A rare cause of respiratory distress in infants: tracheal compression due to anomalous course of innominate artery

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Incomplete vascular ring is a rare congenital vascular anomaly causing tracheal compression. A case of anomalous innominate artery causing tracheal compression and low grade tracheomalacia with remarkable dyspnea and intermittent apnea in a five-month-old infant is reported. Chest X-ray revealed enlarged upper mediastinum. Patent foramen ovale and thymus hyperplasia were detected on echocardiographic examination. Angiography demonstrated left aortic arch with normal arterial branching pattern. Thorax computerized tomography (CT) revealed tracheal compression of more than 50% at innominate artery level and aberrant position of the hyperplastic thymus. Operation was planned for subtotal thymectomy and relocation of innominate artery in order to relieve tracheal obstruction. Postoperative thorax CT demonstrated satisfactory relief of tracheal compression. Patient was discharged from hospital on sixth postoperative day in good condition. On clinical follow-up he is completely asymptomatic without any signs of dyspnea or intermittent reflex apnea. In severely symptomatic cases, surgical treatment offers excellent results.

Key words: respiratory distress, infants, tracheal compression, anomalous course of innominate artery.

Incomplete vascular ring is a rare congenital vascular anomaly. In many cases symptoms are mild, but some patients may develop severe symptoms of airway obstruction and reflex apnea. We would like to report a case of anomalous innominate artery and aberrant hyperplastic thymus causing tracheal compression with remarkable dyspnea and intermittent apnea in an infant.

Case Report

The patient was a five-month-old boy who had progressive respiratory distress and intermittent reflex apnea. He was referred to our clinic for detailed examination of the underlying etiology including congenital heart disease. On his physical examination noisy respiration with biphasic stridor and mild perioral cyanosis were noted. Chest X-ray revealed enlarged upper mediastinum (Fig. 1). Patent foramen ovale and thymus hyperplasia were detected on echocardiographic examination. Angiography demonstrated left aortic arch with normal arterial branching pattern. Thorax computerized

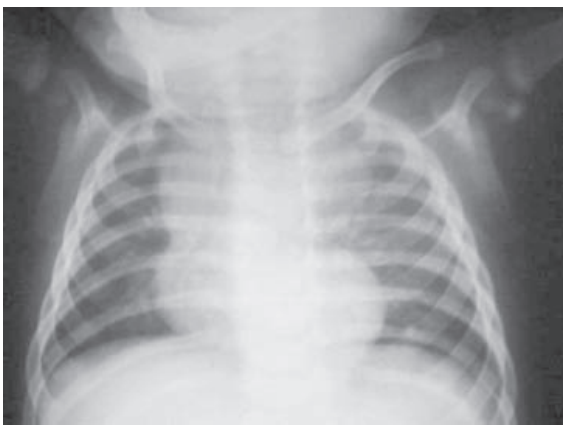

Fig. 1. Preoperative chest X-ray demonstrating enlarged upper mediastinum (aberrant position of hyperplastic thymus).

tomography (CT) revealed tracheal compression of more than 50% at innominate artery level and aberrant position of the hyperplastic thymus (Fig. 2a). Direct laryngoscopy demonstrated grade 1 tracheomalacia. Operation was planned for subtotal thymectomy and relocation of innominate artery in order to relieve tracheal obstruction.

After median sternotomy and subtotal resection of thymus gland, anomalous origin and course of the innominate artery from the left aortic arch were detected. The innominate artery was crossing over the trachea from left to right in an oblique fashion and the central portion of the trachea above the bifurcation of left and right main bronchus was compressed (Fig. 3a, 3b). Relocation of innominate artery by arteriopexy technique was preferred as the surgical treatment. To accomplish suspension of the artery, several superficial sutures were placed on the innominate artery and then these sutures were fixed to the posterior aspect of the right side of the upper sternum.

Postoperative period was uneventful and the patient was extubated three hours after the operation without any problem. Postoperative thorax CT demonstrated satisfactory relief of tracheal compression (Fig. 2b). Patient was discharged from hospital on sixth postoperative day in good condition. On clinical follow-up he is completely asymptomatic without any signs of dyspnea or intermittent reflex apnea.

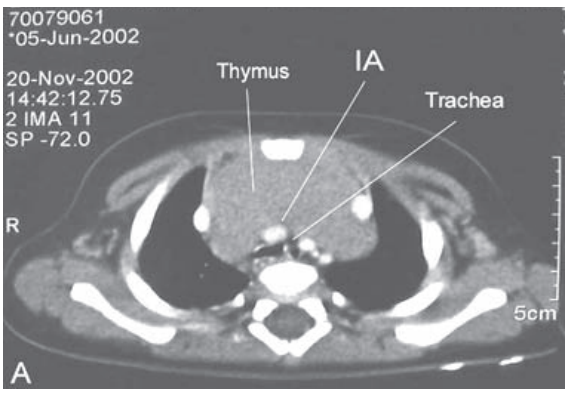

Fig. 2a. Preoperative thorax CT demonstrating enlarged thymus and innominate artery compressing the trachea. IA: Innominate artery.

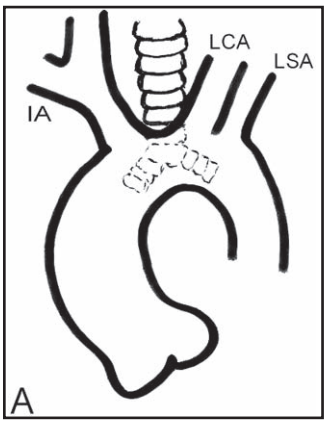

Fig. 3a. Normal branching pattern of left aortic arch. IA: Innominate artery, LCA: Left carotid artery, LSA: Left subclavian artery.

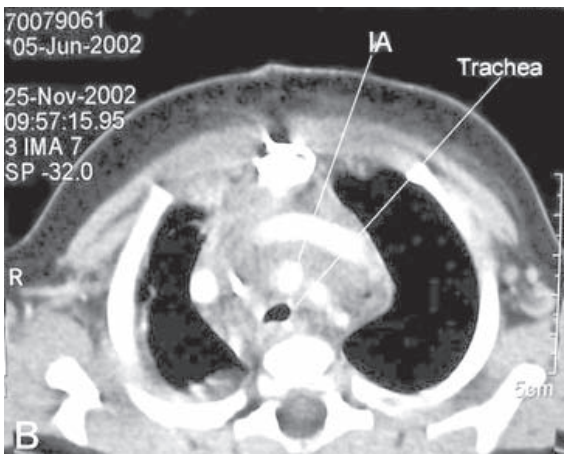

Fig. 2b. Postoperative thorax CT demonstrating position of innominate artery and decompressed trachea after subtotal thymectomy and arteriopexy. IA: Innominate artery.

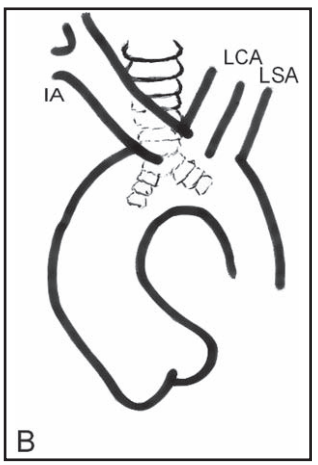

Fig. 3b. Anomalous origin and course of innominate artery resulting in tracheal compression. IA: Innominate artery, LCA: Left carotid artery, LSA: Left subclavian artery.

Discussion

Tracheobronchial compression of vascular origin is an uncommon but important cause of respiratory distress in infants and children. Nonspecific respiratory symptoms may result in delayed diagnosis[1,2]. Anomalous course of innominate artery is a rare form of this entity. Most of the patients have mild symptoms but in some cases it can cause severe stridor, cyanosis and apnea. Failure to diagnose and treat this pathology may result in progressive respiratory failure and even death. Tracheoscopy is a standard method for diagnosis but in recent years magnetic resonance (MR) and CT imaging of the thorax is more frequently used for the evaluation of vascular and airway abnormalities[3]. In severely symptomatic cases surgical treatment is mandatory. Several techniques have been described for the correction of the pathology, and suspension of the innominate artery to the sternum is the widely accepted treatment[4]. It is a simple and effective technique with low risk and good long-term results. Reimplantation technique is another alternative but division and reimplantation of innominate artery carries potential risks of bleeding, stroke, and early and late stenosis at the anastomotic site. Another anomaly that may coexist with anomalous course of the left innominate artery is aberrant position of hyperplastic thymus. The degree of tracheal stenosis may become significant with compression of the artery and thymus. Subtotal thymectomy and arteriopexy procedure might be necessary as in our case[5,6].

Left innominate artery is a rare form of incomplete vascular ring anomalies and most of the patients have mild symptoms of tracheal compression. In severely symptomatic cases, surgical treatment is needed. Relocating the innominate artery without ligation or reimplantation is a safe and effective method.

REFERENCES

1. Bove T, Demanet H, Casimir G, Viart P, Goldstein JP, Deuvaert FE. Tracheobronchial compression of vascular origin. Review of experience in infants and children. J Cardiovasc Surg 2001; 42: 663-666.
2. Woods RK, Sharp RJ, Holcomb GW 3rd, et al. Vascular anomalies and tracheoesophageal compression: a single institution's 25-year experience. Ann Thorac Surg 2001; 72: 434-438.
3. Faust RA, Rimell FL, Remley KB. Cine magnetic resonance imaging for evaluation of focal tracheomalacia: innominate artery compression syndrome. Int J Pediatr Otorhinolaryngol 2002; 65: 27-33.
4. Adler SC, Isaacson G, Balsara RK. Innominate artery compression of the trachea: diagnosis and treatment by anterior suspension. A 25-year experience. Ann Otol Rhinol Laryngol 1995; 104: 924-927.
5. Hennington MH, Detterbeck FC, Molina PL, Wood RE. Innominate artery and tracheal compression due to aberrant position of the thymus. Ann Thorac Surg 1995; 59: 526-528.
6. Mandell GA, McNicholas KW, Padman R, Harcke HT. Innominate artery compression of the trachea: relationship to cervical herniation of the normal thymus. Radiology 1994; 190: 131-135.