We present the case of a 19-years-old female patient with history of mucopolysaccharidosis type VI (Maroteaux-Lamy syndrome) diagnosed in 2001 and that presents the typical multisystemic features of this pathology. She is under enzyme substitution treatment (galsulfase) since 2006.

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The patient underwent a surgical tracheostomy following an elective procedure that induced major airway edema and made extubation impossible. Ventilatory weaning progressed favorably after that and she was discharged. The patient never complained of swallowing difficulties.

One month later she was brought into the emergency department with a tracheostomy bleeding. At this time a fibroscopic evaluation through the tracheostomy cannula revealed a bulging of the posterior tracheal wall, just below
the inferior limit of the cannula, that obliterated about 75% of the tracheal lumen (Fig. 1).

A CT scan was performed and showed an external compression of the posterior tracheal wall, immediately bellow the inferior limit of the cannula, caused by an aberrant right subclavian artery originating from the aortic arch (Arteria Lusoria – Figs. 2 and 3).

The estimated incidence of this aortic arch anomaly is 0.5–2%. Only in 15% of these cases the aberrant artery passes between the esophagus and trachea (as it is in this patient).