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Gangliocapsular Bleed with Ipsilateral Internal Carotid Artery Aplasia

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Summary

Background:

Agenesis requires an extensive work-up as a number of associated other vascular and nonvascular anomalies can be expected. In this scenario, an associated ipsilateral basal ganglia bleeding with subarachnoid haemorrhage with no aetiology is uncommon. We present such a case of moderate ipsilateral ganglio-capsular bleed of unknown cause with associate aortic arch vessel anomaly.

Case Report:

A 45-year-old diabetic man of Indian origin with complaints of a sudden onset of giddiness, left-sided weakness and slurring of speech. Motor system examination revealed power of grade 2. Computed tomography scan revealed a moderate bleeding in the basal ganglia and the right temporo-parietal lobe. Angiography revealed unilateral aplasia of the internal carotid artery.

Patient improved symptomatically with a motor system power of grade 4 after hematoma evacuation and treatment with antibiotics, anti-edema measures and neuroprotective drugs.

Conclusions:

Developmental anomalies of the carotid and aortic arch with intracranial bleeding is a rare occurrence and any arterial anomaly requires extensive evaluation.

MeSH Keywords:

Carotid Artery, Internal • Cerebral Angiography • Intracranial Hemorrhages

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Background

Unilateral agenesis of the internal carotid artery is an extremely rare anomaly with an incidence of 0.01%, and in most cases it is an incidental finding. The presence of extensive collateral circulation ensures that the brain's blood supply is not compromised in almost all such case reports.

Case Report

A 45-year-old man of Indian origin was brought to our hospital with a history of sudden onset of giddiness and fall, following which he had left-sided weakness and slurring of speech. The patient is a known diabetic for 8 years and he was under treatment with an oral hypoglycaemic agent (Metformin 500 mg twice daily). No history of hypertension. The patient is a known alcoholic and smoker for about 15 years. On general examination, his pulse rate was 86/

min and blood pressure was 140/90 mmHg. Auscultation revealed normal cardiovascular and respiratory systems. On palpation, the abdomen was soft, no tenderness, no organomegaly. On auscultation, bowel sounds were normal. The patient was conscious, drowsy, pupils were equal and reacting to light. Left upper motor neuron palsy and left hemiplegia were noted with a power of grade 2 during motor system examination. Biochemical investigations revealed normal parameters. Computed tomography (Figure 1) revealed a moderate bleeding in the basal ganglia and the right temporo-parietal lobe causing a mass effect in the right lateral ventricle and a midline shift of 5 mm to the left. There was diffuse subarachnoid haemorrhage in the basal cistern, tentorial leaflets, interhemispheric fissure, and temporo-parietal lobe. Angiography revealed unilateral aplasia of the Internal Carotid Artery (ICA) (Figure 2). Ipsilateral middle cerebral artery (MCA) was supplied by posterior circulation through the posterior

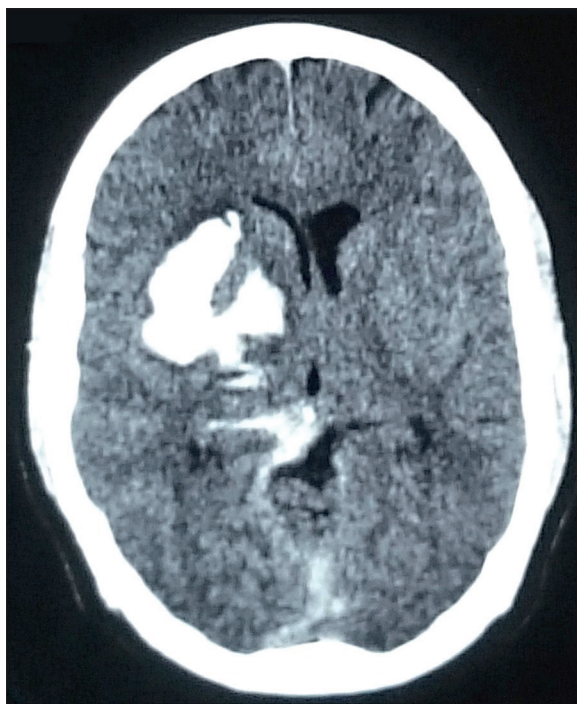


Figure 1. Computed tomography scan showing a moderate sized bleed in basal ganglia and right temporo-parietal lobe.



Figure 2. Angiography showing unilateral aplasia of internal carotid artery.

communicating artery (Figure 3). Common origin of the right innominate artery and the left common carotid artery was seen (Figure 4). The MCA branches were compressed by the hematoma with the presence of vasospasm in the MCA branches and anterior cerebral arteries (ACA). No aneurysm or arteriovenous malformation pathology was identified.

Results

Patient managed by evacuation of hematoma through right fronto-temporal craniotomy and treated with antibiotics,



Figure 3. Ipsilateral middle cerebral artery being supplied by posterior circulation through the posterior communicating artery.

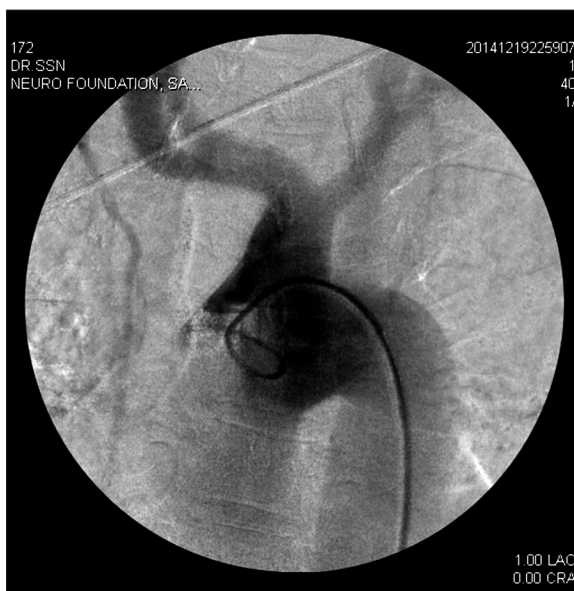


Figure 4. Common origin of right innominate artery and left common carotid artery.

anti-edema measures, neuroprotective drugs (citicoline), mesna nebulisation. The patient improved symptomatically with resolution of bleeding. On discharge the patient was conscious, well oriented and had an improved power of the motor system to grade 4 on the left side.

Discussion

Internal carotid artery is the terminal branch of the common carotid artery and its congenital anomalies have been reported with great rarity and till today the first documented case of ICA agenesis has been by postmortem examination [2]. With just over 100 cases reported in literature, the first cerebral angiography documentation of

ICA agenesis was reported by Verbiest [3]. Vascular anomalies can include agenesis, aplasia or hypoplasia [3]. Agenesis refers to a complete failure of development of ICA with hypoplasia indicating a partially developed small-calibre artery [4]. The suspected event of insult is considered to occur at 4 to 8 weeks of embryonic development with proximal ICA developing from the 3rd aortic arch, and distal ICA from the cranial part of the dorsal aorta at approximately 4 to 5 mm embryonic stage [1,5]. By 6 weeks, development of ICA can be noted and the Circle of Willis gets completed at the stage of 7–24 mm [1,6].

In the presence of aplasia, adequate collateral development occurs with restoration of intracranial flow [7]. As per Lie TA and Padget DH, collaterals may either arise from patent posterior communicating artery and anterior communicating artery (type A) or from patent anterior communicating artery only (type B) [4,6]. At times bilateral agenesis of the anterior cerebral artery will occur with supply of the anterior circulation from carotid vertebrobasilar anastomosis (type C). Segmental internal carotid artery agenesis involving the cervical segment only can occur with intercavernous communication between internal carotid arteries restoring intracranial blood flow in bilateral hemispheres (type D). The diminutive anterior communicating artery is supplied by the bilateral hypoplastic internal carotid artery and the middle cerebral artery is supplied by the enlarged posterior communicating artery in type E. Collateral from the internal maxillary artery extends intracranially in type F. Hence, the collateral blood flow in ICA agenesis or aplasia or hypoplasia depends predominantly on the circle of Willis with rare occurrence of type E and type F. The persistent trigeminal artery is another collateral expected in ICA anomaly [8].

With the absence of the cervical and petrous segment of the internal carotid artery, carotid canal agenesis can be expected [9]. Carotid canal absence indicates a congenital etiology. As ICA is a part of aortic arch development, other associated arch anomalies can be expected. Hasan Dinc et al. reported a case of Horner's syndrome with ICA agenesis

and left-sided origin of the right subclavian artery [10]. Hence, the occurrence of associated aortic arch anomalies can be expected in cases of ICA agenesis. Anomalies such as Tetralogy of Fallot, ventricular septal defect, coarctation have been associated with ICA agenesis along with a higher incidence (25–30%) of intracranial aneurysms [9,11]. CP Hess et al. reported dysgenesis of ICA in PHACE syndrome which involves hemangiomas, posterior fossa anomalies, arterial anomalies, cardiac and eye abnormalities [12]. Dysfunction of cervical neural crest cells which are responsible for muscular and other connective tissue development of the ICA wall is the proposed cause for ICA agenesis in PHACE syndrome [12]. Jelena Djokic Kovac et al. reported that the vascular variation has a significant clinical impact because of some patients being predisposed to cerebrovascular ischemic attacks [13]. Our patient did not have any history of drug intake leading to intracerebral haemorrhage [14].

Many patients with ICA agenesis or arch anomalies are asymptomatic due to good collateral flow and these conditions are usually diagnosed incidentally. However, a patient can present incidentally with infarct, seizure, transient ischemic attacks and subarachnoid haemorrhage [15,16]. Our case is unique with ICA agenesis being associated with ipsilateral gangliocapsular bleeding, subarachnoid haemorrhage, and a common origin of the innominate and the left carotid artery.

Conclusions

Aortic arch and carotid anomalies with intracranial bleeding are a rare occurrence and any arterial anomaly requires an extensive evaluation to rule out associated syndromes, anomalies and other co-morbidities and to initiate any preventive measures, if necessary.

Conflict of interest

The authors declare that they have no conflict of interest.

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