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Laryngeal Obstruction due to Blue Rubber Bleb Nevus Syndrome

Vittorio D'Aguanno, MD[1], Massimo Ralli, MD, PhD[1], Antonio Greco, MD[1], and Marco de Vincentiis, MD[2]

Blue rubber bleb nevus syndrome (BRBNS) is a rare vascular disorder characterized by multifocal venous malformations, occurring in any tissue, although skin, soft tissues, and gastrointestinal (GI) tract are the most frequently involved areas. Involvement of head and neck region is common. However, occurrence in upper airways may be challenging because of the risk of airway obstruction and bleeding.

A 66-year-old woman was admitted to the emergency department of our hospital for a severe GI hemorrhage. In her medical history, the patient reported previous episodes of GI hemorrhage and intermittent dysphagia to solid foods, dry cough, foreign-body sensation in the throat, and dyspnea on exertion for 4 months. Chemoembolization of the GI bleeding lesions was planned. Before undergoing the procedure, the patient presented with a worsening of her dyspnea. Flexible laryngoscopy of upper airway (Figure 1) was performed that revealed the presence of a blue-reddish, large, vascularized lesion involving the left supraglottic region with severe compromise of the respiratory airway space. A smaller lesion was observed in the posterior pharyngeal wall. The head and neck examination revealed bilateral parotid gland enlargement. The patient was informed of her condition and tracheotomy was performed under general anesthesia, before proceeding with chemoembolization of the GI bleeding lesion. After successful treatment of intestinal bleeding, a magnetic resonance (MR) scan with contrast (Figure 2) confirmed the supraglottic soft tissue highly vascular mass, measuring 46 × 41 mm. STIR (Short-TI Inversion Recovery) sequences showed the extent of the venous malformation, which appears as a hyperintense, multilobulated, septated mass involving the larynx, with severe reduction of the airway space. No clear signs of cartilage erosion were reported. Similar lesions were observed in the tongue, right parotid gland, posterior pharyngeal wall, and subcutaneous tissue of the neck. Surgical excision was recommended, although the patient refused further treatments. The patient had no further bleeding; however, tracheostomy has not been removed.

Blue rubber bleb nevus syndrome is a rare condition also known as ''Bean syndrome'' since William Bean fully characterized this disorder in 1958. Because of its appearance was termed ''Blue rubber bleb nevus syndrome.''[1] Skin manifestations appear as small, blue, compressible lesions of 1 to 2 cm.[2] Occurrence in oral cavity is reported in 59% to 64% of cases.[3]

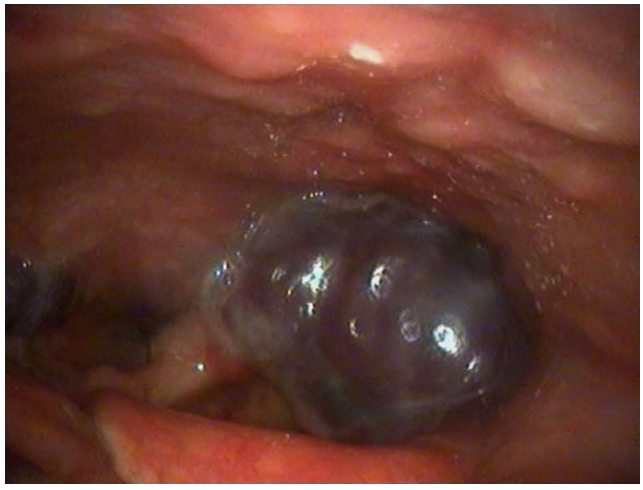

Figure 1. Flexible laryngoscopy showing a large, blue-reddish vascularized lesion, involving the left supraglottic region with a severe reduction of the airway space.

These lesions may develop at birth or in early childhood, growing with age,[3] and typically they occur on the skin of

[1] Department of Sense Organs, Sapienza University of Rome, Roma, Italy
[2] Department of Oral and Maxillofacial Sciences, Sapienza University of Rome, Roma, Italy

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Corresponding Author:
Vittorio D'Aguanno, MD, Department of Sense Organs, Sapienza University of Rome, Viale del Policlinico 155, Rome 00186, Italy.
Email: vittorio.daguanno@uniroma1.it

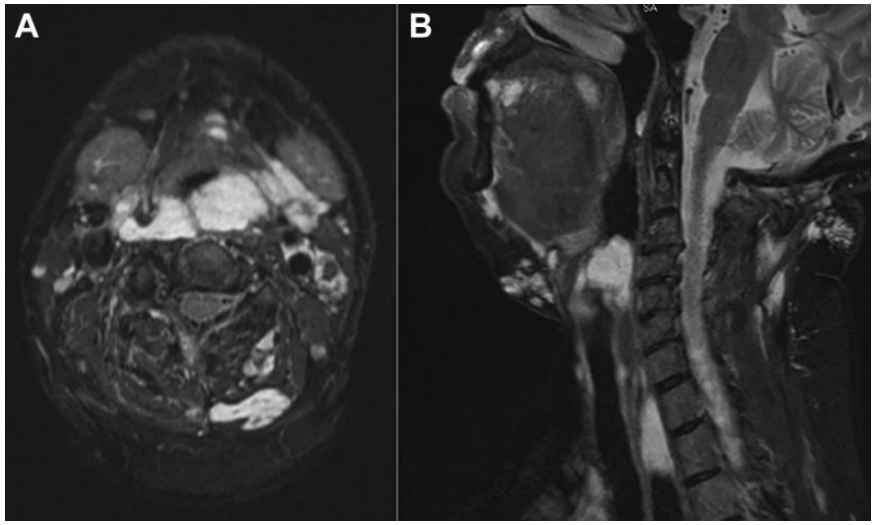

Figure 2. Axial (A) and sagittal (B) magnetic resonance (MR) scan, STIR images, showing a supraglottic soft tissue high-vascularized mass, measuring 46 × 41 mm. No evidence of bony or cartilage erosion were observed.

upper limbs, trunk, and perineum, although they can arise on skin throughout the body. Blue rubber bleb nevus syndrome has occurred in liver, spleen, heart, lung, pleura, kidney, thyroid, parotid, skeletal muscle, bladder, and the central nervous system.[3]

Blue rubber bleb nevus syndrome may occur as a sporadic condition or as inherited disorder with an autosomal dominant transmission pattern,[4] often associated with Maffucci syndrome. Histologically, these lesions appear as dilated vascular spaces lined with cuboidal epithelium with an increased number of sweat glands and presence of smooth muscle fibers.[5]

Morbidity of BRBNS is associated particularly with GI involvement, with chronic silent bleeding at an early age, evolving in some cases in an acute anemia.[2] Treatment is based on control of GI bleeding, with symptomatic treatment using transfusions or iron supplementation, and with regular blood tests. Surgery is not considered definitive treatment because of the high rate of recurrence and the large number of bowel lesions. However, in selected cases, surgical excision of bowel lesions may be considered as a preventive measure against either massive hemorrhage or obstruction.[3] Several antiangiogenic agents as corticosteroids, interferon, and octreotide have been proposed for BRBNS patients.[2]

Local treatments including laser photocoagulation,[2] cryosurgery, and sclerosing agents have been used widely, mainly for cutaneous lesions considered unacceptable cosmetically or functionally. Possible involvement of the upper airways should be considered in all patients with BRBNS. Symptomatic laryngeal lesions should be treated to prevent airway obstruction or troublesome bleeding. A case of BRBNS requiring surgery has been described previously in a 16-year-old girl with a supraglottic mass.[6] Thus evaluation of upper airways should be performed in BRBNS patients undergoing surgery anywhere in the body to evaluate the risk of airway obstruction and injury during intubation, causing a potentially catastrophic hemorrhage,[7] even in patients without laryngeal symptoms. If laryngeal lesions are present, they should be treated before elective surgery is performed elsewhere, and blind nasal intubation should be avoided. Intubation should be performed only with good visualization of the larynx and related structures.

Authors' Note

The data sets used and/or analyzed during the current study are available from the corresponding author on reasonable request. All the clinicopathologic investigations detailed in the manuscript have been conducted in accordance with the Declaration of Helsinki and its later amendments or comparable ethical standards. Written informed consent for publication of data and images was obtained from the patient.

Declaration of Conflicting Interests

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ORCID iD

Vittorio D'Aguanno https://orcid.org/0000-0001-5682-1009
Massimo Ralli https://orcid.org/0000-0001-8776-0421

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