Significant narrowing and deviation of the trachea in an Osteogenesis Imperfecta Type III patient: previously unreported

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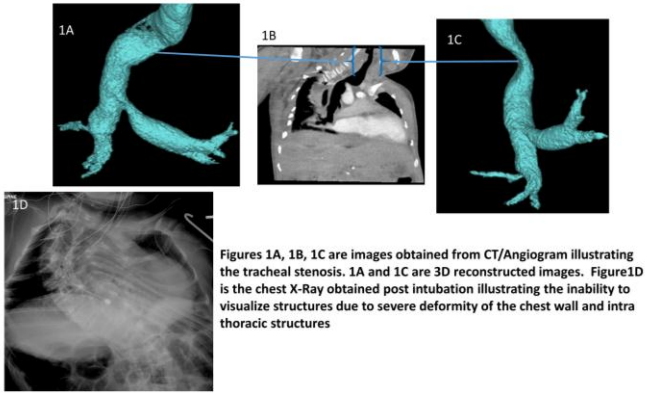
INTRODUCTION

- Osteogenesis imperfecta (OI) is an autosomal dominant disorder affecting type I collagen. OI affects 6-7 individuals per 100,000 worldwide.[1]
- OI type III is characterized by persistent fractures, growth delay, and kyphoscoliosis leading to cardiac and respiratory comorbidities.
- Tracheal abnormalities have never been reported in patients with OI.

CASE REPORT

- A nineteen-year-old male with OI type III and severe thoracic and thoracolumbar scoliosis presented to our institution for a T2-L4 posterior spinal fusion.
- Due to the severity of the spinal curvature and the small thoracic cavity, the trachea was not well visualized in any preoperative X-rays(Figure 1A).
- Shortly after induction, hand ventilation became difficult and an oral airway was placed - chest rise was observed but no end-tidal carbon dioxide tracing was seen.
- Direct laryngoscopy was performed with a Macintosh #3 blade and a 6.5mm cuffed endotracheal tube (ETT) was passed through the vocal cords; however, it could not be advanced further into the trachea.
- During this time the pulse-oximeter reached a nadir of 82% and remained in the high 80s for approximately 4 minutes
- Fiberoptic examination revealed a severely deviated, torturous, and narrow trachea past the thoracic inlet.
- The ETT was taped at 17cm at the gums and a portable CXR confirmed position.
- At the end of the procedure, the trachea was extubated once the patient was awake and following commands.
- Postoperatively, CT angiogram of the chest showed significant tracheal narrowing at the level of the thoracic inlet (Figure 1B,C, D).

Figure 1



Figures 1A, 1B, 1C are images obtained from CT/Angiogram illustrating the tracheal stenosis. 1A and 1C are 3D reconstructed images. Figure1D is the chest X-Ray obtained post intubation illustrating the inability to visualize structures due to severe deformity of the chest wall and intra thoracic structures

DISCUSSION

- Two aspects of this case are unique and have not been reported regarding OI.
- First, loss of end-tidal carbon dioxide after placement of an oral airway despite normal chestrise seen during the same time.
- This may be due to narrow or stenotic trachea which may have a component of malacia which causes it to obstruct during exhalation.[2,3]
- It is also possible that the oral airway caused the epiglottis to buckle backwards and obstruct the laryngeal inlet, which has been described in children with mucopolysaccharidoses (MPS).[4]
- Second, we were surprised by the difficulty encountered due to the tracheal abnormality in passing the ETT beyond the cords.
- Tracheal abnormalities including deviation, buckling, and narrowing have been best described in children with MPS, specifically Morquio Syndrome(MS).[5,6,7]
- The prevailing theory is that different growth rates between the trachea, neck, and chest cavity plus pectus carinatum and a narrow thoracic inlet lead to severe airway distortion.[19]
- Combined with a kyphotic curve of the spine compromising the space for major airways and vessels, this may have caused the anatomy seen in our patient.
- The severity and uniqueness of the tracheal abnormalities in this patient may become more prevalent as patients continue to live longer due to newer medical therapies such as bisphosphonate treatments.

CONCLUSION

- Any suspicious finding on CXR such as deviation, narrowing, or new onset of noisy or difficult breathing should be further evaluated for tracheal abnormality.
- Any patient with OI who has a significant kyphotic curve, or new respiratory symptoms should undergo imaging such as CT angiogram using dose reduction techniques.
- Anesthesiologists should be prepared to manage the airway with difficult airway equipment including appropriately sized fiberoptic scopes.

REFERENCES

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