Separate Origins of Left Internal and External Carotid Arteries from the Aortic Arch and Cervical ICA Aneurysm in a Patient With Noonan Syndrome: Case Report and Review of Literature

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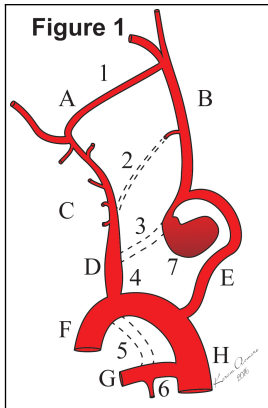
CASE PRESENTATION

47-year-old male with Noonan syndrome who presented for evaluation of neck pain and dysphagia. Physical examination revealed a left pulsatile neck mass. CT and cerebral angiograms demonstrated independent origins of the left ICA and ECA from the aortic arch and absence of a common carotid artery and a large saccular aneurysm was identified originating from the cervical left ICA. It also revealed a normal appearing left ECA whereas the cervical ICA was noted to be tortuous, dysplastic, and of smaller caliber. 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.

SURGICAL TREATMENT

Considering the size of the aneurysm and associated symptoms, treatment was undertaken. The patient underwent balloon test occlusion of the proximal ICA which he tolerated without neurologic symptoms. Subsequently, he underwent resection of the aneurysmal sac and end to end anastomosis of the proximal and distal ICA segments. 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. 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 4).



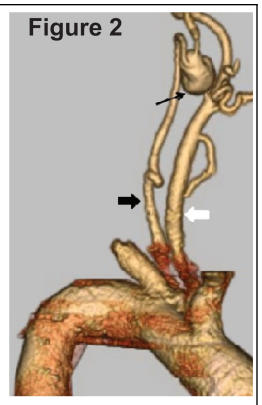


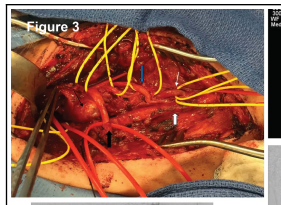
Figure 1 - Artist re-creation of abnormal morphology of the patient's aortic arch: A) Accessory meningeal a., B) ICA, C) ECA, D) Carotid Bulb, E) Ductus Caroticus (persistent), F) Ascending Aorta G) Pulmonary a., H) Descending Aorta, 1) Persistent 1st aortic arch giving ECA-ICA anastamosis via Accessory Meningeal a. and Inferolateral Trunk, 2) 2nd Aortic Arch (regressed), 3) Persistent 3rd Aortic Arch as Carotid Bulb, 4) Persistent 4th aortic arch as normal left aortic arch, 5) Regressed 5th aortic arch, 6) Persistent 6th aortic arch as pulmonary a., 7) Aneurysm from incomplete regression of 3rd aortic arch.

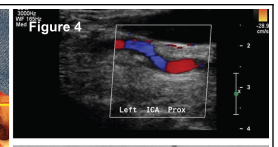
Figure 2 - 3D CT Angiogram: Cervical ICA (thick black arrow) and ECA (white arrow) with clear distinct origins. Aneurysm (thin black arrow) arising from the cervical ICA.

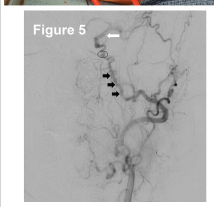
DISCUSSION

A literature review of separate origins of the ICA and ECA from the aortic arch revealed few sporadic case reports. Associated vascular anomalies such as persistent trigeminal artery, persistent proatlantal artery, cervical aortic arch...and double aortic arches have been described. In one report, 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 (figures 1 and 2). 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, 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.

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. It appears plausible that the genetic disorder is linked to our patient's underlying vascular abnormalities.







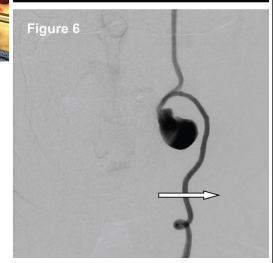


Figure 3 - Intraoperative photograph demonstrating neck dissection with vessel loops identifying the ICA aneurysm (thin black arrow), the proximal ICA (thin white arrow), the ECA (thick white arrow), the hypoglossal nerve (thick black arrow), and the vagus nerve (blue arrow).

Figure 4: Postoperative Doppler ultrasonography demonstrating the patency of the left ICA following surgical resection of the aneurysm.

Figure 5: Digital subtraction angiography. AP view of the ECA injection, demonstrating a tortuous accessory meningeal artery (thick black arrows) from the ECA filling thedistal portion of the ICA (white arrow). The ECA–ICA anastomosis occurs through the foramen of Vesalius (circle) via the inferolateral trunk.

Figure 6: Digital subtraction angiography of the left ICA demonstrating tortuosity and a large saccularaneurysm (white arrow).