CASE REPORT

A new syndrome: right-sided arcus aorta syndrome

Sevket Ozkaya,¹ Adem Dirican,¹ Tibel Tuna,² Ferah Ece³¹Department of Pulmonary Medicine, Samsun Medical Park Hospital, Samsun, Turkey²Department of Pulmonary Medicine, Samsun Chest Diseases and Thoracic Surgery Hospital, Samsun, Turkey³Department of Pulmonary Medicine, Faculty of Medicine, Bahcesehir University, Istanbul, Turkey**Correspondence to**Dr Sevket Ozkaya,
ozkayasevket@yahoo.com**SUMMARY**

Right-sided arcus aorta (RSAA) is a rare congenital anomaly that may cause chronic cough and dyspnoea. We aimed to define RSAA as a new syndrome characterised by RSAA, dyspnoea and cough during exercise. RSAA syndrome should be included in the differential diagnosis of asthma.

BACKGROUND

Right-sided arcus aorta (RSAA) is a rare congenital anomaly that is usually asymptomatic. However, it may cause chronic cough and dyspnoea, especially during exercise. Two recent reports suggested its role in asthmatic attacks as in our case that justifies its inclusion in the differential diagnosis of asthma.^{1 2} It is reported to occur in 0.1–0.2% of adults.³ Our aim is to define RSAA as a new syndrome characterised by RSAA, dyspnoea and cough during exercise.

CASE PRESENTATION

A 4-year-old girl was admitted to the hospital with chronic cough and dyspnoea, which started 6 months ago. She was receiving asthma treatment, but she suffered from dyspnoea, especially during exercise. She had begun training in gymnastics in the last 6 months.

OUTCOME AND FOLLOW-UP

The physical examination was normal. Chest roentgenogram showed the absence of arcus aorta on the left side and there was an enlargement on the shadow of right upper mediastinal area (figure 1).

Figure 1 Chest roentgenogram showed that there is no shadow of arcus aorta on the left side, with enlargement on shadow of the right upper mediastinal area.

Figure 2 The flow–volume curves on spirometry pointed out tracheal compression.

The flow–volume curves on spirometry pointed out tracheal compression (figure 2). We suspected an RSAA from the tracheal compression. A thorax CT confirmed the RSAA and the tracheal compression (figure 3).

Written informed consent was obtained from patients and institutions to use their records for the study. Consent was also obtained for publication of the case report with the accompanying images.

DISCUSSION

Assman first described the roentgenographic studies of this anomaly and Renander described the first case reported in the literature.⁴ We previously reported the incidence of RSAA as 0.16%. RSAA is usually asymptomatic. According to previous reports, some patients exhibit dysphagia and dyspnoea during exercise.^{1 2 4 5} Some patients are clinically diagnosed with exercise-induced asthma, as in our case. We reported 13 patients with RSAA and seven (54%) of them were symptomatic due to external compression of the trachea and oesophagus as a result of RSAA.² The RSAA anomaly may be more common in the community than the ratio estimated from the number of patients who are detected. Our patient was born in 2008 in Samsun (Turkey). In the same year, the number of new born was 18 916. The estimated incidence of RSAA was 1/18 916.

We suspected the patient to have RSAA because of lack of arcus aorta on the left side of mediastinum with chest radiography. Thorax CT and thorax MRI are the best methods for diagnosis.

MRI is the preferable diagnostic tool. We chose CT because our patient was a child. In addition the external tracheal compression due to RSAA was visible in the CT scan of the thorax.

To cite: Ozkaya S, Dirican A, Tuna T, et al. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2013-009569

Figure 3 (A and B) Thorax CT scans showed right-sided arcus aorta and tracheal compression.

Learning points

- ▶ We think this is a new clinical entity including right-sided arcus aorta (RSAA), dyspnoea and cough during exercise.
- ▶ RSAA syndrome should be included in the differential diagnosis of asthma, and all patients with intractable and exertional dyspnoea should be evaluated for RSAA anomaly.
- ▶ We suggest that RSAA together with dyspnoea and cough during exercise is a new clinical entity that should be called RSAA syndrome.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

REFERENCES

- 1 Ozkaya S, Sengul B, Hamsici S, *et al.* Un usual cause of dyspnea. *J Asthma* 2010;47:946–8.
- 2 Ozkaya S, Sengul B, Hamsici S, *et al.* Right sided arcus aorta as a cause of dyspnea and chronic cough. *Multidiscip Respir Med* 2012;7:37.
- 3 D'Souza VJ, Velasquez G, Glass TA, *et al.* Mirror image right aortic arch: a proposed mechanism in symptomatic vascular ring. *Cardiovasc Intervent Radiol* 1985;8:134–6.
- 4 Bevelaqua F, Schicci JS, Haas F, *et al.* Aortic arch anomaly presenting as exercise-induced asthma. *Am Rev Respir Dis* 1989;140:805–8.
- 5 Lunde R, Sanders E, Hoskam JAM. Right aortic arch symptomatic in adulthood. *Neth J Med* 2002;60:212–15.

Copyright 2013 BMJ Publishing Group. All rights reserved. For permission to reuse any of this content visit <http://group.bmj.com/group/rights-licensing/permissions>.
BMJ Case Report Fellows may re-use this article for personal use and teaching without any further permission.

Become a Fellow of BMJ Case Reports today and you can:

- ▶ Submit as many cases as you like
- ▶ Enjoy fast sympathetic peer review and rapid publication of accepted articles
- ▶ Access all the published articles
- ▶ Re-use any of the published material for personal use and teaching without further permission

For information on Institutional Fellowships contact consortiasales@bmjgroup.com

Visit casereports.bmj.com for more articles like this and to become a Fellow