CASE REPORT

Separate origins of the left internal and external carotid arteries from the aortic arch and cervical internal carotid artery aneurysm in a patient with Noonan syndrome

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SUMMARY

Distinct origins of the external carotid artery and the internal carotid artery (ICA) from the aortic arch have been rarely described, and represent an aberrant development of the aortic arches during fetal life. This anatomical variation is usually discovered incidentally; infrequently, an aneurysm of the cervical ICA might accompany this rare configuration. We describe one such case in a patient with Noonan syndrome who presented with pulsatile neck mass. The diagnostic features and management of the aneurysm and a review of the literature are presented.

INTRODUCTION

Anatomic variants of the origin of the great vessels from the aortic arch are relatively common, and are typically discovered incidentally on routine imaging.[1] A separate origin of the internal carotid artery (ICA) and external carotid artery (ECA) from the aortic arch is a very unusual anatomic configuration that has been rarely reported.[2–4] We describe a patient with Noonan syndrome who presented with a large aneurysm of the left cervical ICA, along with separate origins of the left ICA and left ECA from the aortic arch. We discuss possible underlying aberrancies in the embryological development of the aortic arch that led to this rare anatomical configuration and aneurysm formation, as well as the surgical and endovascular treatment.

CASE PRESENTATION

Our patient is a 47-year-old male with Noonan syndrome who presented for evaluation of neck pain and dysphagia. Physical examination revealed a left pulsatile neck mass. Vascular imaging studies were undertaken to better characterize the lesion. CT angiogram (figure 1) demonstrated independent origins of the left ICA and ECA from the aortic arch and absence of a common carotid artery. In addition, a large aneurysmal sac was identified originating from the cervical left ICA (figure 2). Cerebral angiogram (figure 3) revealed a normal appearing left ECA whereas the cervical ICA was noted to be tortuous, dysplastic, and of smaller caliber. A saccular aneurysm originating off of the distal ICA at the level of the C2–C3 vertebrae was demonstrated. A robust ECA–ICA anastomosis between the accessory meningeal artery (arising from the internal maxillary artery) and the inferolateral trunk of the cavernous ICA at the foramen of Vesalius was contributing blood flow to the ICA terminus. The distal ICA assumed a normal anatomic course entering the carotid canal at the skull base.

Considering the size of the aneurysm and associated symptoms, treatment was undertaken. Prior to the planned operation, the patient underwent

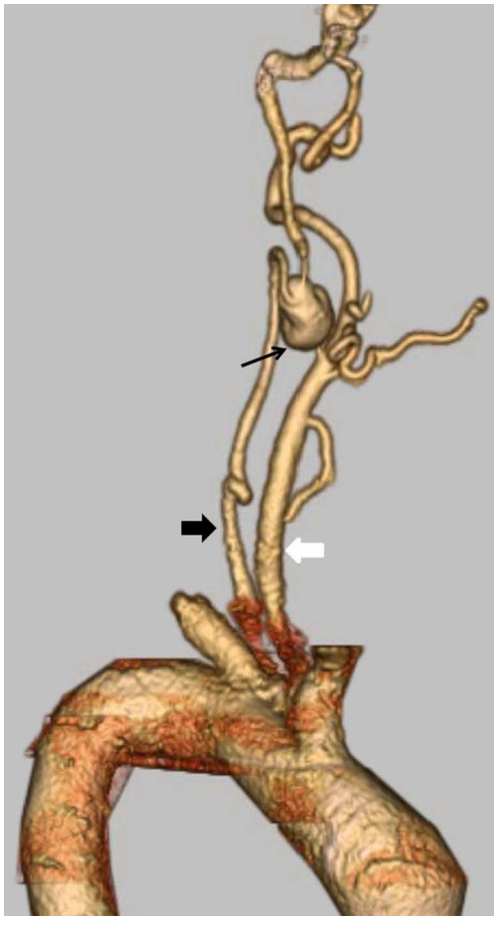

Figure 1 CT angiogram. Three-dimensional reconstruction of the aortic arch and cervical internal carotid artery (ICA) (thick black arrow) and external carotid artery (ECA) (white arrow), demonstrating clear distinct origins of the ICA and ECA from the aortic arch. Aneurysmal sac (thin black arrow) can be seen arising from the cervical ICA.

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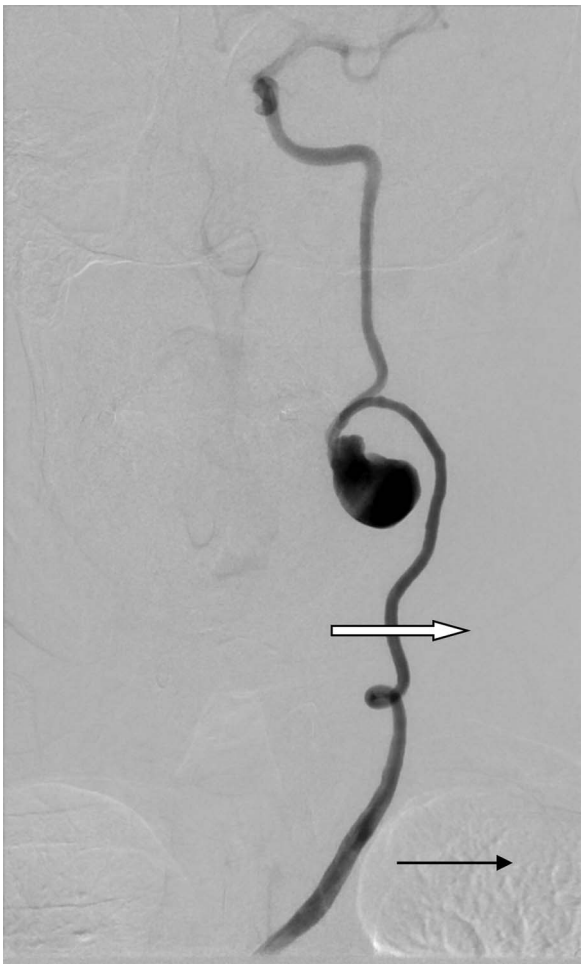

Figure 2 Digital subtraction angiography of the left internal carotid artery (black arrow) demonstrating tortuosity and a large saccular aneurysm (white arrow).

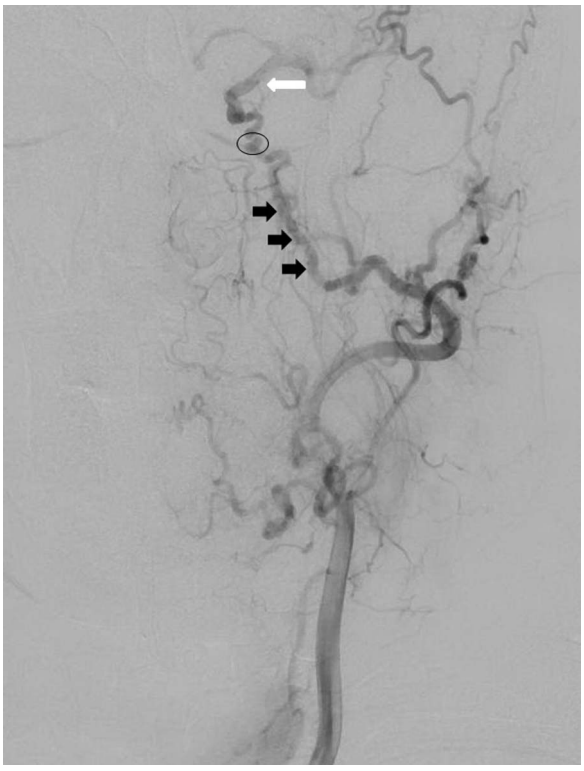

Figure 3 Digital subtraction angiography. Anterior view of the external carotid artery (ECA) injection, demonstrating a tortuous accessory meningeal artery (thick black arrows) from the ECA filling the distal portion of the internal carotid artery (ICA) (white arrow). The ECA–ICA anastomosis occurs through the foramen of Vesalius (circle) via the inferolateral trunk.

balloon test occlusion of the proximal ICA which he tolerated without neurologic symptoms. Subsequently, he was taken to the operating room and underwent resection of the aneurysmal sac and end to end anastomosis of the proximal and distal ICA segments (figure 4). This was performed through a cervical incision along the anterior border of the sternocleidomastoid and, to facilitate distal ICA exposure, left intraoperative mandibular subluxation was performed. The aneurysmal sac was noted to have separate entry and exit openings, and contained a significant amount of thrombus. Histopathological examination revealed fragments of thick walled vessel with chronic inflammation, blood clot, and reactive changes. Postoperatively, the patient remained neurologically intact with no cranial nerve deficits. Postoperative imaging revealed continued patency of the ICA by means of Doppler ultrasonography with no areas of flow acceleration (figure 5).

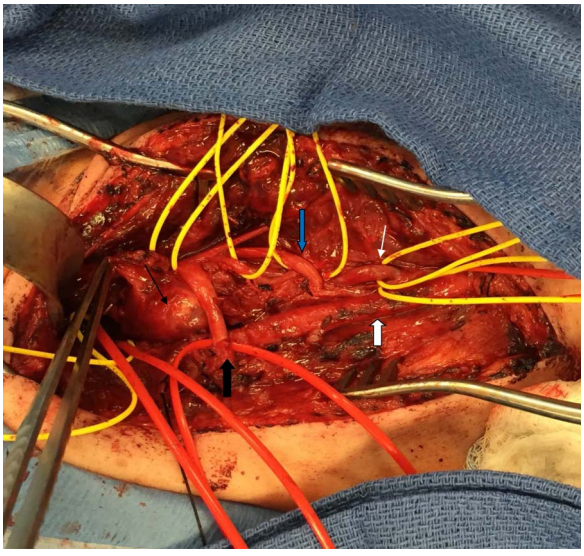

Figure 4 Intraoperative photograph demonstrating neck dissection with vessel loops identifying the internal carotid artery aneurysm (thin black arrow), the proximal internal carotid artery (thin white arrow), the external carotid artery (thick white arrow), the hypoglossal nerve (thick black arrow), and the vagus nerve (blue arrow).

DISCUSSION

Adult configuration of the head and neck vessels is the consequence of a complex process of remodeling, annexation, and involution of the paired ventral and dorsal aortae and six connecting aortic arches (figure 6). The first and second aortic arches involute by the 29th day of development, while forming the future ECA. The third aortic arch gives rise to the carotid bulbs. The fourth aortic arch on the left side remains patent and continues as the normal (left) aortic arch. The dorsal aorta regresses between the third and fourth aortic arch. If the dorsal aorta fails to regress, it is termed ductus caroticus and represents the origin of the left ICA from the arch (figure 7).

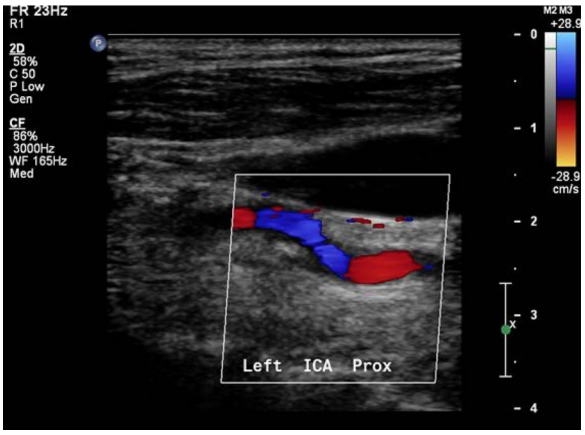

Figure 5 Postoperative Doppler ultrasonography demonstrating the patency of the left internal carotid artery (ICA) following surgical resection of the aneurysm.

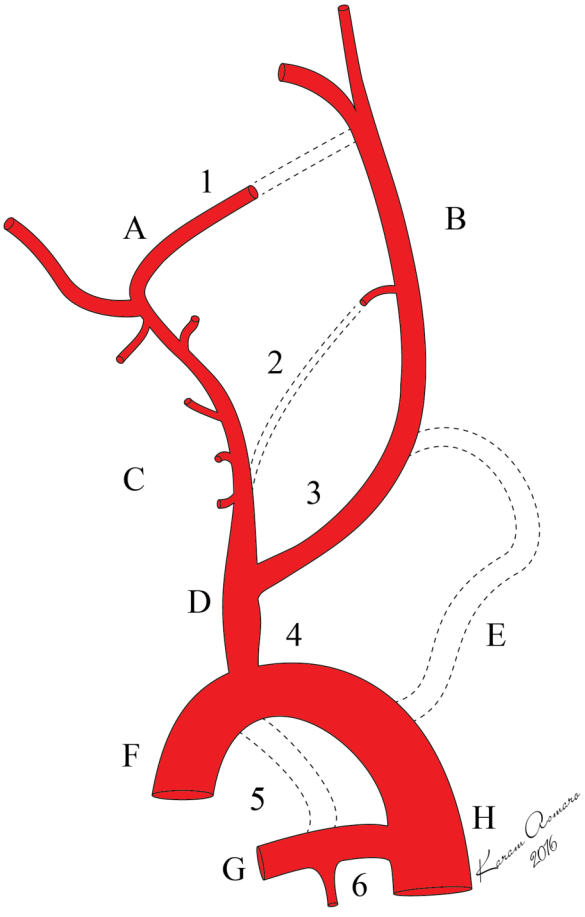

Figure 6 Artist re-creation of the normal aortic arch with common carotid artery. (A) Accessory meningeal artery; (B) internal carotid artery; (C) external carotid artery; (D) carotid bulb; (E) regressed dorsal aorta; (F) ascending aorta; (G) pulmonary artery; and (H) descending aorta. 1=First aortic arch (regressed); 2=second aortic arch (regressed); 3=persisting third aortic arch as carotid bulb; 4=persisting fourth aortic arch as normal left aortic arch; 5=regressed fifth aortic arch; and 6=persisting sixth aortic arch as pulmonary artery.

A literature review of separate origins of the ICA and ECA from the aortic arch revealed few sporadic case reports. A first literature summary of 20 cases was provided by Lie.[2] Since then, only a handful of instances of separate origins from the

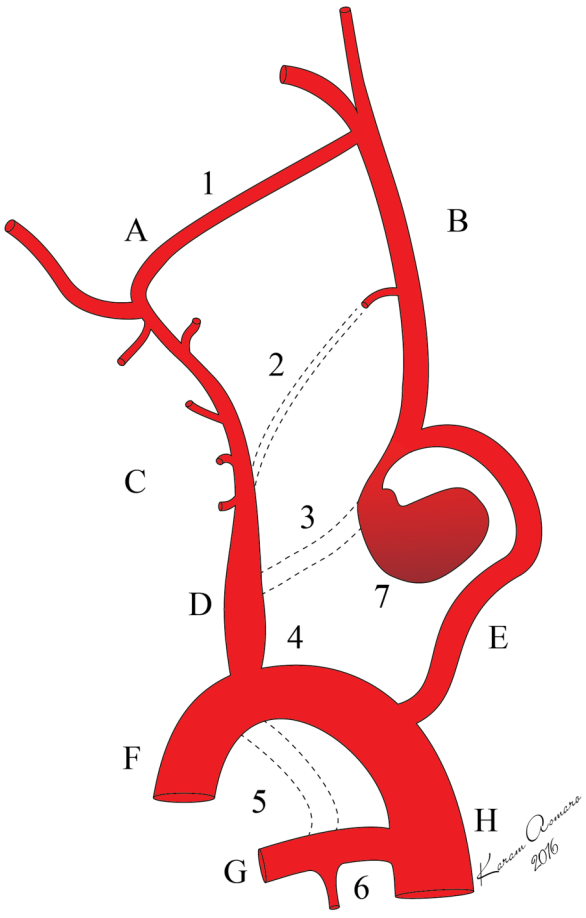

Figure 7 Artist re-creation of abnormal morphology of the patient's aortic arch. (A) Accessory meningeal artery; (B) internal carotid artery (ICA); (C) external carotid artery (ECA); (D) carotid bulb; (E) ductus caroticus (persisting); (F) ascending aorta; (G) pulmonary artery; and (H) descending aorta. 1=Persisting first aortic arch giving ECA–ICA anastomosis via the accessory meningeal artery and inferolateral trunk; 2=second aortic arch (regressed); 3=persisting third aortic arch as carotid bulb; 4=persisting fourth aortic arch as normal left aortic arch; 5=regressed fifth aortic arch; 6=persisting sixth aortic arch as pulmonary artery; and 7=aneurysm sac from incomplete regression of the third aortic arch.

arch have been reported. Associated vascular anomalies such as persistent trigeminal artery, persistent proatlantal artery, cervical aortic arch, and double aortic arches have been described. In one report,[4] a case similar to ours with an aneurysm of the cervical ICA was described.

Our case demonstrates the absence of the common carotid artery with agenesis of the carotid bulb from regression of the third aortic arch with separate origins of the ECA and ICA from the arch (figure 4). The ICA below the aneurysm has a redundant dysplastic appearance. We postulate that the dorsal end of the third aortic arch on the left side failed to regress completely which subsequently led to the aneurysm formation of the ICA. Our hypothesis is based on the location of the aneurysm, at the level of the C3 vertebra, which is one of the more common positions of the carotid bulb,[5] interposed between the ICA and ECA. Interestingly, on imaging and intraoperatively, separate entry and exit openings of the ICA were noted inside the aneurysm, which also supports our hypothesis.

Because of the tortuosity and small caliber of the ICA and the delay in flow related to the large aneurysm, the intracranial circulation on the ipsilateral side was supplemented by a robust

ECA–ICA anastomosis through the foramen of Vesalius between the accessory meningeal artery arising as a tortuous vessel from the internal maxillary artery and the inferolateral trunk. This connection is through persistence of the anatomic pathway of the first aortic arch.[6]

Our patient also carried a diagnosis of Noonan syndrome, a genetically heterogeneous, pleomorphic autosomal dominant disorder, with gene mutations altering proteins involved in the RAS/mitogen activated protein kinase signal transduction pathway leading to several cardiovascular morphological defects as well as other cerebrovascular abnormalities, such as intracranial aneurysms, cavernous hemangiomas, or arteriovenous malformations.[7–9] It appears plausible that the genetic disorder is linked to our patient's underlying vascular abnormalities.

CONCLUSION

In this report, we present a very rare anatomic variant of the aortic arch and its branches in a patient with Noonan syndrome along with a large cervical ICA aneurysm leading to dysphagia. Given the overall anatomy of the carotid arteries and location of the aneurysm, we believe that incomplete regression of the third fetal aortic arch, which normally persists to form the carotid bulb, is the underlying process that resulted in distal ICA aneurysm formation.

Learning points

- The vascular anatomy of the head and neck is the result of several embryologic modifications to the ventral and dorsal aorta and the paired arches.
- Incomplete or lack of normal embryologic modifications to the arches may result in pathology in the vasculature of the head and neck.
- A combined open vascular and endovascular treatment plan is often required to treat vascular pathology in the head and neck.

Contributors RAT and AP contributed manuscript composition and review. KA provided/created the illustrations for the paper. MK and TN were responsible for patient care. Final manuscript review and edits were provided by RAT, TN, and HM.

Competing interests None declared.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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