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Case Report

Horseshoe adrenal gland associated with retro-aortic right diaphragmatic crus and several vascular variants: Report of two cases ^{☆,☆☆}

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ABSTRACT

Horseshoe adrenal gland (HA) refers to congenital fusion of the adrenal glands across the midline - a rare anatomical variant often found in association with other congenital anomalies. Here we report 2 cases of HA associated with retro-aortic course of the right diaphragmatic crus, yet another rare anatomical variant, in a 61 year-old male and a 69-year-old female, who underwent CT examinations for unrelated reasons. Both patients also had additional vascular and vertebral anomalies. To the best of our knowledge, this is the first report to document association of 2 rare congenital anomalies: HA and retro-aortic right diaphragmatic crus.

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Introduction

Horseshoe adrenal gland (HA) is a rare congenital condition in which adrenal glands are fused across the midline, connected by an isthmus posterior to the aorta. Most reported cases of HA were seen on ultrasound or at autopsy in fetuses and infants. HA was found to be associated with other congenital anomalies, including asplenia (right atrial isomerism), neural tube defects and renal agenesis [1].

To our knowledge, there are a total of 7 published cases of HA in adults, usually reported as an incidental finding on cross-sectional imaging. Of the 7 reported cases of adult HA, 3 were associated with posterior diaphragmatic defect, 2 with vascular variants and 4 with spinal variants, as summarized by Khurram et al. [2].

However, none of the reported cases of HA were associated with a retro-aortic course of the right diaphragmatic crus, yet another rare congenital anomaly. Similarly, there are no pub-

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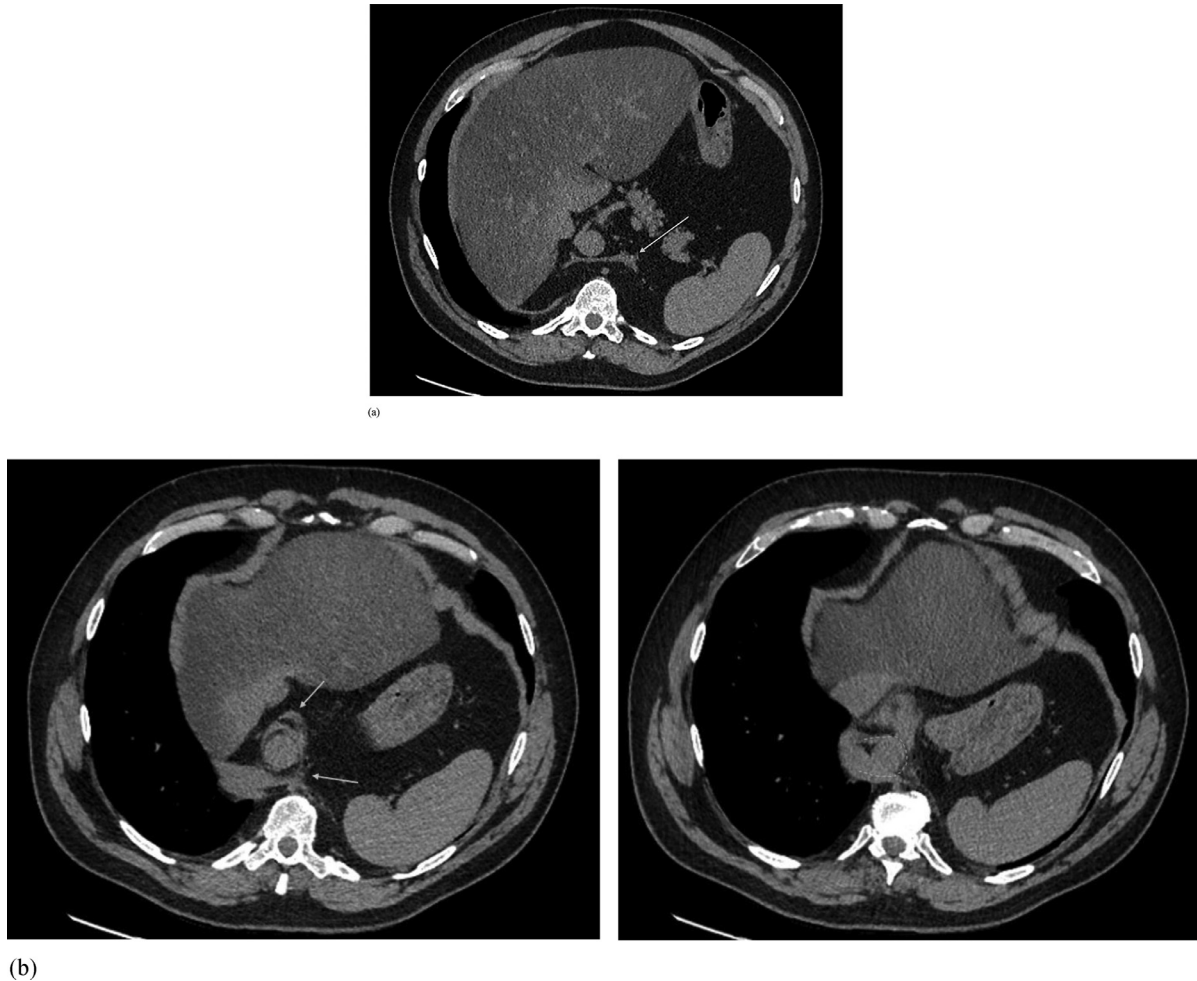


Fig. 1 – Non-contrast-enhanced CT chest of the upper abdomen demonstrating (A) fused (horseshoe) adrenal gland (arrow), and (B) retro-aortic course of the right diaphragmatic crus, forming an inverted C-shaped soft tissue density (arrows), resulting in a sling-like soft tissue structure (dotted outline) around the aorta.

lished reports of HA in association with aberrant right subclavian artery, bi-carotid trunk or absent celiac trunk.

Here we present 2 cases of HA associated with a retro-aortic right diaphragmatic crus and aberrant right subclavian artery, found incidentally on computed tomography of adult patients. Additionally, one of our cases also featured bi-carotid trunk and absent celiac trunk.

Case presentations

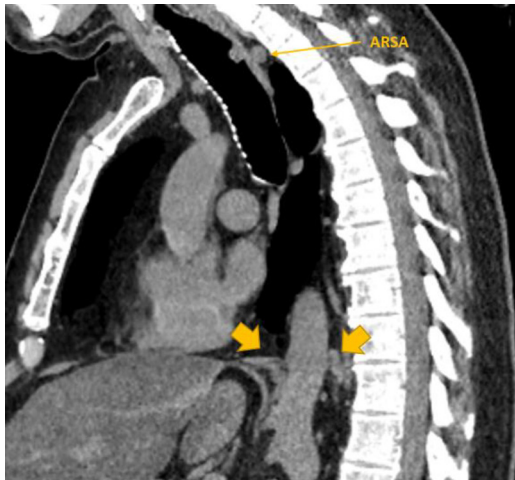
Case 1

A 61-year-old man with a 5 pack-year history of smoking, known for lumbosacral spina bifida and remote resection of a conus medularis lipoma, was referred for a CT chest to rule out interstitial lung disease. Non-contrast CT chest was performed, demonstrating no evidence of interstitial lung disease, but revealing an incidental HA (Fig. 1A), abnormal retro-

aortic course of the right diaphragmatic crus (Fig. 1B), and an aberrant right subclavian artery (Fig. 2).

Case 2

A 69-year-old woman, known for chronic obstructive pulmonary disease, hypertension and atrial fibrillation, presented to our emergency department with vertigo and pleuritic chest pain. A contrast-enhanced CT pulmonary angiogram (CTPA) was requested to rule out pulmonary embolism. The CTPA showed no evidence of pulmonary embolism and no acute intrathoracic abnormality. However, it demonstrated incidental HA (Fig. 3A) and retro-aortic right diaphragmatic crus (Fig. 3B). Additionally, the patient was found to have an aberrant right subclavian artery (Figs. 4 A-B), bi-carotid trunk (Fig 4B), and absent celiac trunk, with left gastric, splenic, common hepatic and superior mesenteric arteries arising independently from the aorta (Fig. 5). The patient was also found to have fused T10 and T11 vertebral



(a)



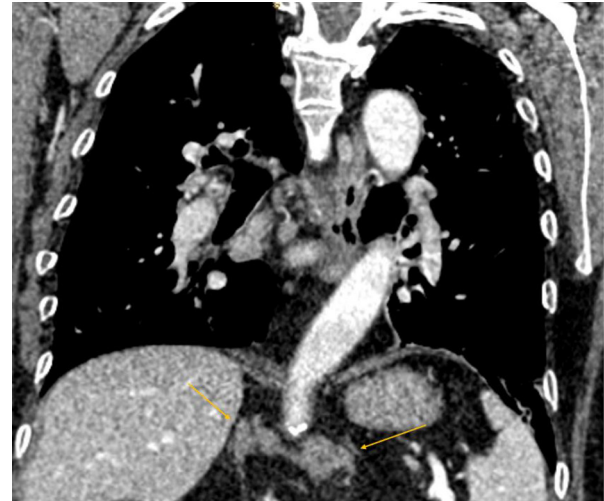
(b)

Fig. 2 – Non-contrast-enhanced CT of the chest. (A) Sagittal reformation demonstrating retro-esophageal course of the aberrant right subclavian artery (thin arrow). Broad arrows are pointing to the right diaphragmatic crus, which is both anterior and posterior to the aorta. **(B)** Axial image at the aortic inlet demonstrating branches of the aortic arch, including retro-esophageal course of the aberrant right subclavian artery. RCA: right common carotid artery, LCA: left common carotid artery, LSA: left subclavian artery, ARSA: aberrant right subclavian artery.

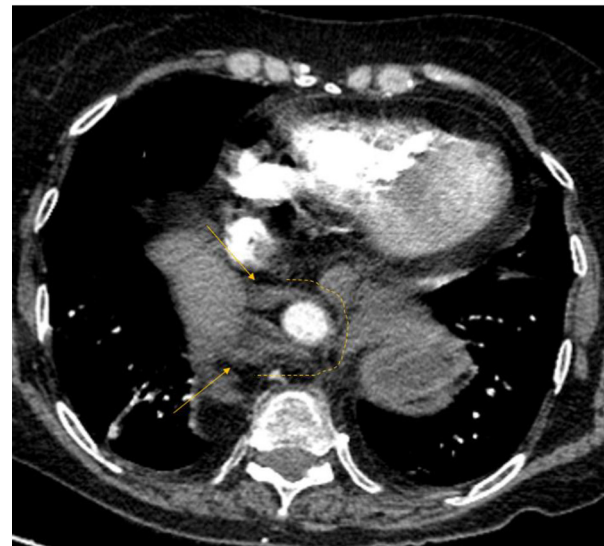
bodies (Fig. 4A). The patient was managed conservatively for her symptoms and subsequently discharged.

Discussion

Here we present 2 cases of HA associated with a retro-aortic right diaphragmatic crus and aberrant right subclavian artery, found incidentally on computed tomography of adult patients.



(a)



(b)

Fig. 3 – Contrast-enhanced CT of the chest. (A) Coronal reformation showing midline fusion of the adrenal glands (horseshoe adrenal). **(B)** Axial image of the lower chest demonstrating retro-aortic course of the right diaphragmatic crus, forming an inverted C-shaped soft tissue density around the aorta (line arrows), giving rise to a sling-like soft tissue structure (dotted line).

Additionally, one of our cases also featured bi-carotid trunk and absent celiac trunk.

Horseshoe or butterfly adrenal gland is the term used to describe a midline fusion of the adrenal glands. It has been reported in the adult population in association with other congenital anomalies involving genitourinary, cardiovascular, musculoskeletal, and central nervous systems [2].

Association of the HA with diaphragmatic abnormalities was first reported by Feldmann et al. in 2009, who observed medial defects of the posterior diaphragm with a “floating crura” appearance and postulated that both abnormalities probably resulted from embryological failure of midline

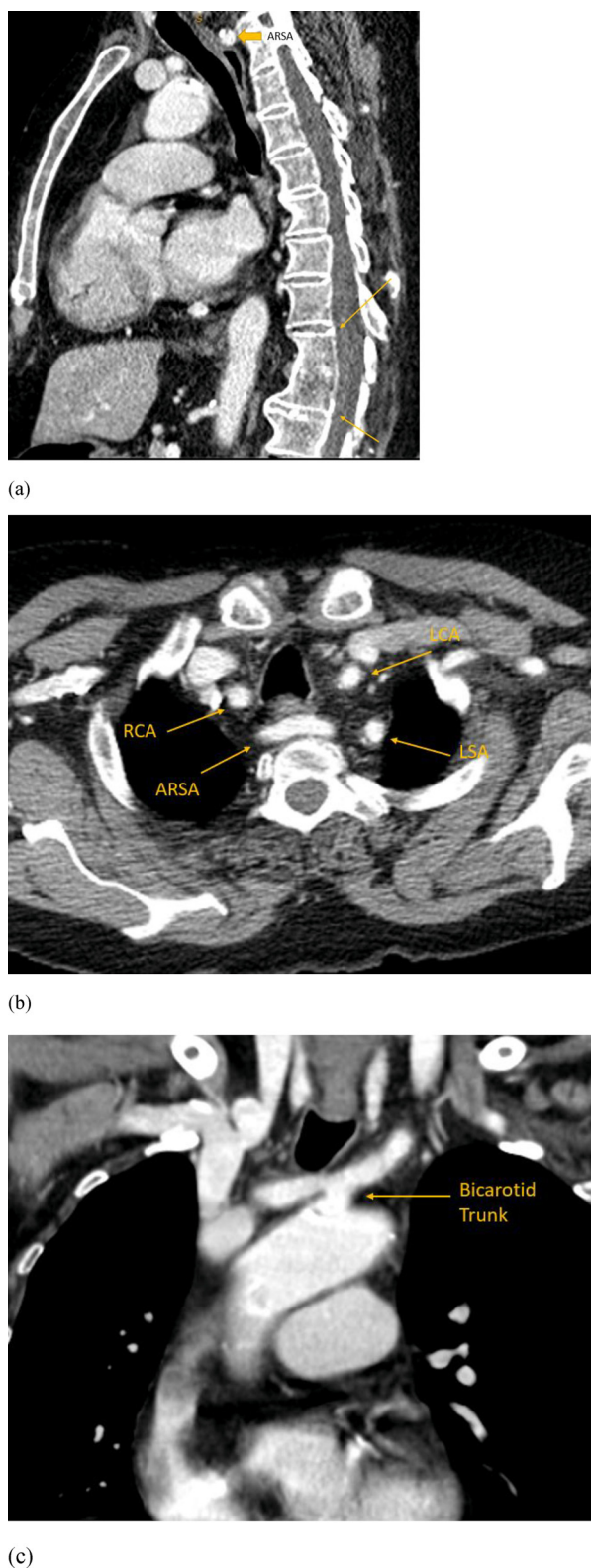


Fig. 4 – Contrast-enhanced CT of the chest. (A) Sagittal reformation demonstrating retro-esophageal course of the aberrant right subclavian artery (broad arrow), and fused T10 and T11 vertebral bodies (thin arrows). (B) Axial image at the aortic inlet demonstrating branches of the aortic arch and retro-esophageal course of the aberrant right subclavian artery. (C) Coronal reformation demonstrating the first branch of the aortic arch which bifurcates into right and left common carotid arteries.

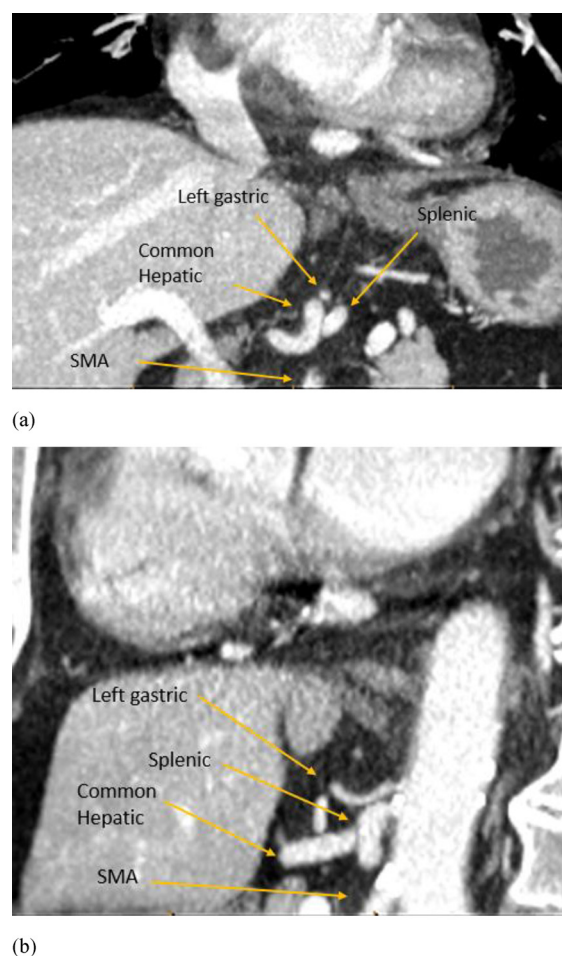


Fig. 5 – Contrast-enhanced CT of the upper abdomen. (A) Coronal and (B) saggital zoomed-in reformations showing absent celiac trunk, with independent origins of the left gastric, splenic, common hepatic, and superior mesenteric (SMA) arteries from the aorta.

structures to separate, given shared embryological precursor, anatomical proximity, and concurrent development during 4th-6th weeks of gestation of both the adrenals and diaphragm [3].

Subsequently, another group reported a case demonstrating similar findings with additional congenital vertebral abnormalities [4]. This finding provided additional support to the assumption that HA and diaphragmatic defects result from failure of separation of midline structures during early gestation. The hypothesis being that since the vertebrae start to develop during the 3rd week of gestation, before adrenal and diaphragmatic development (4th week), failure of normal vertebral development can result in a persistent notochord which is postulated to be underlying factor for subsequent failure of adrenal gland separation and failure of diaphragmatic fusion. In addition, the authors proposed a possible role of defective cellular proteins and transcription factors such as Pax and Sox to explain the etiology of these combined incidental abnormalities in healthy adults [4].

In agreement with the above hypothesis, we have observed similar spinal anomalies in both our cases of HA: our first case had

history of spina bifida and the second case - vertebral fusion at the level of the diaphragm.

Additionally, both our cases demonstrated concomitant HA and unusual posterior diaphragmatic abnormality, that is not a defect but an abnormal course of the right diaphragmatic crus, posterior to the aorta – an association, which to our knowledge, has never been previously described in English-language medical literature. Moreover, our search in PubMed and Google Scholar search engines, failed to identify a single published case of a retro-aortic diaphragmatic crus. There has been a considerable number of published cadaveric and cross-sectional imaging anatomic studies of the diaphragm focusing on variants and variable appearances of the diaphragmatic crura, however none have described this rare variant [5].

Aside from the abnormal right retro-aortic diaphragmatic crus, both our cases of HA also featured aberrant right subclavian artery (ARSA), which is the most common congenital anomaly of the aortic arch, with a prevalence of 0.5%-2% in the general population [6]. We also found a common carotid trunk (CCT) in our second case, which is seen in 20.6% of patients with aberrant right subclavian artery [7,8]. However, given relatively high prevalence of these vascular variants, both ARSA and CCT could have been coincidental.

Our second case also demonstrated absence of the celiac trunk with independent origins of left gastric, splenic, common hepatic and superior mesenteric arteries from the aorta, which is a rare variant with reported incidence of 0.1%-2% [9]. It is thought to result from complete resorption of the longitudinal anastomosis uniting the ventral segmental arteries of the aorta, without regression of the roots of the ventral segmental arteries, which is thought to take place during 4th-5th weeks of gestation [10].

Although we are unable to provide a definite explanation of the connection between HA, diaphragmatic and vascular abnormalities we observed in our 2 cases, we hypothesize that these associations are not random and are probably all related to a common mechanism; failure of normal development of posterior midline structures during fetal development.

Conclusion

In conclusion, horseshoe adrenal gland is an extremely rare entity that is getting more frequently detected and recognized with the increased use of cross-sectional imaging. We

report 2 cases of a previously unreported association of HA with unusual diaphragmatic variant of retro-aortic right diaphragmatic crus. Additionally, both our cases featured spinal anomalies (spina bifida and vertebral fusion), aberrant right subclavian artery, and, in one case, common carotid trunk and absent celiac trunk. Early embryological event leading to these abnormalities is a plausible explanation, however the precise mechanism remains unknown.

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