

CASE REPORT

Chronic cough: a herald symptom of thoracic aortic aneurysm in a patient with a bicuspid aortic valve

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SUMMARY

Chronic cough has a wide differential, of which thoracic aortic aneurysm is a rare but potentially devastating cause. We present a patient with previous aortic valve replacement for a bicuspid valve who had been suffering from a chronic cough for 8 months and who developed subsequent voice hoarseness. This had been initially managed in the community with trials of steroid inhalers, steroid nasal sprays and proton pump inhibitors to no avail. He was referred to cardiology and chest clinics. An urgent CT aortogram was requested given his widened mediastinum on chest radiograph, cardiac history of bicuspid valve and symptoms. This revealed a large aneurysm of the thoracic aorta with chronic dissection that required urgent operative intervention. His cough resolved 6 weeks postoperatively. The purpose of this report is to highlight thoracic aortic aneurysms as a potential rare differential for chronic cough and as a complication of patients with bicuspid aortic valves.

BACKGROUND

Cough is a common presentation in all areas of medicine. There is a wide differential and often the cause is respiratory or cardiac in origin. In primary care initial investigations can be performed and it is not unusual to trial treatments for common causes. This includes gastro-oesophageal reflux, postnasal drip and asthma.¹ This is dependent on investigations and clinical suspicion.² Rarer causes must also be considered especially when there is treatment failure. Cough has been reported to be secondary to thoracic aortic aneurysms in the literature.³ It is presumed to be secondary to bronchial compression from local mass effect.⁴

We present a patient with a bicuspid aortic valve, thus prone to aortic root enlargement, presenting with chronic cough with no apparent cause. Given widening of the mediastinum on his chest radiograph in conjunction with his new symptoms there were concerns there may be local compression from a thoracic ascending aortic aneurysm. Urgent imaging was requested which revealed the diagnosis. There had been a significant delay of 8 months to diagnosis, which is considerable when there was a clear risk factor. Timely diagnosis of thoracic aneurysm is important given that early medical or surgical intervention can prevent the considerable morbidity and mortality associated with rupture and dissection.

CASE PRESENTATION

A 73-year-old man had been referred to secondary care, including a cardiology clinic, by his general

practitioner (GP) for further clinical assessment of his chronic cough. He had a background of a bicuspid aortic valve for which he had had a metallic aortic valve replacement in 2002 for mixed aortic valve disease. He also had a medical history of atrial fibrillation. The patient was a non-smoker with no history of asthma. His regular medications included bisoprolol and warfarin.

He had been experiencing chronic cough for approximately 8 months. The cough was predominantly dry but occasionally productive of clear sputum. The cough occurred at rest and on exertion. There was associated voice hoarseness in the preceding months. No chest pain, haemoptysis or wheeze had been reported.

He had been treated with proton pump inhibitors, steroid nasal sprays and steroid inhalers for some time by his GP with no success in alleviating his symptoms. He had also been referred for a chest radiograph and spirometry which were reported as unremarkable.

On examination his pulse was irregularly irregular. There was an audible metallic second heart sound with no associated diastolic murmur. Chest auscultation revealed normal breath sounds throughout, there was no peripheral oedema and no evidence of lymphadenopathy.

INVESTIGATIONS

From clinic he was sent for a repeat chest radiograph that showed evidence of a widened mediastinum with no evidence of pulmonary oedema or effusions (figure 1). Given this finding on a background of surgically replaced bicuspid aortic valve he was referred for urgent CT aortogram which showed evidence of a fusiform aneurysm of the



Figure 1 Chest radiograph demonstrating widened mediastinum.



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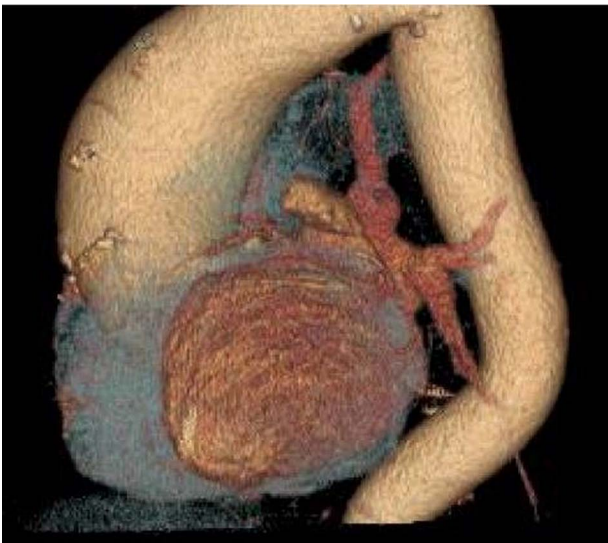


Figure 2 CT three-dimensional volume rendered reconstruction of thoracic aortic aneurysm.

ascending aorta extending from the aortic root to the region of the origin of the left common carotid artery. It had maximum dimensions of 6.9 cm×6.5 cm (anteroposterior×lateral) in the mid ascending aorta and 6×7 cm (anteroposterior×lateral) at the aortic root. A CT three-dimensional volume rendered reconstruction of the thoracic aortic aneurysm demonstrated the aneurysm well (figure 2). There was an associated small dissection flap in the ascending thoracic aorta (figure 3). Interestingly the cross-sectional imaging showed evidence of compression of the right bronchus intermedius (figures 4 and 5). He was referred for surgical intervention. Prior to this he was also seen by a respiratory physician, given cough was his primary symptom, who excluded an underlying respiratory cause.

DIFFERENTIAL DIAGNOSIS

The differential is broad, including cardiac causes such as pulmonary oedema, and respiratory causes such as pulmonary malignancy, postnasal drip, interstitial lung disease and asthma.



Figure 3 Cross-sectional CT image of ascending thoracic aneurysm with associated dissection flap (red arrow indicates dissection flap).

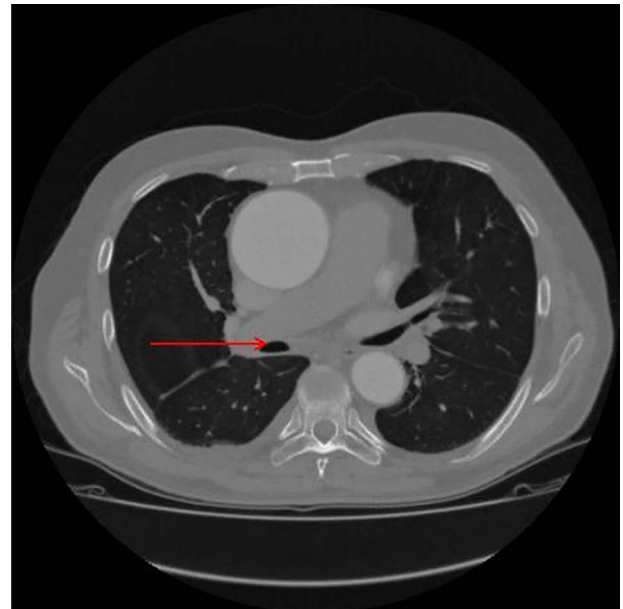


Figure 4 Cross-sectional CT image demonstrating ascending aortic aneurysm with associated mild to moderate right bronchus intermedius compression (red arrow indicates right bronchus intermedius). Image represents the lung window.

Gastrointestinal causes such as gastro-oesophageal reflux disease also need to be considered.

TREATMENT

This patient was optimised medically prior to referral for aortic root replacement.

OUTCOME AND FOLLOW-UP

The patient made a good recovery post aortic root replacement. Interestingly approximately 6 weeks postsurgery his cough

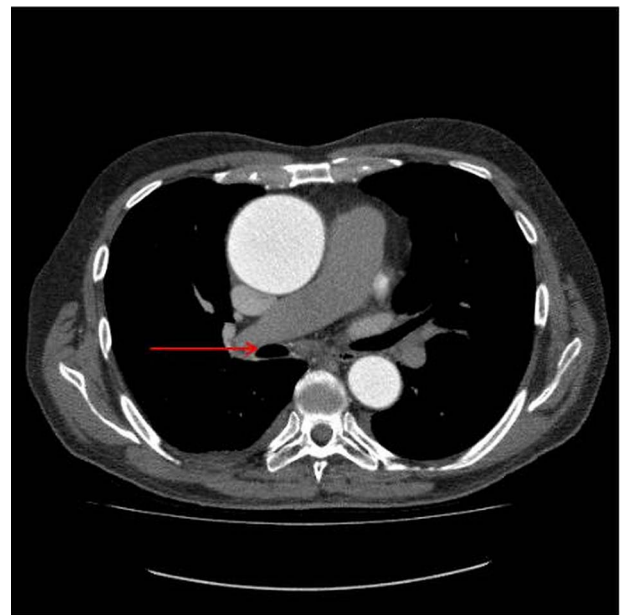


Figure 5 Cross-sectional CT image demonstrating ascending aortic aneurysm with associated mild to moderate right bronchus intermedius compression (red arrow indicates right bronchus intermedius). Image represents the mediastinal window.

resolved to great relief of the patient. He is now being followed up routinely in cardiology clinic.

DISCUSSION

Cough is a very common symptom assessed and investigated both in a primary and secondary care setting. It is considered chronic when experienced for a duration of greater than 8 weeks.² There are a myriad of causes ranging from the benign to the potentially life threatening. Therefore, a detailed history and examination must be performed alongside basic investigations. A chest radiograph and spirometry are recommended as compulsory.² In conjunction, the consultation and basic investigations can identify many potentially important causes. These include respiratory causes of cough such as malignancy, pulmonary fibrosis and chronic obstructive pulmonary disease. This also includes important cardiac and vascular causes such as pulmonary oedema and thoracic aortic aneurysms. Trials of treatment can be used to help identify the cause of cough such as in suspected cases of gastro-oesophageal reflux (GORD), sinusitis and asthma. The initial investigations and management may not yield a diagnosis and further referral to specialist clinics/secondary care must be considered.

In this case, in primary care, relevant investigations were performed as well as treatment trials for asthma, postnasal drip and GORD. The initial chest radiograph was reported as unremarkable. It is possible that the mediastinal widening was more subtle at this time. Interestingly the chest radiograph with mediastinal widening, requested by our team (figure 1), was reported as an unfolded aorta. This is a common non-specific finding that includes aneurysm and tortuous aorta as causes for mediastinal widening. This case serves to demonstrate that genuine aneurysmal widening can be reported radiologically as aortic unfolding. Thus the pretest probability of chronic thoracic aortic aneurysm is very important when reviewing films demonstrating mediastinal widening. In this case the pretest probability was much higher with the presence of a bicuspid valve as a risk factor.

It is always important to follow recommendations and diagnostic pathways in finding any underlying diagnosis. However, in clinical medicine every aspect of a patient's case must be considered to generate the differential diagnosis and avoid diagnostic delay. Specifically, in this case of cough, this patient had a bicuspid valve which is not uncommon, with a population prevalence estimated at 0.5–2%.¹ These patients are known to be prone to aortic root dilation and thoracic aortic aneurysms. One study showed that bicuspid aortic valve patients with no thoracic aneurysm at diagnosis had a 26% risk of aneurysm formation after 25 years follow-up.⁵ If this risk factor was considered earlier this may have avoided significant diagnostic delay, approximately 8 months, that could have had devastating consequences.

In bicuspid valve patients some evidence suggests that the aortic wall is histologically similar to Marfan patients,^{6,7} which may explain the predisposition to aneurysm formation. In the bicuspid population regular surveillance of both the valve and aorta are recommended.¹

There are multiple other risk factors for thoracic aortic aneurysms with associations including hypertension, atherosclerosis and connective tissue disorders such as Marfan syndrome. Other causes of aortitis can predispose including giant cell arteritis and Takayasu arteritis.^{6,8}

Thoracic aneurysms can be asymptomatic and are often diagnosed incidentally.^{4,8} They can result in aortic regurgitation and result in subsequent symptoms of cardiac failure. A myriad of symptoms can occur from local mass effect. In this case the underlying anatomical reason for cough as a presenting factor is

due to presumed local mass effect causing compression of the trachea or bronchi.⁴ The patient also presented with a hoarse voice and it is postulated that stretching of the recurrent laryngeal nerve maybe the underlying pathophysiology for this phenomena.⁸ Cases may present with other symptoms of local mass effect including dysphagia secondary to oesophageal compression.⁸ Chest pain can be a symptom from mass effect or can signal onset of dissection or rupture.

Echocardiography typically can only visualise the proximal several centimetres of the aorta.⁹ Thus importantly, as with this case, complete imaging of the thoracic aorta should occur at serial intervals in order to identify subsequent thoracic aorta pathology or aneurysms. This can be achieved with contrast-enhanced CT or MR angiography. In this case, his symptoms of cough and voice hoarseness suggested that he may have developed an aortic aneurysm.

Medical management of patients with thoracic aortic aneurysms mainly includes management of hypertension as well as β -blockade to reduce aortic wall stress during systole. Second-line therapy includes ACE inhibitors and angiotensin receptor blockers in appropriate patients. Smoking cessation is also recommended.⁶

Often patients with bicuspid valves requiring valvular surgery will need aortic root replacement at the time of surgery. An aortic root diameter of 5 cm is an indication for surgery, or 4.5 cm if valvular surgery is also planned concurrently.¹ This surgery is undertaken to prevent future aneurysm complications including dissection or rupture.

It would appear in this case that cough and voice hoarseness represented the compressive effects of an expanding underlying aortic aneurysm. Along with the radiographic demonstration of bronchial compression and the resolution of his cough postsurgery it suggests that the aneurysm was the likely cause for his cough.

Learning points

- ▶ Ascending thoracic aortic aneurysm is a rare cause of cough.
- ▶ Congenital bicuspid valves are associated with aortic root dilation and thoracic aortic aneurysm.
- ▶ When confronted with a chest radiograph with widened mediastinum the pretest probability of thoracic aneurysm must be considered.
- ▶ Imaging modalities for the aorta include echocardiogram, CT and MRI.
- ▶ Management of thoracic aortic aneurysms has both medical and surgical components.

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Contributors CPM was directly involved in the case and is the main author and researcher of the article. SF contributed to the clinical narrative and gave input on suggestions and amendments to the article. EW contributed to the clinical narrative and gave input on suggestions and amendments of the article, he also obtained images and provided captions and explanations. AA helped contribute and give suggestions and amendments to the article.

Competing interests None.

Patient consent Obtained.

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