Tracheal Compression in Marfan Syndrome

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The causes of benign upper airway obstruction are numerous. They include compression by bronchogenic cysts, thyroid goiter, and vascular structures, inflammatory diseases such as relapsing polychondritis and Wegener granulomatosis, postintubation tracheal stenosis, tracheomalacia, and megaesophagus secondary to advanced achalasia. Compression of the trachea by vascular structures has been best described in the pediatric literature (1).

A 19-year-old woman was referred for a preoperative pulmonary evaluation before explorative laparotomy for an abdominal cystic mass.

At the age of 16 years she was diagnosed as kyphoscoliosis and a brace was prescribed for thirty months. The patient denied any symptoms except mild wheezing on exertion and low abdominal pain.

Physical examination revealed arachnodactylia, a moderate right thoracic kyphoscoliosis and mild pectum excavatum. Lung auscultation revealed mild wheezing. Chest Rx indicated right kyphoscoliosis and probable stenosis of the trachea. Pulmonary function tests done on admission demonstrated an FEV₁ of 1.30 L (34.9% of predicted), an FVC of 1.32 L (31.1% of predicted) and an FEV₁/FVC ratio of 98.26, with a peak expiratory flow of 3.63 L/s. The flattening of the initial portion of the expiratory loop suggested an obstruction of the large airways (Fig. 1). Echocardiography revealed aneurism of sinus of Valsava with a diameter of aorta root of 4.4 cm, mild tricuspid and mitral valve regurgitation and dilatation of the proximal main pulmonary artery of 3.7 cm. Oculistic exam showed myopia. The patient fulfilled the Ghent criteria for a sporadic form of Marfan syndrome (2). Dynamic chest CT demonstrated a compression of the trachea at the level of the third thoracic vertebra by an innominate artery, as the trachea was moved away from the thoracic spine compression (Figs. 2, 3). At that point the diameter of the trachea was 4 mm, for a length of 2.6 cm, immutable with respiration.

This is the first case of concomitant tracheal compression caused by a normal innominate artery anteriorly, and by the thoracic spine posteriorly, in Marfan syndrome in an adult. In fact, in this case of right scoliosis, the leftward displacement of the trachea from the vertebra causes the innominate artery to run over the trachea and compress it. It is likely that in this case, with the worsening of scoliosis, there may

Figure 1. Flow-volume loop of the patient. Abnormal shape at the expiratory limb of the flow-volume loop (black arrow).

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Figure 2. Dynamic chest CT on inspiration (2a); innominate artery (white arrow) and third thoracic vertebra (black arrow) are identified. Dynamic chest CT on expiration (2b).

Figure 3. 3D spiral CT reconstructions. Anterior (3a) and lateral (3b) views.

have been significant compression on the trachea for a prolonged period. It is important to undertake a full pulmonary

evaluation in all patients with scoliosis who are scheduled to undergo surgery in order to prevent complications.

References

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