Tracheostomy Cuff Induced Tracheomegaly in a Female Carrier of Duchenne’s Muscular Dystrophy

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Intro Abnormal tracheal enlargement or tracheomegaly has been reported in congenital tracheobronchomegaly, Ehlers-Danlos syndrome and diffuse acquired tracheomalacia from a multitude of inflammatory conditions (2). The average tracheal diameter for an adult is 10-20 mm. The upper limits in men are 25-27 mm and 21-23 mm in women (1). We present a case of tracheomegaly in a carrier of Duchenne’s muscular dystrophy (DMD) secondary to a tracheostomy cuff. Case Report The patient is a 58-year-old female carrier of DMD and chronic respiratory failure that presented with shortness of breath. Due to her manifesting carrier status, she required a chronic tracheostomy with ventilator support. She had become progressively more difficult to ventilate, with high leak, requiring increased ventilator pressures. On CT imaging, she was found to have localized tracheomegaly at the site of her cuff. The area of tracheomegaly was nearly spherical, and was measured 42x42x46mm. The trachea was visually inspected under flexible bronchoscopy. Given the location of dilation, a tracheostomy tube with an extended length and distal cuff was found to be the most appropriate for adequate ventilation. Discussion DMD is an x-linked recessive disease typically affecting only males. However, in rare instances (8%) it can also affect female carriers with variable manifestation. (3) In DMD there is a disruption of the dystrophin protein which acts to preserve muscle cells. In some conditions, the primary cause of tracheal dilatation is thought to be a congenital deficiency in the tracheal and bronchial cartilaginous and membranous tissues. In prolonged endotracheal intubation, acquired tracheomalacia may result from pressure necrosis, impairment of blood supply, infection, and cyclic friction on dry tracheal mucosa. This usually results in focal stenosis of the trachea, but may occasionally cause focal or even diffuse tracheomegaly (2). While there have been previous cases reports associating tracheostomy cuffs with tracheomegaly it continues to be a rare complication. In patient’s that already have increased muscle breakdown, they are unable to compensate for the pressure of the cuff by increasing muscle production. Signs of this development are increasing leaks, or ventilator pressure with a tracheostomy balloon that is properly positioned and inflated. Conclusion Tracheostomies are not uncommon, but tracheomegaly is a rare complication. While we should remain cautious of this complication, we must be even more vigilant with patients with increased muscle breakdown or impaired muscle production.

Resources

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