A Case of Hoarseness with Acute Back Pain - Cardiovocal Syndrome Revisited

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ABSTRACT

A previously well 81-year-old Chinese male presented with hoarseness and low back pain for one month. Chest radiography at presentation revealed widening of the mediastinal silhouette. Nasopharyngoscopy detected left vocal cord paralysis. CT aortogram revealed a large saccular aortic arch aneurysm with a dissection flap extending distally down to the aortic bifurcation. The combination of clinical and imaging findings was consistent with cardiovocal syndrome. In view of good premorbid function, surgical repair was offered, and the patient underwent surgical repair and recovered well with no further back pain. A review of cases of cardiovocal syndrome suggest that prognosis of recurrent laryngeal nerve paralysis is dependent on the degree and duration of compression, and usually persists despite treatment of the underlying aneurysm.

CASE REPORT

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An 81-year-old Southern-Chinese man presented with hoarseness and paravertebral mechanical low back pain of 1 month's duration. He had hyperlipidemia and was being treated with lifestyle and diet modifications, with no other significant medical or surgical history. There was no associated cough, sore throat or fever, or symptoms suggestive of an underlying infection. He did not have any hemoptysis, odynophagia or dysphagia and there were no significant constitutional symptoms such as loss of appetite or weight. He also did not have any personal or family history of cancer. He had a 4-pack year smoking history, but claimed to have quit 35 years ago, and did not take any alcohol. There was no neck trauma, intubation, or recent head and neck surgery.

Initial blood investigations were normal. Chest radiography done at presentation revealed widening of the mediastinal silhouette (Figure 1). Lumbar spine radiographs showed chronic degenerative changes with grade 1 anterolisthesis of L4 on L5 with preserved disc spaces, mild anterior wedging of L1 and multilevel endplate osteophytes. In view of prolonged hoarseness in an elderly Southern-Chinese male where incidence of nasopharyngeal carcinoma is high, an

otolaryngologic review was obtained to rule out a head-and-neck cancer. Bedside fiberoptic nasopharyngoscopy detected left vocal cord paralysis with a glottic gap of 2mm on full adduction. The anterior and posterior nasal space, and pharynx were normal, and no masses were noted. The impression was left vocal cord paralysis, complicated by moderate dysphonia.

Contrast enhanced CT Neck and CT aortogram revealed anteromedial deviation of the arytenoid cartilage and dilatation of the left laryngeal ventricle, consistent with left vocal cord paralysis (Figure 2) [1]. There was no laryngeal edema, vocal cord or anterior commissure laceration, soft tissue mass to suggest a nasopharyngeal malignancy, or arytenoid cartilage avulsion suggesting laryngitis or laryngeal trauma. The CT also revealed a large 9-centimeter saccular aneurysm with mural thrombus in the aortic arch (Figure 3) [2]. This aortic arch aneurysm was associated with a dissection flap beginning distal to the left subclavian artery origin and extending down to the level of the aortic bifurcation (Stanford type B) (Figure 4). There was no periaortic fluid, pericardial effusion, abnormal gas collection, fistula, or contrast extravasation noted. This saccular aortic arch aneurysm likely compressed the left recurrent laryngeal nerve, resulting in left vocal cord palsy. Careful attention was paid to review the entire pathway of the vagal nerve from skull base to the recurrent laryngeal nerve course in the thorax and no further lesions were noted. A 2D Echocardiogram confirmed involvement of the aortic arch but not the ascending aorta or aortic valves. Duplex ultrasound of his carotid arteries confirmed vascular patency. The etiology of the aortic arch aneurysm was likely secondary to atherosclerosis. There was no evidence to suggest that this was secondary to mycosis, syphilis or vasculitis.

In view of good premorbid function, the patient underwent a Thoraflex Hybrid Endovascular Frozen Elephant Trunk replacement of ascending aorta and aortic arch. The aortic arch was accessed via median sternotomy while under cardiopulmonary bypass and intraoperative hypothermia, and major arch vessels clamped and dissected out. A 28mm/30mm Thoraflex Frozen Elephant Trunk Device was then deployed through the true lumen of the aortic arch and its cuff sewn onto the distal arch, while a 10mm Gelweave graft was sewn onto the axillary artery. Proximal anastomosis was performed, with the branches anastomosed respectively to the left subclavian artery, left common carotid and the innominate artery. The patient was admitted to Intensive Care for post-operative management.

Post-operative CT aortography confirmed graft patency with no significant complications (Figure 5). His hoarseness persisted, and a repeat nasopharyngoscopy revealed persistent left vocal cord palsy. A videofluoroscopic swallow study revealed mild oropharyngeal dysphagia characterized by delayed swallow triggers and reduced epiglottic retroflexion. The patient was initially planned for inpatient rehabilitation but recovered well and was discharged home on post-operative day 27. His back pain resolved. As his hoarseness persisted, he subsequently underwent dermal filler injections for vocal cord medialization. He is otherwise well and back to premorbid function as of time of writing.

DISCUSSION

Etiology & Demographics:

Cardiovocal syndrome, also known as Ortner's syndrome, involves hoarseness due to compression of the left recurrent laryngeal nerve. This compression commonly occurs in the region where the left recurrent laryngeal nerve loops around the inferior aspect of the aortic arch, where it is closely related to the pulmonary artery and left atrium (Figure 6). A literature review revealed five cases documented as aortic dissections presenting with dysarthria, all of which were painless [5-9]. The youngest patient was a 58-year-old woman, and the rest were male, with ages ranging from 68 to 81. Three had Stanford Type A dissections and the remaining two were Stanford Type B aortic dissections. Duration of dysarthria ranged from two days to six months prior to presentation. However, all five cases involved compression of the left recurrent laryngeal nerve via either a concomitant aortic aneurysm [5-6] or an intramural aortic hematoma secondary to aortic dissection [7-9], and not an isolated aortic dissection per se. One should keep in mind that both pathologies, although commonly associated, are not always seen with aortic dissection. In this particular patient, it was likely the large saccular arch aneurysm, rather than the aortic dissection, that led to his vocal cord palsy.

Only thoracic aortic aneurysms (TAA), as opposed to the more common abdominal aortic aneurysms (AAA), are located in the mediastinum and will therefore be able to compress the left recurrent laryngeal nerve, resulting in cardiovocal syndrome. The prevalence of TAAs range from 0.16-0.34% [10-11], although actual values are difficult to ascertain as they are usually slowly expanding and therefore often asymptomatic and insidious [12]. In a retrospective study of 62 thoracic artery aneurysms, only one case (1.6%) presented with dysarthria [13]. Risk factors can be congenital or acquired: congenital risk factors include collagen disorders (e.g. Marfan syndrome, Ehlers-Danlos syndrome), bicuspid aortic valves and Turner syndrome, and acquired risk factors include hypertension, inflammatory large vessel vasculitides (e.g. Takayasu arteritis, syphilitic aortitis) and aortic surgery or instrumentation. Management can be either conservative, medical or surgical, with surgical being further subdivided into open and endovascular approaches [12].

Clinical & Imaging Findings:

Cardiovocal syndrome clinically presents with hoarseness. Other possible symptoms include dysphagia, cough, and shortness of breath [14] In this particular case, contrast enhanced CT imaging revealed anteromedial deviation of the arytenoid cartilage and dilatation of the left laryngeal ventricle, consistent with left vocal cord paralysis. Other radiological findings of vocal cord paralysis include atrophy of the posterior cricoarytenoid muscle and laryngeal edema [15]. Further imaging such as cardiac MRI can also be utilized to identify the underlying etiology (Table 2).

Treatment & Prognosis:

Subsequent management and outcomes of the five cases varied widely, with three patients either refusing repair or deemed not medically fit for surgery, and two eventually undergoing surgical repair. One was a 58-year-old female patient with hypertension who had a Stanford Type A aortic dissection and intramural aortic hematoma presenting with dysarthria for two days [7]. She underwent an aortic root replacement with good resolution of dysarthria two weeks post-operation. The other patient who underwent surgical repair was a 68-year old male with hypertension and chronic kidney disease who had a Stanford Type B aortic dissection with concomitant aortic aneurysm presenting with dysarthria of two months [5]. He underwent an elective aortic arch replacement and aorto-axillary bypass, but eventually succumbed to pneumonia and died from multi-organ failure a few weeks later.

The prognosis of recurrent laryngeal nerve paralysis is dependent on the degree and duration of compression, and usually persists after treatment of the aneurysm [16-17]. The patient who experienced resolution of her dysphagia underwent surgical repair two days after symptoms appeared, while our patient presented with one month of symptoms and only underwent elective repair three months later. The persistence of his dysphagia may be related to the duration of nerve compression.

Differential Diagnoses:

The human voice is modulated by vocal folds located within the larynx, which are controlled by phonatory muscles. These are in turn innervated by the recurrent laryngeal nerve, which adopts a prolonged course through the mediastinum and tracheoesophageal groove. Hoarseness can therefore be caused by pathology of any of the above locations, ranging from inflammatory causes such as laryngitis and caustic agents, neoplastic causes such as nasopharyngeal carcinoma, and neuromuscular disorders such as myasthenia gravis. Other pathologies causing cardiovocal syndrome specifically, include mitral stenosis, pulmonary arterial hypertension and aortotracheal fistulas, and are further discussed below. The varied list of differentials above warrant specialized targeted investigations such as nasopharyngoscopy and radiological imaging.

MITRAL STENOSIS

Cardiovocal syndrome was classically described in a patient with hoarseness caused by the impingement of the left recurrent laryngeal nerve between the aortic arch and an enlarged left atrium due to severe mitral stenosis. X-ray findings include left atrial enlargement, signs of pulmonary hypertension such as upper zone venous congestion and pulmonary edema. Further details regarding size and severity can be obtained through a CT or Cardiac MRI.

PULMONARY ARTERIAL HYPERTENSION

The nerve palsy in pulmonary arterial hypertension is more likely to be due to direct pressure of enlarged pulmonary arteries on the thoracic part of the nerve [1]. X-ray findings include enlarged pulmonary arteries, prominent pulmonary outflow tract, and signs of elevated right heart pressures including an enlarged right atrium and elevated cardiac apex. CT signs can be divided into extracardiac vascular signs, cardiac signs, and parenchymal signs. Extracardiac vascular signs include enlarged pulmonary arteries, increased pulmonary artery to aorta ratio, and an enlarged pulmonary trunk. Cardiac signs include right ventricular wall thickness >4mm and straightening of the interventricular septum. Parenchymal signs include ground-glass nodules and neovascularity. CT may also help to identify the etiology of pulmonary hypertension e.g. interstitial lung disease.

AORTOTRACHEAL FISTULA

A CT Thorax may reveal signs such as ectopic gas surrounding or within the aorta, vascular contrast within the trachea, an aneurysm at the site of the fistula or even visualization of a continuous tract between the aorta and trachea. A Cardiac MRI can also be utilized to further delineate the fistula.

TEACHING POINT

Potentially life-threatening cardiac and vascular pathologies such as aortic aneurysms warrant early imaging and detection and may occasionally present with seemingly unrelated and benign symptoms and signs such as hoarseness. In the approach of an uncommon presentation, it is prudent to revisit fundamental principles of anatomy and physiology such as the course of the recurrent laryngeal nerve in order to recommend appropriate investigations.

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FIGURES

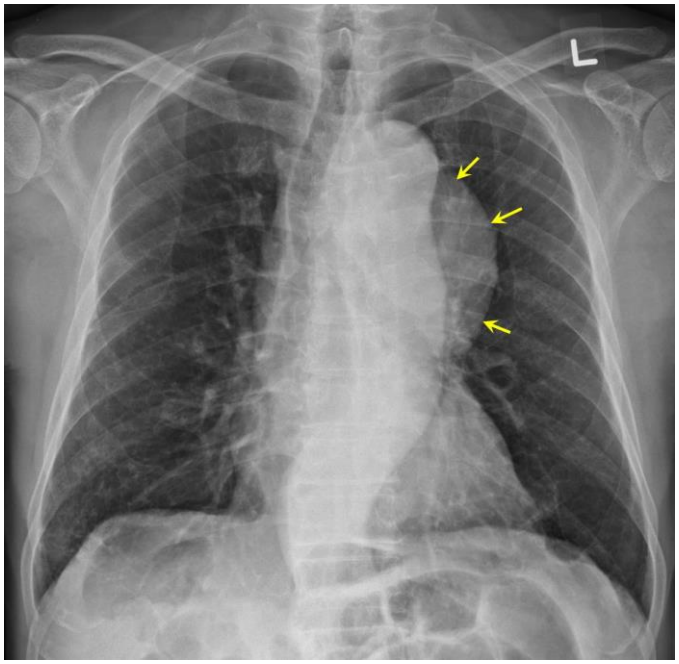

Figure 1: 81-year-old Chinese male with Cardiovocal Syndrome.
Frontal chest radiograph revealed widening of the mediastinal silhouette. There is an abnormal soft tissue density mass projected over the aortopulmonary window (yellow arrows). The margins of the descending thoracic aorta are preserved and the mass appears separate from the descending thoracic aorta.

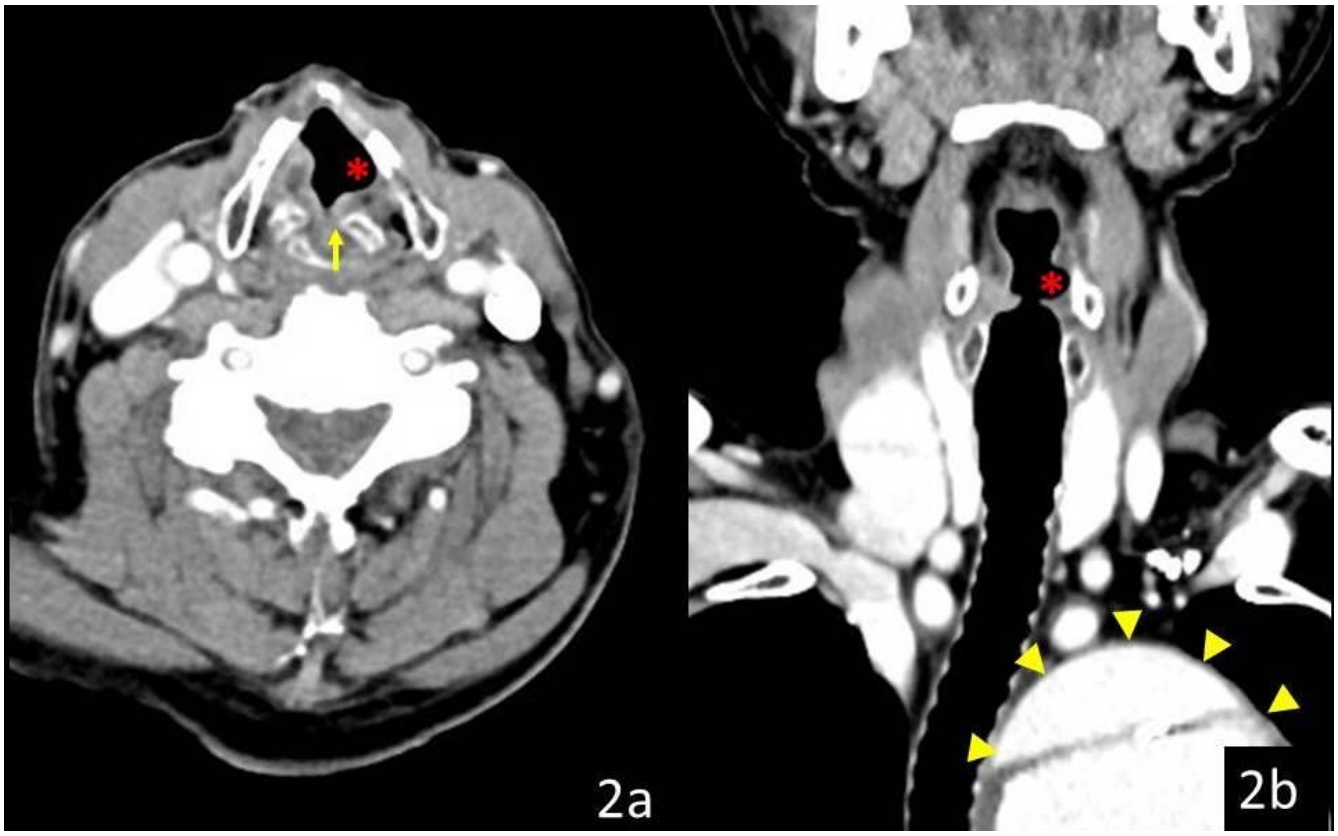

Figure 2: 81-year-old Chinese male with Cardiovocal Syndrome.
Findings: Axial (figure 2a) and coronal (figure 2b) contrast-enhanced CT neck images in a patient with left vocal cord paralysis shows enlargement of the left laryngeal ventricle (red asterisk) and anteromedial deviation of the arytenoid cartilage (yellow arrow). Figure 2b also shows a partially imaged aortic aneurysm (yellow arrow heads), deviating the trachea to the right.

Technique: Axial CT with coronal reconstructions, 90 kV, 117 mAs, 3 mm slice thickness, intravenous contrast 90 ml Omnipaque 350

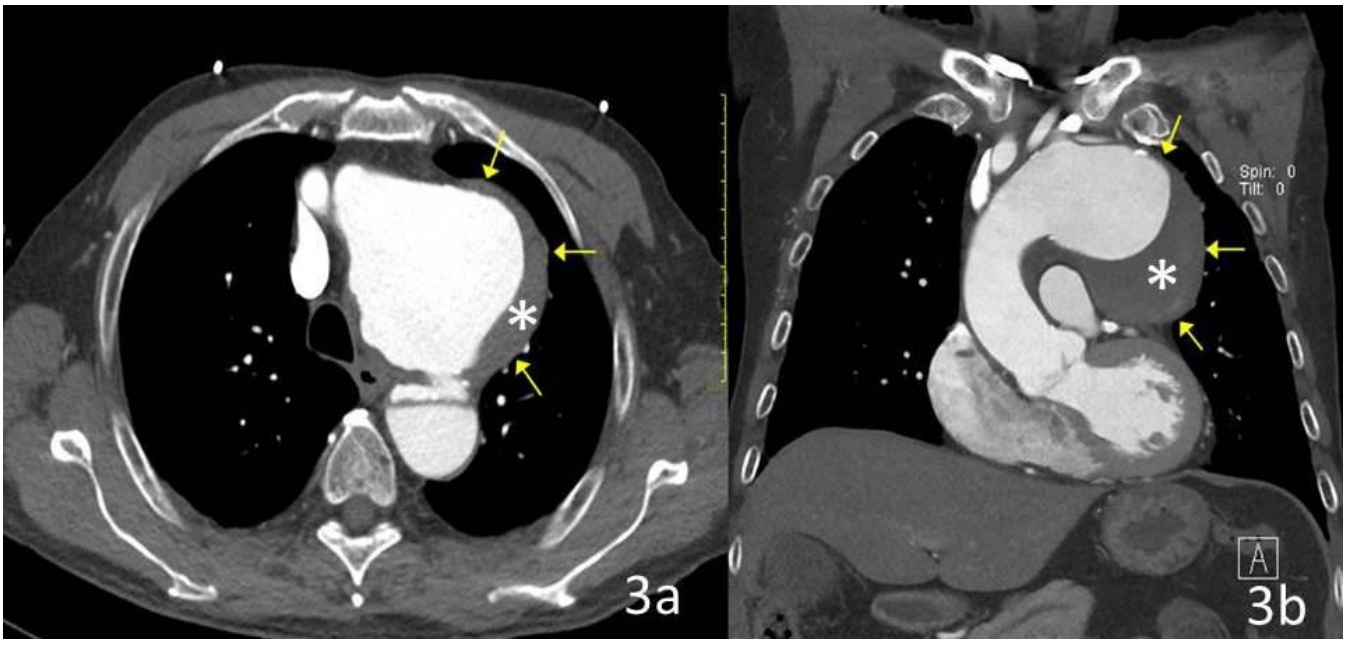

Figure 3: 81-year-old Chinese male with Cardiovocal Syndrome.
Findings: Axial (figure 3a) and coronal (figure 3b) contrast-enhanced CT aortogram reveal a large (about 9 cm diameter) saccular aneurysm of the aortic arch (yellow arrows) with mural thrombus (white asterisk).

Technique: Axial CT with coronal reconstructions, 80 kV, 197 mAs, 3 mm slice thickness, intravenous contrast 90 ml Omnipaque 350

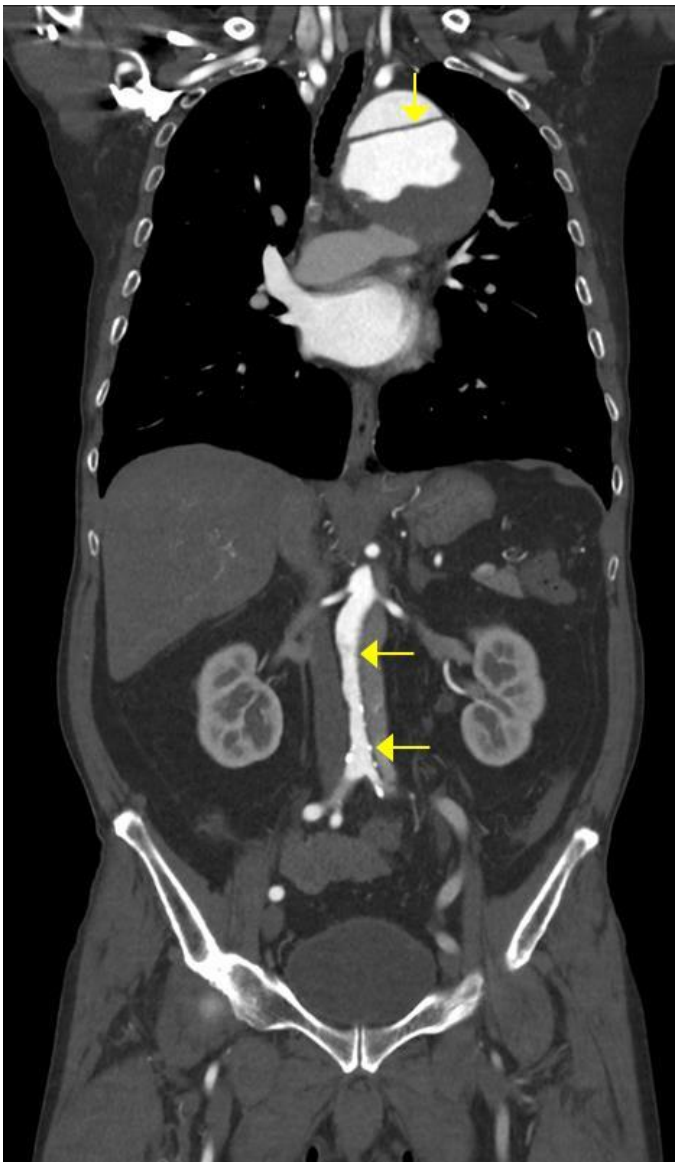

Figure 4: 81-year-old Chinese male with Cardiovocal Syndrome.

Findings: Coronal contrast-enhanced CT aortogram images showing a dissection flap (yellow arrows) extending from the distal aortic arch down to the level of the aortic bifurcation.

Technique: Axial CT with coronal reconstructions, 80 kV, 197 mAs, 3 mm slice thickness, intravenous contrast 90 ml Omnipaque 350

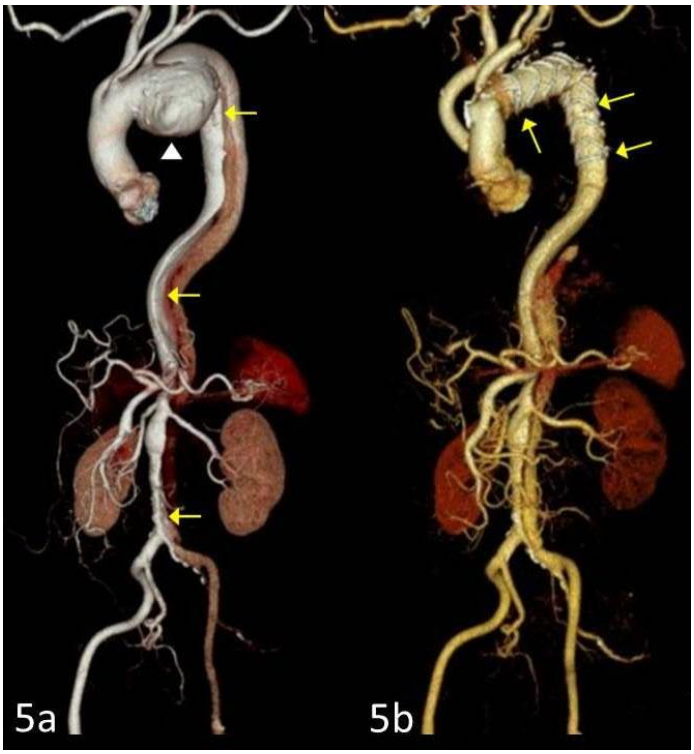

Figure 5: 81-year-old Chinese male with Cardiovocal Syndrome.

Findings: Surface shaded display volume rendered CT images of the aorta before (figure 5a) and after (figure 5b) endovascular repair. Figure 5a shows the saccular aortic arch aneurysm (white arrow head) and associated dissection flap (yellow arrows) down to the level of the aortic bifurcation (Stanford type B). Figure 5b (after aortic stent grafting) shows that the saccular aneurysm no longer fills with contrast. The location of the stent graft is indicated by the yellow arrows.

Technique: Surface shaded volume rendering of a CT, 80 kV, 197 mAs, 3 mm slice thickness, intravenous contrast 90 ml Omnipaque 350

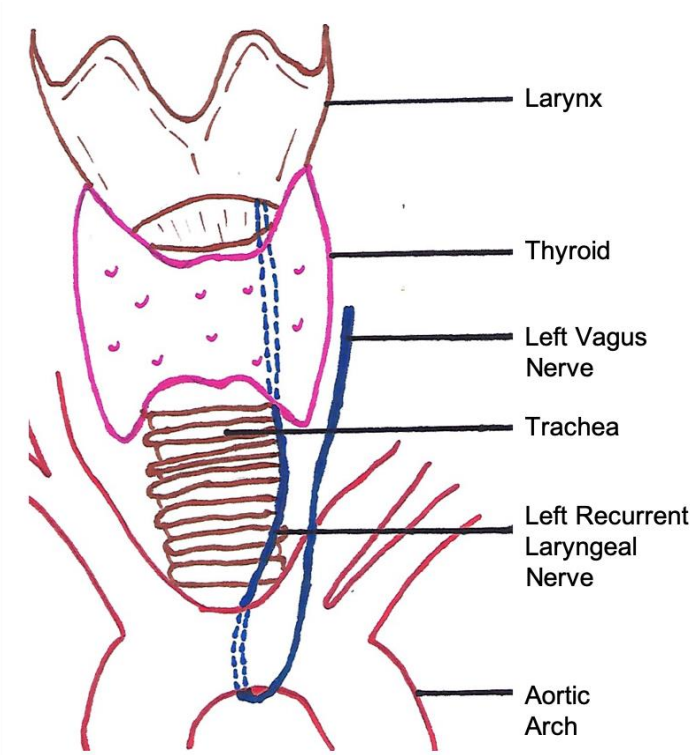

Figure 6 (left): Diagram illustrating the course of the left recurrent laryngeal nerve.
The left recurrent laryngeal nerve arises from the left vagus nerve at the level of the aortic arch. It then loops around the inferior aspect of the aortic arch before ascending on its medial surface to enter the tracheoesophageal groove. The nerve then continues superiorly to pass behind the thyroid gland and terminate in the larynx.

Etiology	Impingement of the left recurrent laryngeal nerve by a thoracic arch aneurysm
Incidence	Incidence of TAAs is 0.16-0.34%, out of which only 1.6% present with cardiovocal syndrome
Gender Ratio	Not available
Age Predilection	Not available, but likely to be more common in the elderly population.
Risk Factors	Risk factors for TAAs are as follows: *Congenital:* Collagen disorders (e.g. Marfan syndrome, Ehlers-Danlos syndrome), bicuspid aortic valve, Turner syndrome *Acquired:* Hypertension, inflammatory large vessel vasculitides (e.g. Takayasu arteritis, syphilitic aortitis), aortic surgery/instrumentation
Treatment	Treatment of cardiovocal syndrome includes management of the patient's symptoms. i.e. hoarseness, and elimination of the etiology. For symptom management, dermal filler injections can be utilized for vocal cord medialization, with an early referral to speech therapy for voice rehabilitation. Addressing the etiology in this case would involve treatment of his aortic aneurysm either via endovascular, surgical or combined approaches.
Prognosis	The prognosis of recurrent laryngeal nerve paralysis is dependent on the degree and duration of compression, and usually persists after treatment of the aneurysm.
Findings on Imaging	*Videofluoroscopic swallow study:* Oropharyngeal dysphagia with delayed swallow triggers and reduced epiglottic retroflexion *Chest X-ray:* Widening of the mediastinal silhouette *Contrast enhanced CT neck and CT aortogram:* Anteromedial deviation of the arytenoid cartilage and dilation of the left laryngeal ventricle indicative of left vocal cord paralysis. The etiology is also often revealed, in this case demonstrating a large saccular aneurysm with mural thrombus and dissection flap beginning distal to the left subclavian artery and extending down to the level of the aortic bifurcation.

Table 1: Summary table for cardiovocal syndrome secondary to thoracic aortic aneurysm.

Differential diagnoses	X-ray	CT	MRI
Aortic Aneurysm	May demonstrate widening of the mediastinum, irregular aortic contour	Modality of choice. Accurately depicts the site and extent of the aortic aneurysm, and any associated dissection. The location of the aneurysm and proximity to the expected course of the left recurrent laryngeal nerve will help confirm the diagnosis.	Good alternative for patients in whom iodinated contrast (used in CT) is contra-indicated, or when radiation concerns exist (e.g. young or pregnant patients).
Mitral stenosis	Non-specific, but may reveal left atrial enlargement, signs of pulmonary hypertension such as upper zone venous congestion and pulmonary edema	Provides clear anatomical delineation of the size and extent of left atrial enlargement. If ECG gated CT obtained, there is also an option for cardiac and valve function to be assessed.	Cardiac MRI can provide further detail for both diagnostic and surgical planning purposes. Severity can be gauged via mitral valve area and pressure.
Pulmonary arterial hypertension	Enlarged pulmonary arteries, prominent pulmonary outflow tract, and signs of elevated right heart pressures including an enlarged right atrium and elevated cardiac apex.	Signs can be divided into extracardiac vascular signs, cardiac signs, and parenchymal signs. Extracardiac vascular signs include enlarged pulmonary arteries, increased pulmonary artery to aorta ratio, and an enlarged pulmonary trunk. Cardiac signs include right ventricular wall thickness >4mm and straightening of the interventricular septum. Parenchymal signs include ground-glass nodules and neovascularity. CT may also help to identify the etiology of pulmonary hypertension e.g. interstitial lung disease.	Cardiac MRI can assist in the assessment of the right ventricle and proximal pulmonary arteries. Shunts, ventricular hypertrophy and impairment, and wall motion abnormalities can be visualized in greater detail.
Aortotracheal fistula	No specific findings to allow diagnosis of this condition.	May reveal signs such as ectopic gas surrounding or within the aorta, vascular contrast within the trachea, an aneurysm at the site of the fistula or even visualization of a continuous tract between the aorta and trachea.	May be utilized to further delineate the fistula.
Laryngitis	May present with tracheal wall thickening, although most likely to appear normal.	May present with tracheal wall thickening. May also reveal signs of distal involvement such as subsegmental atelectasis, peribronchial thickening, and perihilar consolidations.	No further specific findings to allow diagnosis of this condition.
Nasopharyngeal Carcinoma	Not a commonly utilized modality to allow diagnosis of this condition.	Soft tissue masses may be noted, most commonly at the fossa of Rosenmüller. Can also be utilized to delineate the extent of growth, and to assess for early bone and lymphatic involvement.	Can be used to assess for early perineural spread.

Table 2: Differential diagnosis table for cardiovocal syndrome.

ABBREVIATIONS

2D = 2-Dimensional
AAA = Abdominal Aortic Aneurysms
CT = Computed Tomography
TAA = Thoracic Aortic Aneurysms

KEYWORDS

Cardiovocal syndrome; Ortner syndrome; Aortic dissection; Hoarseness; CT Aortogram

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