Aberrant right subclavian artery syndrome: A case of chronic cough

LTC Patricio Rosa, MD, LTC David L. Gillespie, MD, LTC James M. Goff, MD, LTC Sean D. O'Donnell, MD, and **MAJ Benjamin Starnes, MD,** *Tripler Army Medical Center, Hawaii; and Washington, DC*

A young, otherwise healthy man had chronic cough of 16 months' duration. Evaluation revealed an aberrant right subclavian artery. Kommerell's diverticulum without aneurysmal degeneration was present. Imaging studies showed compression of the esophagus but not the trachea. Results of methacholine challenge test were negative for evidence of reactive airway disease, but suggested mild variable intrathoracic obstruction. While aberrant right subclavian artery syndrome most commonly involves dysphagia, our patient's only symptom was cough. Right subclavian artery to right common carotid artery transposition was performed, with oversewing of the subclavian artery stump to the left of the esophagus through a right supraclavicular incision. This treatment was curative, with complete resolution of symptoms. (J Vasc Surg 2003;37:1318-21.)

The right subclavian artery has an aberrant origin in 0.5% to 1% of the population.[1-3] Presence of associated symptoms is, however, even more uncommon. The most common symptoms include dysphagia, cough, and stridor, which are usually associated with evident compression of the appropriate structure. Of these, dysphagia is probably the most recognized symptom, and gives rise to the term, dysphagia lusoria.[2] The origin of the aberrant artery is usually broad based and is known as Kommerell's diverticulum.[1,4] The diverticulum may undergo aneurysmal degeneration, which is associated with a high incidence of rupture if untreated. Our patient had chronic cough only and no evident tracheal compression on any imaging study. Kommerell's diverticulum was present, but without aneurysmal degeneration. Right subclavian artery to carotid artery transposition was curative.

CASE REPORT

A 23-year-old male active duty soldier had a 1-year history of chronic persistent nonproductive cough. He denied dysphagia, pyrosis, or chronic nasal congestion. He was otherwise healthy, and had quit smoking 6 months earlier. He frequently had as many as 15 small coughs per minute, which he thought originated in the throat. As part of his duties he was required to run on a regular basis. The cough, however, interfered with his ability to run, increasing his 2-mile time by 4 minutes over the course of 1 year. Physical examination revealed clear bilateral breath sounds, with no wheezes, ronchi, rales, or cardiac murmurs. The initial differential diagnosis included asthma, gastroesophageal reflux disease, and, less likely, a vascular malformation. A radiograph of the chest was normal. Pulmonary function tests revealed forced expiratory volume in 1 second (FEV_1) of 4.64 L and forced vital capacity (FVC) of 5.56 L. Results of methacholine challenge test were negative for any evidence of reactive airway disease, although there was some flattening of the middle and late test flow loops, suggestive of mild and variable intrathoracic airway obstruction. A barium esophagram showed extrinsic compression of the esophagus at the thoracic inlet (Fig 1), but failed to demonstrate any evidence of reflux or esophagitis. Esophagogastroduodenoscopy confirmed a pulsatile extrinsic compression at 23 cm from the gums, without associated mucosal abnormalities or evidence of esophagitis. Omeprazole, 20 mg twice a day, was empirically started, with no change in symptoms. A computed tomography (CT) scan of the chest showed an aberrant right subclavian artery with a broad base, or Kommerell's diverticulum, but no aneurysm. There was mild compression of the esophagus, but no compression of the trachea (Fig 2). Fiberoptic bronchoscopy showed no tracheomalacia, obstruction, mucosal abnormalities, or any findings suggestive of chronic aspiration. Contrast medium-enhanced arch arteriography (Fig 3) confirmed presence of the aberrant right subclavian artery, as well as a common carotid trunk.

After reviewing all data and after an extensive counseling session with the patient, the decision was made to proceed with surgical intervention. Right subclavian artery to right common carotid artery transposition was performed, with oversewing of the subclavian artery stump to the left of the esophagus. This was done through a right supraclavicular incision. This technique has been previously described by Kieffer et al.[2] The cough completely resolved immediately postoperatively. Recovery was complete and uneventful, except for a minor superficial wound problem that healed satisfactorily. On follow-up at 3 and 5 weeks postoperatively the patient remained cough-free.

DISCUSSION

Aberrant right subclavian artery is the result of an embryologic defect, although its embryologic origin remains a subject of debate.[1,2,5] When aberrant, however, the artery comes off the aortic arch beyond the left subclavian artery and crosses the midline, most often dorsal to the esophagus (80%), but may also pass between the esophagus

From Tripler Army Medical Center and Walter Reed Medical Center.

Competition of interest: none.

Reprint requests: LTC Patricio Rosa, MD, Chief, Vascular Surgery Service, MCHK-DSV, 1 Jarrett White Rd, Tripler Army Medical Center, HI 95859 (e-mail: Patricio.Rosa!haw.tamc.amedd.army.mil).

0741-5214/2003/$30.00 + 0
doi:10.1016/S0741-5214(02)75464-5

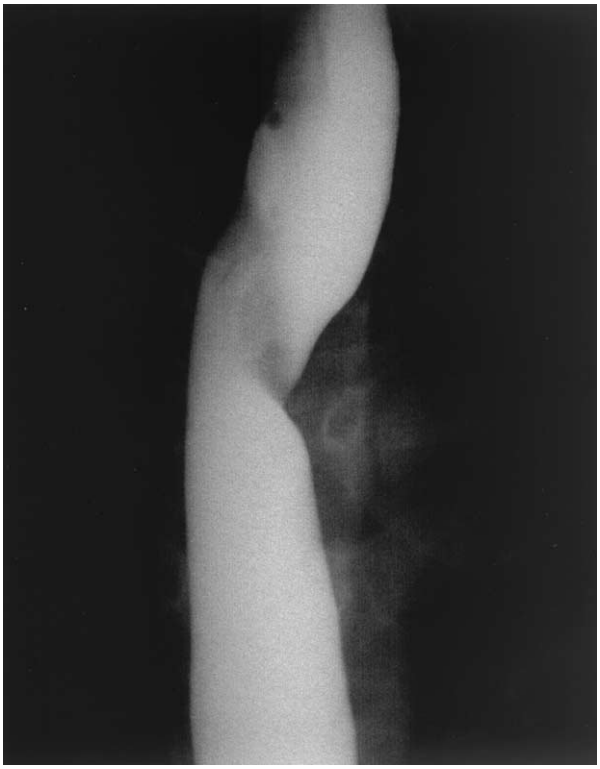

Fig 1. Barium esophagogram demonstrates extrinsic compression.

and the trachea (15%) or even anterior to the trachea (5%).[1,4,5] The artery most commonly arises from the posterolateral portion of the aortic arch. It is the persistent proximal portion of the dorsal aorta that results in formation of a diverticulum of Kommerell,[1,4,6,7] a tapered broadening of the proximal portion of the artery. This is the most common congenital arch abnormality,[4] present in 0.5% to 1% of the population.[1-3] Most patients are free of symptoms, and the diagnosis is made by incidentally finding the abnormality on imaging studies obtained for other reasons. Confusion seems to also exist in the literature regarding the term, Kommerell's diverticulum. Some authors use the term to refer to the aneurysmal degeneration sometimes found at the origin of the artery,[2,4] whereas others use it to refer to the natural broadening of the origin of the aberrant artery as a consequence of the persistent dorsal aorta.[1,3,6,7]

Other abnormalities commonly associated with aberrant right subclavian artery may be of interest to the surgeon. A common carotid trunk may be present. In such cases the ipsilateral vertebral artery may arise from the common carotid artery and may have an anomalous point of entrance into the cervical spine. The thoracic duct may end in the right jugulosubclavian junction. A nonrecurrent laryngeal nerve, important during thyroid surgery, will extend almost directly across from the vagus nerve to the laryngeal territory.[1,2]

Symptoms associated with aberrant right subclavian artery involve the gastrointestinal and respiratory tracts. Symptoms usually arise from the compressive effect of the artery or aneurysm on the surrounding structures. Dysphagia is the most common and best recognized symptom, and gave rise in the past to the term, dysphagia lusoria.[2] Pres-

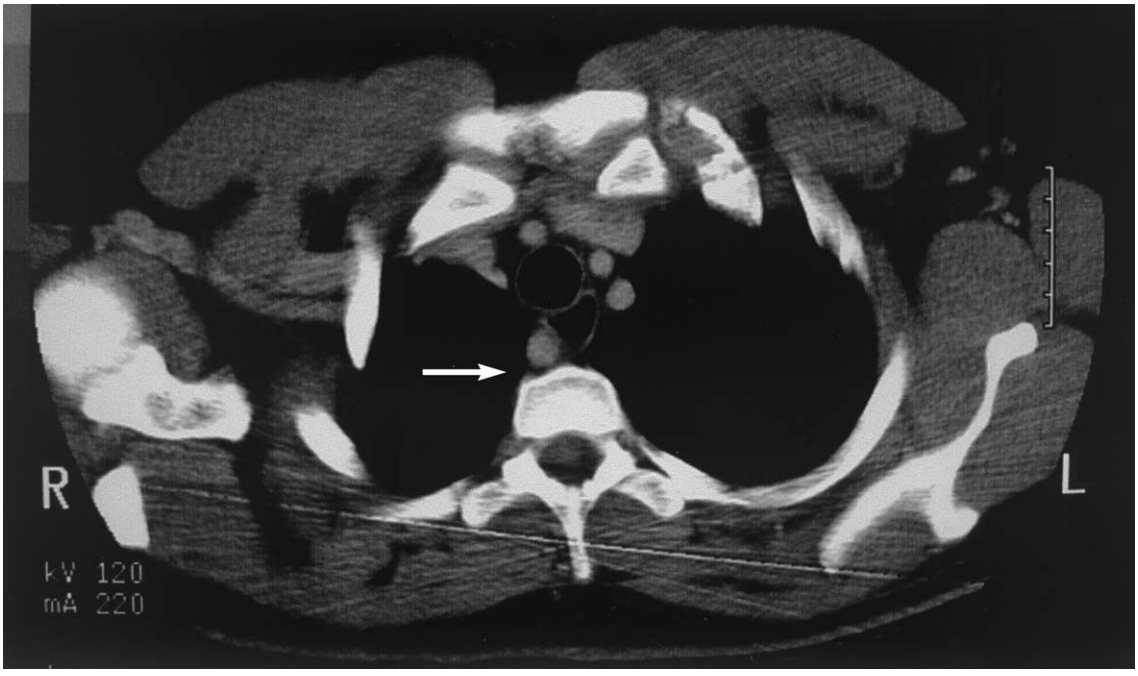

Fig 2. CT scan demonstrates aberrant right subclavian artery *(arrow)*.

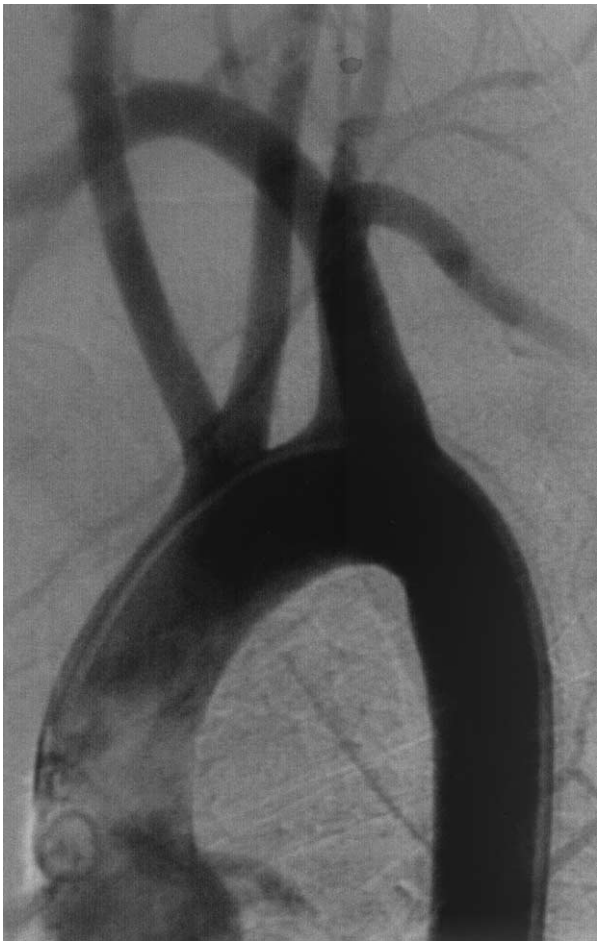

Fig 3. Arch aortogram demonstrates aberrant right subclavian artery and common carotid trunk.

ence of aneurysmal dilatation of the aberrant right subclavian artery or Kommerell's diverticulum seem to serve as very clear explanations for the symptoms of dysphagia. The presence of a common carotid trunk has also been implicated as a possible contributing factor. When either of these structures is present and compresses the tracheobronchial tree, presence of respiratory symptoms also seems to have a clear explanation. In the absence of these abnormalities, the explanation for symptoms is not clear.

Once the diagnosis of aberrant right subclavian artery has been entertained or made, further evaluation is indicated only in patients whose symptoms are suspected to be related to the condition or who have associated aneurysmal degeneration. Aneurysms are almost always found at or near the origin of the artery, and are suspected to result from degeneration of a diverticulum of Kommerell. These aneurysms should always be treated[2] because, even if asymptomatic, they can rupture, thrombose, or embolize.[8] Further evaluation should be guided by the symptoms, and should be oriented toward identifying other potential causes for the symptoms. If no other disease explains the symptoms, preoperative surgical evaluation should include CT and biplanar arteriography.[9] These two studies combined will provide detailed information about the size of an aneurysm, if present, as well as to the relationship between the aorta and its branches. Magnetic resonance imaging does not usually add any further information than gained from CT and arteriography.[2] The key issue in treatment involves determination of whether an aneurysm is present. Although multiple approaches have been reported,[1] in the absence of an aneurysm the condition is successfully treated with right subclavian artery to right common carotid artery transposition. This can be accomplished through a right supraclavicular incision.[10-12] Dissection and transection of the artery should be carried over to the left side of the esophagus. This accomplishes removing the compression effect from the esophagus and trachea, and revascularization of the right upper extremity simultaneously. The presence of Kommerell's diverticulum should not modify this approach, because the theoretical problem of aneurysmal degeneration of the diverticulum should be averted by thrombosis of the stump. When the aberrant right subclavian artery is aneurysmal, the aneurysm needs to be addressed. Various approaches have been described.[11,13,14] The preferred approach is left posterolateral thoracotomy.[2] This approach provides direct access to the origin of the aberrant right subclavian artery. If the aberrant right subclavian artery has not been previously reconstructed, vertebrobasilar insufficiency is a risk, and cardiopulmonary bypass should be considered.[2] In our patient the smooth, tapered dilatation of the origin of the aberrant right subclavian artery is consistent with Kommerell's diverticulum without aneurysmal degeneration. The patient was advised to undergo long-term follow-up to ensure that the diverticulum remains unchanged.

Our case differs from others we have reviewed in the literature in that the cough, which was the patient's only symptom, occurred in the absence of radiographically evident tracheal compression. It is notable, however, that the flow volume loops suggested variable intrathoracic obstruction. This is a common finding in clear cases of vascular compression,[15] and may serve as sufficient evidence of compression of the trachea by the aberrant artery in the absence of radiographic evidence. Although the possibility of aberrant right subclavian artery was considered in the initial differential diagnosis, absence of evident tracheal compression, and absence of literature suggesting resolution of symptoms after surgery in such circumstances, made us initially uneasy about intervening. The literature review clearly indicates that aberrant right subclavian artery is often associated with cough, most often in the presence of dysphagia, but also sometimes by itself, but this is typically associated with evident tracheal compression. As further evaluation continued, however, all other possible explanations were ruled out. Furthermore, the patient was a young, otherwise healthy adult whose activities were limited by chronic cough, with no other explanation than aberrant right subclavian artery. After a long counseling session with the patient, the decision was made to

proceed with surgical treatment of the aberrant right subclavian artery. Symptoms completely resolved. It is not clear exactly what the mechanism for the cough was, in the absence of visible tracheal compression. We hypothesize, however, that pressure of pulsation of the artery being transmitted to the airways caused the cough reflex. Presence of a vascular ring would more easily explain the symptoms; however, all imaging studies failed to demonstrate a vascular ring or any anterior compression. One could perhaps hypothesize that the common carotid trunk anteriorly, in combination with the aberrant right subclavian artery posteriorly, constituted a functional vascular ring. Once the aberrant subclavian artery was divided, compression was relieved. Still, however, no evidence of anterior compression is demonstrated. In our patient, the common carotid trunk appeared to be of no consequence because, although this has been implicated in dysphagia, our patient did not have dysphagia.

We recommend that, although the decision to intervene in a patient with chronic cough and aberrant right subclavian artery without evident tracheal compression must be individualized, intervention in the absence of other explanations should be considered. The presence of variable intrathoracic tracheal obstruction in flow volume loops obtained at spirometry may support this decision.

REFERENCES

1. Stone WM, Brewster DC, Moncure AC, et al. Aberrant right subclavian artery: varied presentations and management options. J Vasc Surg 1990;11:812-7.
2. Kieffer E, Bahnini A, Koskas F. Aberrant subclavian artery: surgical treatment in thirty-three adult patients. J Vasc Surgery 1994;19:100-11.
3. Bisognano JD, Young B, Brown JM, et al. Diverse presentation of aberrant origin of the right subclavian artery: two case reports. Chest 1997;112:1693-7.
4. Brown DL, Chapman WC, Edward WH, et al. Dysphagia lusoria: aberrant right subclavian artery with a Kommerell's diverticulum. Am Surg 1993;59:582-6.
5. Hamilton WJ, Mossman HW. Cardio-vascular system. In: Human embryology. 4th ed. Cambridge, England: W Heffer & Sons Ltd; 1972. p 267-71.
6. Copleman B. Anomalous right subclavian artery. Am J Roentgenol 1945;54:270-5.
7. Cina CS, Arena GO, Bruin G, Clase CM. Kommerell's diverticulum and aneurysmal right-sided aortic arch: a case report and review of the literature. J Vasc Surg 2000;32:1208-14.
8. Dougherty MJ, Allegro KD, Savarese RP, et al. Atherosclerotic aneurysm of the intrathoracic subclavian artery: a case report and review of the literature. J Vasc Surg 1995;21:521-9.
9. Jebara VA, Arnaud-Crozat E, Angel F, et al. Aberrant right subclavian artery aneurysm: report of a case and review of the literature. Ann Vasc Surg 1989;3:68-73.
10. Valentine RJ, Carter DJ, Clagett GP. A modified extrathoracic approach to the treatment of dysphagia lusoria. J Vasc Surg 1987;5:498-500.
11. Harrison LH Jr, Batson RC, Hunter DR. Aberrant right subclavian artery aneurysm: an analysis of surgical options. Ann Thorac Surg 1994;57:1012-4.
12. Taylor M, Harris KA, Casson AG, et al. Dysphagia lusoria: extrathoracic surgical management. Can J Surg 1996;39:48-52.
13. Kalke BR, Magotra R, Doshi SM. A new surgical approach to the management of symptomatic aberrant right subclavian artery. Ann Thorac Surg 1987;44:86-9.
14. Pome G, Vitali E, Mantovani A, et al. Surgical treatment of the aberrant retroesophageal right subclavian artery in adults (dysphagia lusoria): report of two new cases and review of the literature. J Cardiovasc Surg (Torino) 1987;28:405-12.
15. Parker JM, Cary-Freitas B, Berg BW. Symptomatic vascular rings in adulthood: an uncommon mimic of asthma. J Asthma 2000;37:275-80.

Submitted Aug 29, 2002; accepted Nov 26, 2002.