

Blue Rubber Bleb Nevus Syndrome

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BLUE Rubber Bleb Nevus Syndrome is a rare anomaly of developmental vascular biology that can occur in any part of the body, most commonly the skin and gastrointestinal tract. The accompanying images show the typical appearance of these firm but compressible lesions (*short arrows*) in the pyriform

sinuses, the anterior laryngeal wall, and in relation to the aryepiglottic fold (left panel, *long arrow*). These locations would be at risk for injury during direct laryngoscopy and intubation, or even with placement of a laryngeal mask, which could result in potentially catastrophic hemorrhage.

Sometimes called “Bean Syndrome” following its original description in 1958, this entity can involve dozens of these lesions, which are felt to be congenital venous malformations rather than true vascular neoplasms.¹ Although cutaneous lesions are often present at birth or develop in early childhood, the diagnosis has been made in an octogenarian. Of approximately 300 cases in the literature, approximately 10% showed definite airway involvement; central nervous system involvement was almost twice as common.² Clinical manifestations are secondary to chronic occult blood loss, or rarely, intussusception from a bleb acting as a lead point. Although surgical extirpation of symptomatic lesions has been the mainstay of therapy, recent reports describe regression after sirolimus treatment.³ Patients presenting for endoscopy or surgical intervention should be assessed preoperatively for anemia and assumed to have lesions in the upper aerodigestive tract. Blind nasal intubation should be avoided, and because involvement of the lower airways has been observed, fiberoptic intubation may be prudent.

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Competing Interests

The author declares no competing interests.

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