical information, lesions were considered as generalized PWS. Skin biopsy was performed and a diagnosis of congenital generalized PWS was made. Bronchoscopy demonstrated a simple weblike subglottic stenosis 1.5 cm below vocal cords. Regarding risks of laser ablation, she underwent rigid bronchoscopy with mechanical dilation. She has had no recurrence after 6 months of follow-up.

Discussion

Idiopathic subglottic stenosis is a rare condition. The exact incidence and etiology are obscure. Previous studies suggested GERD as a causative factor, with a single case report documenting successful therapy of a subglottic stenosis after GERD therapy [7]. In our report, the patient does not have any symptoms of GERD.

Post intubation trauma is the most usual cause of tracheal stenosis and is thought to be related to ischemic damage and the mechanical effects induced by prolonged or traumatic intubation [8]. The clinical presentation of post-intubation tracheal stenosis is stridor and shortness of breath after a recent prolonged intubation. It occurs most commonly in the first 2 months after extubation after mechanical ventilation of 5 to 7 days [9]. No case of permanent tracheal stenosis has ever been described in patients with intubation for less than 18 hours [10].

Rosen and Smoller later theorized that both acquired and congenital PWSs were the result of malformed sympathetic innervation. In the case of congenital PWS, there was a maturational defect in local sympathetic nervous system, whereas loss of sympathetic innervation, possibly through trauma, could lead to acquired PWSs [11].

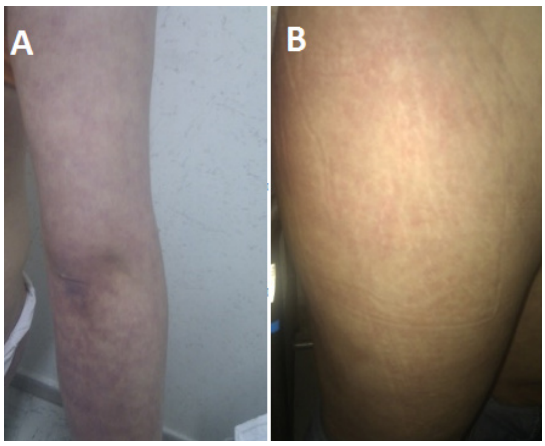

Figure 1. Examination of lesions shows dark red color patches and the muscles on her body. A; Dorsal surface of arm and forearm. B; Ventral surface of thighs.

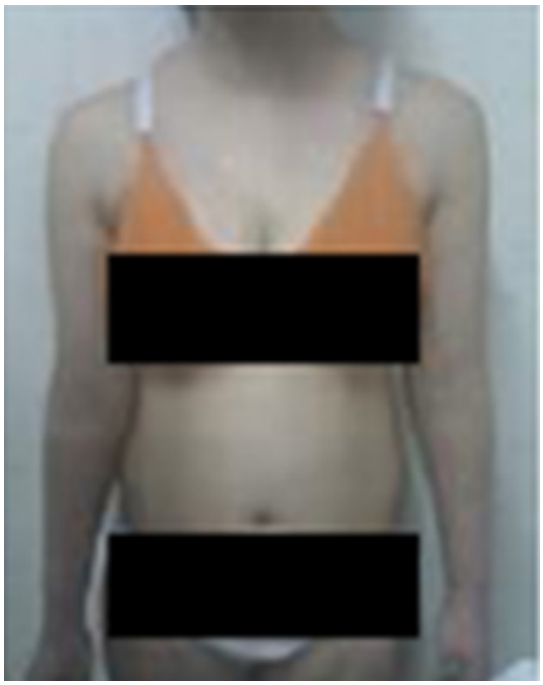

Figure 2. Typical PWS lesions extended to both upper and lower limbs.

It has been demonstrated that vessels within a PWS do not respond normally to vasoactive stimuli, further supporting the role of altered autonomic innervation [12].

PWS is a small dermis venous malformation which is present at birth. They may be localized or generalized affecting a whole limb. These are best considered as low-flow vascular malformations that may occur on any part of the body but commonly affect face. Initially, these lesions are pale pink patches, but with time, they may mature into a violaceous color, remain static or even lighten and may become nodular [13].

Idiopathic tracheal stenosis is a condition that can be challenging to treat. Its management includes an endoscopic approach with mechanical dilation using rigid bronchoscopy or balloon dilation with flexible bronchoscopy, but the rate of recurrence has been reported as high as 87% in 5 years. Crico-tracheal resection is an alternative approach with a recurrence rate that varies between 10% and 40% in 8 years [14].

Our case had a sign of hemangioma on her body (generalized PWS) but no vascular lesion on her larynx, bringing up the possibility that vascular lesions might have been disappeared during time and subglottic stenosis is a sequel.

Conclusion

Accordingly, subglottic stenosis is a rare but a curable cause of shortness of breath that can be associated with different clinical entities. Hence, physicians should be aware that recognition and understanding of vascular abnormalities associated with this condition might include PWS in their differential diagnoses.

Conflicts of Interest

None declared.

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