Case Report

Right-sided aortic arch with mirror image branching and situs solitus: a case of a 79 years old woman

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A B S T R A C T

Right aortic arch with mirror image branching (RAMI) is a rare congenital defect of the aorta. The exact incidence of RAMI in the general population is unclear. In RAMI the first branch arising from the arch is the left innominate artery, followed by the right carotid artery and right subclavian arteries. We report a case of an adult female patient with RAMI discovered as an incidental finding during radiological investigations for suspected pulmonary embolism in emergency department. No other congenital malformations were reported. It is important to recognize congenital variants of the aortic arch, as they can have relevant implications for patients' prognosis and management. Therefore, being aware of these conditions is key to avoid any mistakes or surgical and endovascular complications.

Introduction

Congenital anomalies of the aortic arch complex include left-sided, right-sided, and double aortic arches, with various branching patterns of the great vessels. They result from aberrant development of one or more components of the embryonic pharyngeal arch system [1]. According to Edwards' hypothetical embryonic double aortic arch model, each primitive aorta consists of a ventral and a dorsal segment [1]. Six paired primitive aortic arches develop between the ventral and dorsal aortae. It is possible to describe most anomalies of the aortic arch by postulating regression of a segment that would normally persist, and/or persistence of a segment that would normally regress [2]. In accordance with this, the Edwards classification describes 3 types of right aortic arch (RAA): RAA with mirror image branching (RAMI), RAA with aberrant left subclavian artery (ALSA) and RAA with isolation of the LSA [3]. RAMI

is the second most common form of a right-sided arch, after right arch with ALSA. This anomaly results from regression of the left dorsal aorta distal to the origin of the seventh intersegmental artery, so that the left fourth arch becomes the proximal subclavian artery rather than the definitive aortic arch [4]. We report a case of a 79 years old woman with right aortic arch with mirror image branching (RAMI) discovered as incidental finding.

Case report

A 79-years-old-female was admitted to the Emergency Department for dyspnea and chest pain.

She had no history of cardiovascular disease. In medical history, the patient referred to be affected by left renal oncocytoma, not subjected to any type of therapy, monitored, and stable in the various controls.

Blood laboratory test showed: creatinine: 0.80 mg/dL (normal value (nv): 0.55-1.22 mg/dL); fibrinogen 365.00 mg/dL (nv 200-400 mg/dL); D-Dimer: 3243.00 ng/mL (nv 0-500 ng/mL); international normalized ratio: 0.97 (0.8-1.2); creatinin kinase: 0.90 mg/mL (nv <3.4); myoglobin: 38.00 ng/mL (nv <108); troponin 6.0 ng/mL (nv <15.6).

Pulmonary embolism was suspect and a contrast-enhanced computed tomography (CT) chest examination was performed by a 64-slice CT scanner (Lightspeed; General Electric Healthcare, Waukesha, WI). The protocol included a noncontrast CT scan, with 5 mm of thickness, and dynamic
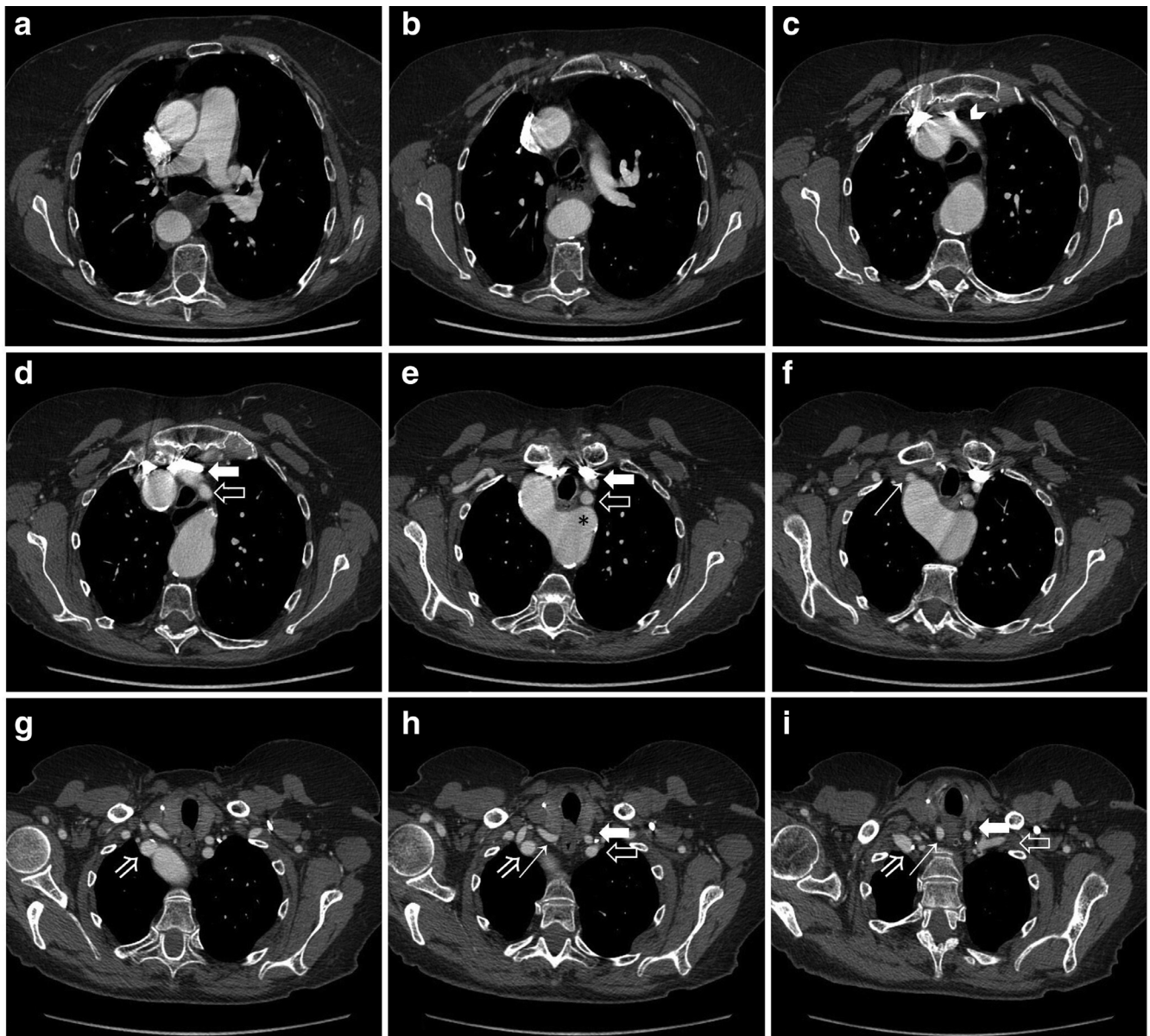
Fig. 1 – Nine axial computed tomography sections listed in craniocaudal direction show epiaortic vessels origin and development: (c) show origin of left innominate artery (white arrowhead); (d) show left carotid artery (white arrow) and left subclavian artery (white outline arrow); (e) show focal ectasia on the anterior arch aortic wall (*); (f) show origin of right carotid artery (north east arrow), (g) show origin of right subclavian artery (double lines arrow).

Fig. 2 – Figures show descending thoracic aorta development and dilatation of the lower esophagus with evidence of swallowed food stasis (*).
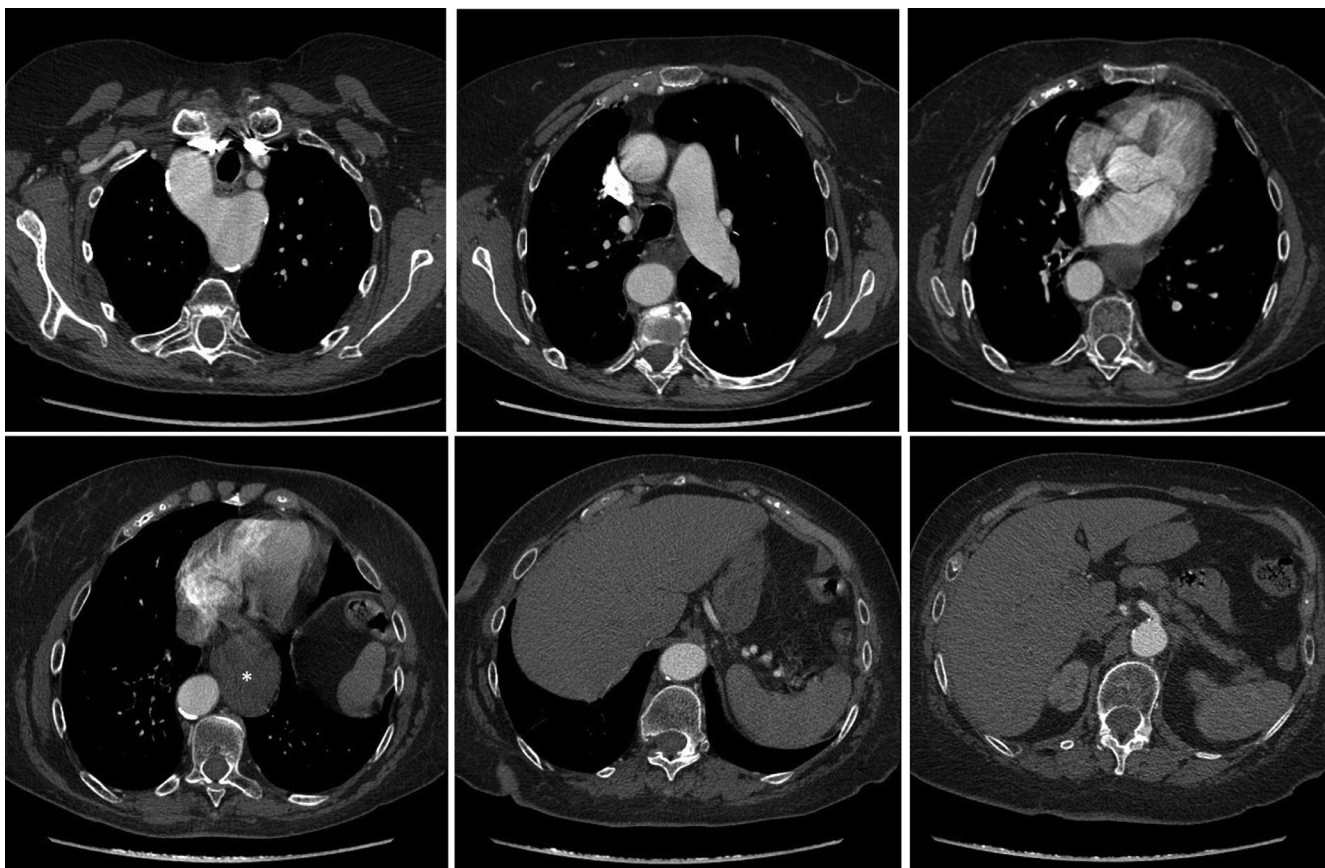
acquisition with bolus tracking after administration of iodine contrast medium (Iobitridolo; volume: 70 mL; flow rate: 3.5 mL/s), slice thickness: 1.25 mm. CT exam was negative for pulmonary embolism. There were evident submantellar emphysema blisters as for parasectal emphysema and massive sliding hiatal hernia with dilatation of the lower esophagus and swallowed food stasis.

As additional finding, contrastographic CT acquisition showed a curvy right-sided thoracic aorta with anomalous RAA and origin of epiaortic vessels from it. Aorta originates from the left ventricle, ascends on the right of trachea and esophagus and then descends posteriorly to the esophagus, after showing a focal ectasia on the anterior aortic wall, maybe because of an embryonal residual (Fig. 1). Descending thoracic aorta goes down right-sided to the rachis, and then, at its distal third, with a curvy path, leads on the left and descend with its usual development (Fig. 2).

The first branch arising from the left lateral-anterior wall of the arch is the left innominate artery, which, after a brief course, splits into the left common carotid and LSA that move left where they proceed with their usual development.

The second vessel is the right common carotid artery that originates from the anterior wall, presents kinking at the proximal third where it shows a paraesophageal course and then goes up into the neurovascular bundle of the neck.

The third vessel originating from the right lateral-anterior wall of the arch is the right subclavian arteries which, with horizontal path, lead into the axillary region (Figs. 1,3, 4-6).

This congenital defect is known as right aortic arch with mirror image branching (RAMI). No other malformations were reported. The patient was informed of this condition.

Discussion

Congenital anomalies of the aortic arch can occur with a frequency ranging from 0.5% to 3.0% [5]. Right-sided aortic arch (RAA) is a rare congenital defect of the aorta. It is present in 0.05% to 0.1% of adult radiology series and in 0.04%-0.1% of autopsy series [5,6]. In literature, the exact incidence of RAMI in the general population is unclear because usually they remain undetected. According to Hayashi et al., RAMI cases detected in adult CT examinations represented 0.012-0.018% [7]. In pediatric age, instead, there are not several studies reporting RAMI incidence. In their work, Oztunc et al. reviewed 653 foetal and neonatal echocardiographic examinations performed in 8 years and founded only 15 cases of RAMI [8]. In RAMI the left innominate artery is the first branch arising from the arch, which is followed by the right carotid artery and right subclavian arteries.
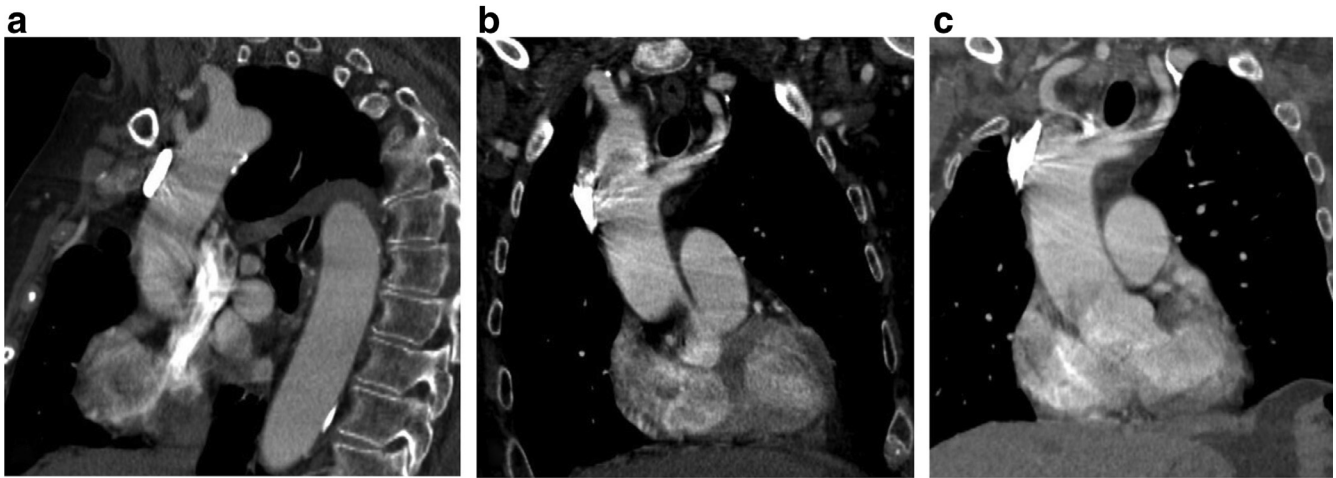
Fig. 3 – (a) shows origin of right carotid common artery (RCCA) and right subclavian artery (RSA) on sagittal section; (b) shows origin of right subclavian artery (RSA) and left innominate artery (LIA) on coronal section; (c) shows origin of right carotid common artery (RCCA) and left innominate artery (LIA) on coronal section.
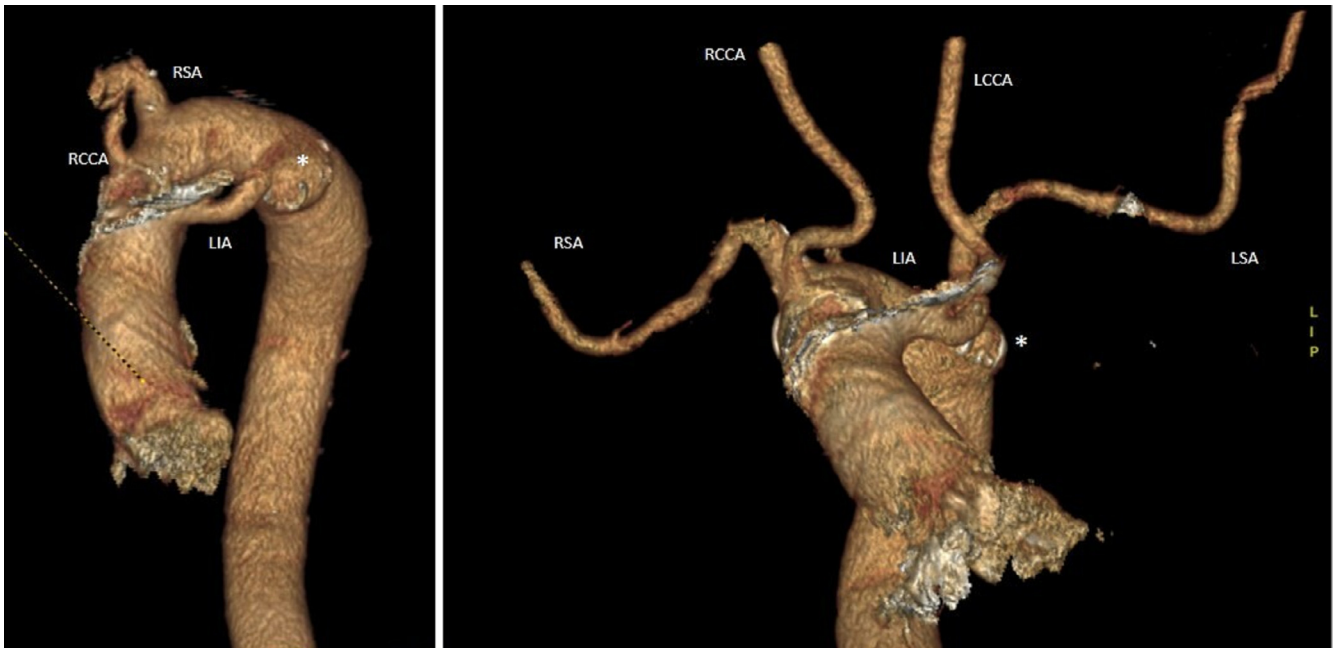

Fig. 4 – Three-dimensional reconstruction of our case of RAMI where is shown epiaortic vessels origin and focal ectasia on the anterior arch aortic wall (*).

This anomaly rarely produces symptoms and is usually an incidental radiological finding.

In adulthood, symptoms associated with this type of RAA are often the result of early atherosclerotic changes of anomalous vessels or aneurysmal dilatation accompanied by compression of surrounding structures such as the trachea or the esophagus [9,10]. It can be associated with dysphagia lusoria or respiratory distress. The development of aneurysm usually occurs at the level of origin of an ALSA and is known as Kommerell's aneurysm [11]. In this situation, long-term Doppler ultrasound monitoring may well be a valuable diagnostic method.

In infancy, symptoms of RAMI are related to cyanotic congenital heart diseases (CHD). RAMI is strongly associated with CHD in greater than 98% of cases, especially tetralogy of Fallot and truncus arteriosus [12]. According to Tawfik et al., 18% of children with CHD present a RAA [13]. Evans et al. hypothesized that a right aortic arch in situs solitus, with or without an associated cardiovascular malformation, is often associated with a vascular ring [14]. In our patient, no congenital heart anomalies were found.

Despite RAMI without CHD is considered a benign condition by most radiologists, adults with RAMI may have increased risk of acquired aortic disease and should be moni-
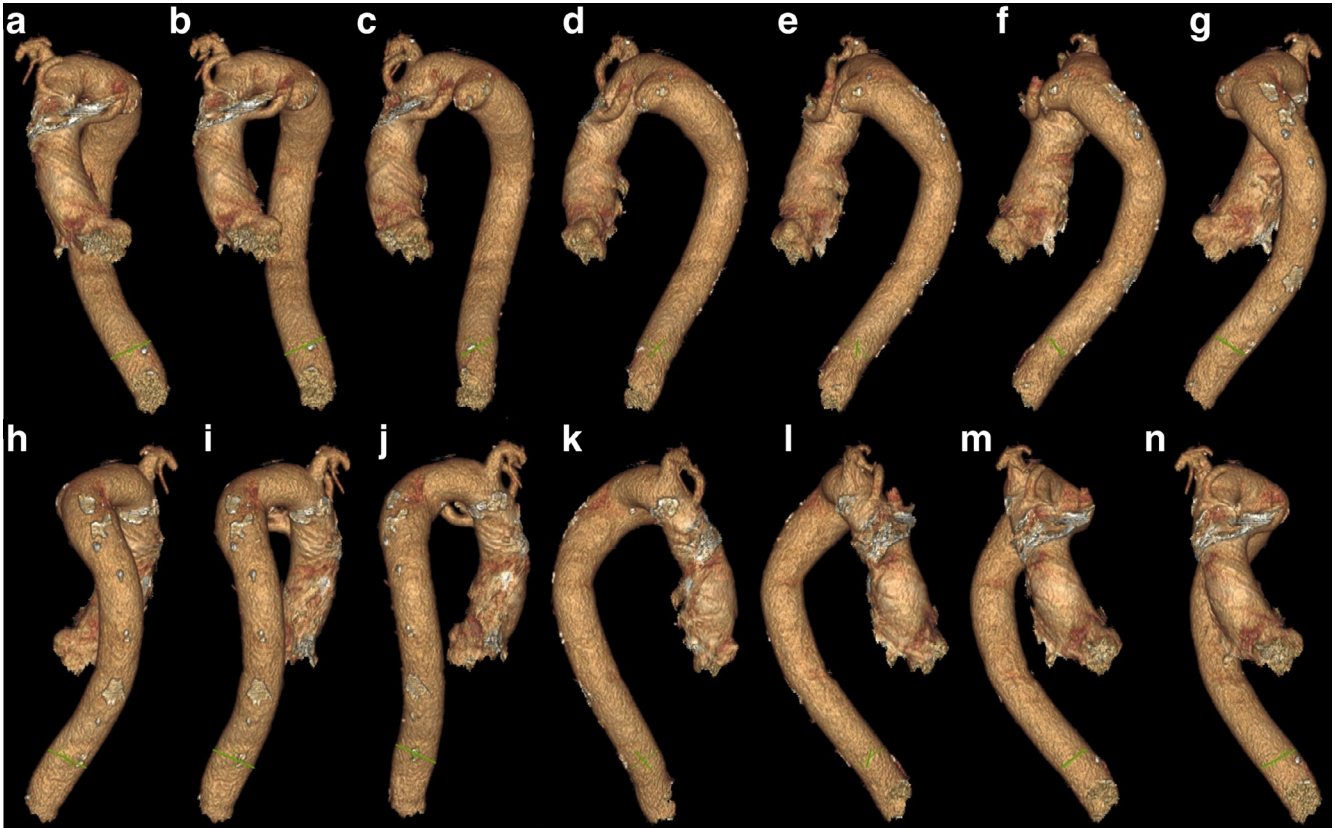
Fig. 5 – Figure shows several views of a 3-dimensional reconstruction of the thoracic aorta.

Fig. 6 – Computed tomography scout view shows upper mediastinal contours anomalies.

tored carefully. Echocardiography, cardiac magnetic resonance (MR) imaging, and computed tomographic (CT) angiography are important imaging modalities used to identify and diagnose aortic arch variants. Moreover, knowledge of the associations' specific to a particular aortic arch anomaly is important in directing further detailed search for other associated defects and anomalies. Identification of these malformations allows for accurate surgical and intervention planning and helps avoid potential complications [15]. Detailed evaluation of aortic arch anatomy is important for planning thoracic surgery and endovascular intervention, as the presence and pattern of arch variants and anomalies may influence the surgical incision, cardiopulmonary bypass cannulation, and/or interventional approach. Arch anomalies may increase the technical difficulty of endovascular treatment, including carotid stent procedures, and the risk of neurologic complications [16–18].

In conclusion, familiarity with the spectrum and imaging appearance of aortic arch anomalies and malformations is essential for accurate diagnosis and classification. It is important to recognize congenital anomalies of the aortic arch since they can have relevant implications for prognosis and management [19]. Awareness of these variations is significant for clinician, anesthesiologist, and surgeons to avoid any mistakes in long-term management of patients or intraoperative surgical complications.

Declaration of Competing Interest

The authors have declared that no competing interests exist.

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