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Review Article

Unilateral absence of the internal carotid artery associated with anterior communicating artery aneurysms: Systematic review and a proposed management algorithm

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ABSTRACT

Background: Absence or hypoplasia of the internal carotid artery (ICA) is a rare congenital anomaly that is mostly unilateral and highly associated with other intracranial vascular anomalies, of which saccular aneurysm is the most common. Blood flow to the circulation of the affected side is maintained by collateral pathways, some of which include the anterior communicating artery (Acom) as part of their anatomy. Therefore, temporary clipping during microsurgery on Acom aneurysms in patients with unilateral ICA anomalies could jeopardize these collaterals and place the patient at risk of ischemic damage. In this paper, we review the literature on cases with a unilaterally absent ICA associated with Acom aneurysms and provide an illustrative case.

Methods: We combined our experience of one case of a unilaterally absent ICA associated with an Acom aneurysm with the 33 existing publications on the same subject in the literature, for a total of 40 cases. We provide a detailed systematic literature review of this association of vascular anomalies, exploring different aspects regarding the collateral pathways and how they impact management strategies and propose a management algorithm to deal with such association.

Results: The mean age was 48.2 ± 16.5 years. The aneurysmal rupture was the most common presentation (75%). Agenesis was observed in 70% of patients, followed by hypoplasia (20%) and, finally, aplasia (10%). Lie Type A was the most common pattern of collaterals (50%), with Types B and D being of almost equal proportions. Most aneurysms were located at the A1-Acom junction contralateral to the anomalous side (Fisher's Exact test; $P = 0.03$). One case of temporary clipping was reported in the literature.

Conclusion: Acom aneurysms in patients with unilateral ICA anomalies, given they are more commonly present contralaterally, could be of acquired etiology, warranting periodic screening in asymptomatic patients. Temporary clipping might be safe in patients with Type D collateral pattern, while those with Types A or B may require intraoperative rupture risk assessment and a tailored management plan to avoid disrupting collateral flow and causing ischemia.

Keywords: Absence, Aneurysm, Anterior communicating artery, Hypoplasia, Internal carotid artery, Unilateral

INTRODUCTION

Congenital anomalies of the internal carotid artery (ICA) is an umbrella term encompassing three developmentally distinct, although interchangeably termed, conditions affecting the ICA, namely, agenesis, aplasia, and hypoplasia.[30] These anomalies are extremely rare, affecting <0.01% of the population with as few as 200 reported cases in the literature. They occur either unilaterally or much less commonly bilaterally with a reported ratio up to 3:1.[29]

Agenesis refers to the complete absence of an artery or its primordium, while aplasia is used to describe an undeveloped ICA with the presence of some of its primordia, such as remnant vessel segments, or a sign of its presence such as a patent ipsilateral carotid canal. These two terms are often collectively referred to in the literature as congenital absence of the ICA.[8,11] In a subtle yet important contrast, hypoplasia describes an artery that is present but underdeveloped.

These anomalies rarely cause symptoms when present in isolation.[33] This observation implies the provision of sufficient collateral blood flow, served in most cases by the contralateral ICA and the vertebrobasilar system through the circle of Willis, or less commonly by persistent primitive pathways (i.e., intercavernous anastomosis) or transcranial collaterals from the external carotid artery (ECA).[8] Hence, the risk lies not in the ICA anomalies themselves, but rather in other cerebrovascular abnormalities reported to be associated with them. Of those abnormalities, intracranial aneurysms project the heaviest burden, with a much higher reported prevalence averaging at about 27.8% compared to an incidence rate of 2–4% in the general population.[22,59]

The anterior communicating artery (Acom) is reported to be the most common location for intracranial aneurysms associated with congenital ICA anomalies.[59] This artery also happens to be part of the anatomy of a major pathway in two of the six patterns of collateral blood flow described by Lie.[30] Thus, neurosurgeons are faced with a dilemma when managing patients with Acom aneurysms associated with a unilaterally absent ICA, as both microsurgical and endovascular management could compromise the artery, leading to loss of this collateral pathway, and an ensuing vascular insufficiency and iatrogenic ischemia in the affected side.

In this article, we provide a detailed, systematic review of the available literature on the unilateral absence of the ICA associated with Acom aneurysms. Clinical aspects of the available cases are abstracted, statistically analyzed and discussed in regards to their etiology, the possible collateral pathways, and their impact on treatment options. The available data are also used to propose a management algorithm for dealing with such conditions.

Illustrative case

A 51-year-old man with a history of hypertension was referred to the Neurosurgical Teaching Hospital in Baghdad after complaints of sudden, severe headache associated with nausea and forceful vomiting. Physical examination showed intact consciousness and normal vital signs. Pupillary examination showed bilaterally equal and reactive pupils of normal size (about 3 mm) and round shape. Cranial nerves examination was unremarkable. The patient had severe neck stiffness and left hemiparesis (muscle power was 3/5).

Non-contrast computed tomography (CT) scan revealed diffuse thick subarachnoid hemorrhage (SAH) with mild ventricular dilatation. CT angiogram (CTA) shown in [Figure 1] revealed a dome-shaped, wide-neck aneurysm arising from the left A1-Acom junction with right-anterior-superior projection. The right ICA was not visualized down to its origin from the common carotid artery (CCA); both the right anterior cerebral artery (ACA) and the middle cerebral artery (MCA) connected to the contralateral ICA through the left A1-Acom pathway. There was no connection between the right posterior communicating artery (Pcom) and the right MCA. Revision of the plain CT scan using bone window

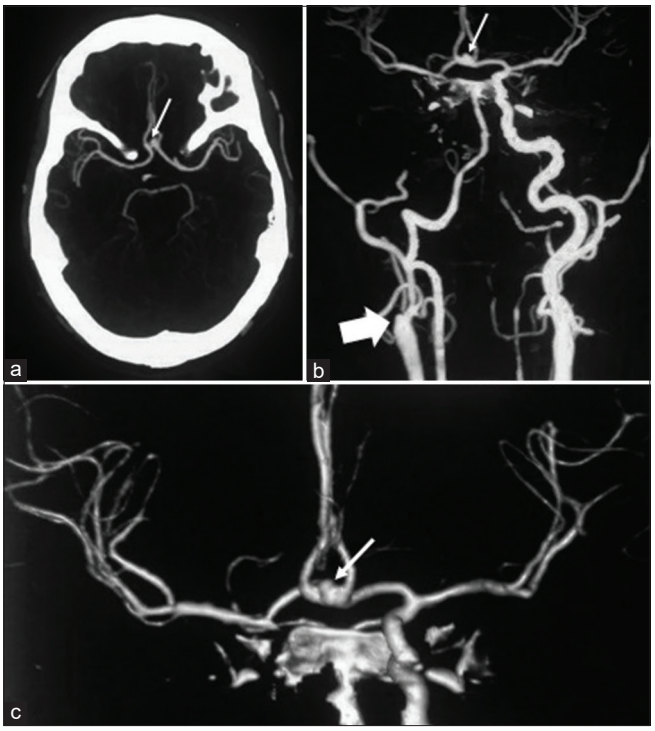

Figure 1: Computed tomography angiogram (CTA) (a) and 3D-reconstructed CTA (b and c) showing a left A1-ACoA junction aneurysm with right superior-anterior projection (thin arrows). The right internal carotid artery (ICA) is not visualized down to its origin from the common carotid artery (thick arrow). The right anterior cerebral artery and middle cerebral artery are supplied by the left ICA through the left A1-ACoA pathway.

revealed an absent carotid canal. Due to the high cost and unavailability of endovascular treatment facilities, the wide neck of the aneurysm, and the need to avoid antiplatelets in the setting of rupture, we were not in favor of endovascular options in this patient and instead opted for surgery.

Microsurgical treatment was performed through a left pterional trans-sylvian approach. During surgery, absence of the right ICA was visually confirmed, and the left A1-Acom complex showed vasospasm. In addition, the aneurysm neck was very wide and its manipulation was difficult without temporary clipping of the left A1, a procedure that would risk blood flow to the right ACA and MCA. Intraoperatively, the aneurysm showed a punctate source of bleeding. A low-voltage bipolar cautery, set at 5–7, was used to shrink the bleeding source. Once bleeding has been controlled, the aneurysm was secured with muscle wrapping. Using low-voltage bipolar cautery to repair vascular injuries or to shrink punctate bleeding is a common practice in cerebrovascular surgery. After surgery, the patient's muscle power improved, he had no further complications, and was discharged on day 7 postoperatively. The lesion remained stable on follow-up imaging.

LITERATURE REVIEW

Methods

A systematic online literature search was performed on the April 14, 2020, for reports on unilateral congenital ICA anomalies associated with Acom aneurysms. PubMed (Medline 1966–2020) and Google Scholar were used for the review, and the search algorithm was as follows: "(((Acom) OR (ACoA) OR (AComA) OR (anterior communicating artery)) AND ((aneurysm) OR (intracranial aneurysm [MeSH])) AND (unilateral) AND (((ica) OR (internal carotid artery) OR (carotid artery, internal [MeSH])) AND ((anomaly) OR (absence) OR (aplasia) OR (agenesis) OR (hypoplasia)))." An additional filter was applied for articles published on humans. The search returned 526 peer-reviewed manuscripts. A systematic abstract screening was performed, and only articles that reported on unilateral congenital ICA anomalies with associated Acom aneurysms were included for final analysis. In addition, the reference list of each article was screened for additional articles. All non-English literature was translated.

The gender, age, presenting signs/symptoms, type, side of the ICA anomaly, and Lie classification for collateral pattern were recorded for each case. Concerning the Acom aneurysm, the side (of its junction with A1), whether it was ruptured or not at presentation, course of treatment, and outcome was recorded. Statistical work was done using IBM Statistical Package for the Social Sciences version 26. Continuous variables were tested for the difference using independent samples *t*-test. Categorical variables were tested for correlation using Fisher's Exact test. The significance level was set at $P < 0.005$.

RESULTS

Our review yielded a total of 33 articles reporting 39 cases of unilateral ICA anomalies associated with Acom aneurysms, aside from the case we present in this article.[1-5,7,9,12-14,18,19,21,24-28,36,37,40,41,43,47-50,52,53,55-58] A table summary of the review is provided in the [Appendix Table 1].

The mean age of patients at presentation was 48.2 ± 16.5 years. Twenty-one (52.5%) of the patients were females, and 29 (72.5%) of cases had a left-sided anomaly of the ICA. Of the aforementioned anomalies, agenesis was most common (70%), followed by hypoplasia (20%). There was no significant difference in age at presentation between males and females ($P = 0.84$) or the types of anomalies ($P = 0.67$). Half of all cases had a Lie Type A collateral pattern, with the remaining half having either Types B or D patterns in almost equal proportions (22.5% and 20%, respectively).

Thirty (75%) of the patients presented with signs of SAH; namely, sudden severe headache with or without nausea and vomiting (three of which were due to ruptured aneurysms other than the Acom aneurysm), while the remaining cases had different presentations (three chronic headache, two incidental, one dysarthria, one seizures and microcephaly, one progressive hearing loss, one numbness, and one unavailable data). Additional vascular anomalies were reported in nine patients; those included absence of the CCA, ECA, and vertebral artery, "cross-over" duplication of the MCA, moyamoya phenomena, and arachnoid cyst. Surgically treated patients had a significantly better outcome compared to non-operative treated patients (Fisher's Exact test, $P = 0.004$).

The demographic data of the study group, as well as characteristics of the Acom aneurysms, are summarized in [Tables 1 and 2], respectively. [Table 3] demonstrates the correlation between the laterality of the ICA anomaly and that of the Acom aneurysm.

DISCUSSION

Given the low prevalence of congential ICA anomalies, the available literature is mainly related to their epidimiology. There has not been much discussion on details of management and follow-up for these patients, particularly for the special circumstance of association with Acom aneurysms. Hence, our review suffers from an inherent lack of clinical data on surgical and long-term follow-up details, and we are therefore forced to depend on anecdotal evidence and single incidental reports from the literature in providing

Table 1: Demographic data of the cases of ICA anomalies associated with Acom aneurysms.

Anomaly	#	Age (years)		Sex			Side of anomaly		Lie classification			
		Mean	SD	M	F	Un.	Lt	Rt	A	B	D	Un.
Agenesis	28	49.9	15.7	10	16	2	20	8	17	6	4	1
Aplasia	4	45	17.6	1	2	1	2	2	1	0	3	-
Hypoplasia	8	44.3	19.9	5	3	-	6	2	2	3	1	2
Total	40	48.2	16.5	16	21	3	29	11	20	9	8	3

SD: Standard deviation, Un.: Unavailable data, M: Male, F: Female, Lt: Left, Rt: Right

Table 2: Characteristics of Acom aneurysms in the study patients.

Aneurysmal characteristics				
Lateralization				
Lt.		Rt.		Un.
11		22		7
Rupture				
Yes		No		Un.
30		9		1
Management				
Clipping	**Coiling**	**Muscle wrapping**	**Conservative**	**Un.**
23	1	3	7	6
Outcome				
Good		**Poor**		**Un.**
27		5		8

Un.: Unavailable data, Lt: Left, Rt: Right

Table 3: Association between the side of ICA anomalies and Acom aneurysms.

ICA anomaly lateralization	Aneurysmal lateralization		
	Left	Right	Total
Left	5	19	24
Right	6	3	9
Total	11	22	**33**

P=0.03, Fisher's Exact test. Seven cases were excluded due to missing data.

recommendations for the management of such patients. In our case, the bleeding point was cauterized, and the aneurysm was muscle wrapped. This management strategy was also adopted by Burmester and Stender. However, other management options, including clipping, and endovascular coiling, should all be considered by the surgeon when faced with such lesions.

Patterns of collateral flow in unilateral absence of the ICA

In his original 1968 thesis, Lie describes six patterns of collateralization to supply blood to the hemisphere affected by ICA agenesis, aplasia, or severe hypoplasia.[30] Types C and E describe the patterns in bilateral ICA anomalies. Type F, characterized by unilateral or bilateral "rete mirabile;" a web-like mesh of thin arteries at the skull base connecting and supplying the anomalous ICA(s) through the ECA(s), has not been documented to be associated with Acom aneurysms. Therefore, the three above-mentioned types were not considered in the context of this study and are henceforth not discussed.

In Type A, the ACA of the affected side is supplied by the contralateral ICA through the contralateral A1 and Acom, while the MCA of the affected side is supplied by the posterior circulation through an enlarged ipsilateral Pcom. In type B, both the ACA and MCA of the affected side are supplied by the contralateral ICA through the contralateral A1 and Acom. Type D describes a "trans-cavernous" pattern of collateralization, in which an embryonic remnant vessel originating from the cavernous portion of the normal ICA persists. This remnant vessel reconstitutes blood flow either by anastomosing with the cavernous portion of the affected ICA (a pattern we refer to as D1), or by directly supplying the MCA of the affected side (D2).

Of particular interest, we observed that patients with Lie Type D collateral pattern, the rarest type in unilateral anomalies, displayed a much higher affinity for Acom aneurysms compared to other types. Of the total 20 cases of ICA anomaly with type D pattern reported in the literature, eight of them had saccular intracranial aneurysms, and all of them had an Acom aneurysm (sole in six cases, associated with other aneurysms in one case).[24]

Etiology of Acom aneurysms associated with unilateral absence or hypoplasia of ICA

Although the exact etiological factors causing intracranial saccular aneurysms in patients with anomalies of the ICA are not well studied, several articles propose two main possible mechanisms. The congenital theory implies the coexistence of the aneurysm alongside the ICA anomaly since fetal life.[51,54] In contrast, the acquired theory implicates the changes in blood flow brought about by the abnormal flow vectors within the collateral pathways as a cause for the development of the aneurysms.[55] The data we reviewed seems to support the latter acquired etiology as a more plausible cause, at least in the particular case of Acom aneurysms. For one, the

mean age at presentation was 48 years, making an acquired etiology more probable. Second, a majority of the cases had the Acom aneurysm at a side opposing that of the ICA anomaly, as shown in [Table 3], which supports the theory that altered blood flow vectors in the enlarged contralateral circulation constitute stress on the vessel wall, leading to the development of aneurysms.

Need for periodic aneurysm screening

Despite the rarity of ICA absence or hypoplasia, which are estimated to be prevalent in <0.01% of the population, the prevalence of intracranial saccular aneurysms in association with them (ranging from 25 to 67%) is much higher compared to that of the normal general population (2–4%).[1,25,53] Some articles suggest screening for intracranial aneurysms in individuals with a higher risk than that of the general population, such as those with positive family history or autosomal dominant polycystic kidney disease.[6,44] The above-mentioned points, alongside the proposition of the aneurysms being of an acquired nature within this context, may support periodic screening of such patients to detect the development of new aneurysms.

Magnetic resonance angiography (MRA) might be preferable in this setting over catheter or CT angiography, given its non-invasive, non-ionizing properties, and good sensitivity profile. The previous studies support the suggestion that MRA is a valid screening tool for intracranial aneurysms, as recent advances have significantly improved its sensitivity and specificity.[6] Despite that, it is recognized that there is an uncertainty concerning the cost-effectiveness of MRA periodic screening and that there is an occurrence of false-positive and false-negative aneurysm detection on MR angiograms, especially for small aneurysms.[16,45] In cases, where there is a particularly high suspicion for the presence of an aneurysm, more sensitive imaging tools like CTA might be a better option.

Preservation of collateral flow during microneurosurgery

Temporary clipping is often employed during microvascular clipping of intracranial aneurysms, either in an elective manner to acquire proximal or distal control of vessels to facilitate the manipulation of the aneurysm complex, or as a rescue measure to control bleeding caused by intraoperative rupture (IOR) of the aneurysm. For Acom aneurysms, the temporary clipping usually involves the A1 segment of the ipsilateral ACA. Temporary clipping is not associated with additional risk of delayed cerebral infarction or delayed ischemic neurological deficits in patients with bilaterally normal ICA formation.[31] However, these findings might not apply to patients suffering from aneurysms associated with unilateral congenital ICA anomalies. Given the anatomical relation of Acom aneurysms to the collateral pathways in several of the patterns described by Lie and illustrated in [Figure 2], temporary clipping during microsurgery of these aneurysms could be of particular concern.

For Type D1, blood flow is reinstituted to the affected ICA at its cavernous portion far from the Acom, and as such, no special considerations need to be taken when managing such a combination. On the other hand, the contralateral A1 segment and Acom constitute a major collateralization pathway in ICA anomaly patients with Type A, B, and D2 patterns.

In Type A, the collateral pathway supplies the ACA, while the MCA is supplied by the posterior circulation through an enlarged ipsilateral Pcom. Disruption of the A1-Acom pathway by temporary clipping in such patients would compromise blood flow to the affected side ACA, sparing the MCA. Bhaskar *et al.*[5] reported a case with such conditions who presented with signs of aneurysmal rupture. IOR occurred during microsurgical clipping, and temporary clipping of the unaffected side A1 segment for 5 min was done. The aneurysm was clipped, and the patient recovered without

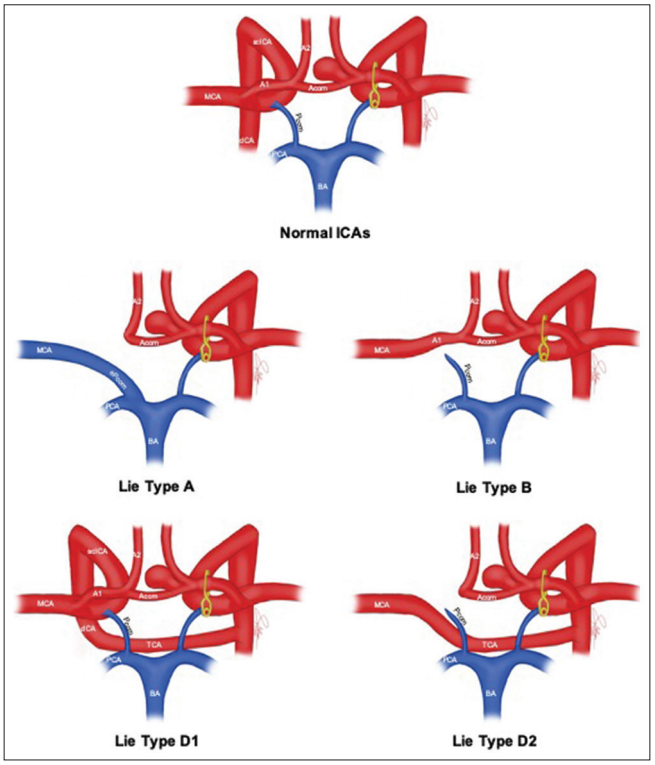

Figure 2: Figure artistic depiction of the anatomical position of ACOM aneurysm in normal and unilateral congenitally absent ICAs and its relation to collateral blood flow patterns. CICA: Cavernous internal carotid artery, SCICA: Supraclinoid internal carotid artery, A1, A2: Segments 1 and 2 of the anterior cerebral artery, ACOM: Anterior communicating artery, MCA: Middle cerebral artery, BA: Basilar artery, PCA: Posterior cerebral artery, PCOM: Posterior communicating artery, EPCOM: Enlarged posterior communicating artery, TCA: Transcavernous anastomosis.

complications. At the 6-month follow-up appointment, the patient had no neurological deficits. The absence of postoperative complications could be attributed to the short duration of temporary clipping. It has been shown that temporary clipping for <20 min under induced hypertension and mild hypothermic technique is an acceptable practice that causes no additional risk of delayed cerebral infarction.[38] In addition, electrophysiological intraoperative monitoring and propofol burst suppression may help ameliorate the risk of hypoperfusion injury and ischemia.[23] However, those studies were conducted in the context of bilaterally intact ICAs. Based on the above-mentioned case report, it can be assumed that temporary clipping can be employed in such patients, at least in an emergency context, to stop bleeding from IOR. The same can be said of Type D2 pattern cases, as the anatomy is similar in the context of collateral flow to the affected side ACA through the contralateral A1-Acom pathway.

In contrast, Type B, in which the collateral pathway supplies blood flow to both the ipsilateral ACA and MCA, the anatomy implies that disruption of these collaterals would cutoff all blood supply to a major portion of the affected hemisphere.

Based on the above flow patterns for Types A, B, and D2, it is a reasonable practice to assess the risk of IOR before intervention and determine the course of action accordingly.

Endovascular intervention is a very viable initial treatment option for intracranial aneurysms, both ruptured and unruptured.[20,34] However, there is still some concern regarding its use in aneurysms with difficult morphological features and the frequent need for antiplatelet therapy, most notably with flow diversion. This often prohibits its use in ruptured cases.[42] In addition, endovascular treatment entails higher costs compared to clipping, and its facilities are not widely available, particularly in developing countries. In this review, we only found one case of Acom aneurysm associated with ICA hypoplasia treated by endovascular coiling.[21] For high-risk patients, endovascular coiling might be considered as an alternative to microsurgery as it has an overall lower risk of rupture and does not require temporary clipping.[15]

Based on all data discussed above, we have devised a simple algorithm for the management of patients presenting with unilateral absence or severe hypoplasia of the ICA in the context of associated aneurysms, as shown in Figure 3.

Figure 3: Proposed outline for anterior circulation aneurysm-contextual management of patients presenting with unilateral congenital ICA anomalies. *For ICA anomalies with no associated aneurysms, we propose periodic screening, preferably using MRA as a non-invasive safe method. **Generally, ruptured aneurysms have a higher risk of IOR compared to unruptured aneurysms. High risk group (unruptured aneurysms): large (for clipping) or very small (for coiling) size, anteriorly directed dome, irregular shape with daughter cysts, high aspect ratio. High risk group (ruptured aneurysms): High modified Fisher's or Hunt and Hess grade, rebleeding before intervention.

Some studies have suggested that ruptured Acom aneurysms have the greatest risk for re-rupture during microsurgery among all aneurysmal sites.[15] This pre-imposed risk necessitates checking for other factors that increase the risk of IOR and thus emergency temporary clipping in patients with anomalous ICA and anterior circulation. Overall, ruptured aneurysms have a higher risk of IOR compared to unruptured aneurysms. The specific risk of IOR during microsurgical clipping for unruptured aneurysms can be assessed in the context of the surgeon's experience on factors such as large (for clipping) or very small (<4 mm – for coiling) size, anteriorly directed dome, irregularly-shaped aneurysms with Murphy's teats (daughter cysts), and a high aspect ratio.[10,17,32,35,46] Additional factors for assessing IOR risk in ruptured aneurysms include high modified Fisher's or Hunt and Hess grades and rebleeding before intervention.[39] If the surgeon deems the case to have a high risk for IOR during microsurgery, then endovascular coiling should be considered, particularly in patients with Lie Types A, B, and D2 collateral patterns.

CONCLUSION

Intracranial saccular aneurysms are a common finding in patients with unilateral congenital anomalies of the ICA and could be of acquired etiology in such patients. Periodic screening for patients with these congenital anomalies for the development of aneurysms could be warranted. Acom aneurysms in association with Lie Type D1 collateral pattern can be managed as in patients with bilaterally normal ICAs. Acom aneurysms associated with Lie Types A, B, or D2 collateral patterns might require special considerations during microsurgical clipping, as the application of temporary clips could compromise collateral blood flow and cause ischemic damage. Endovascular therapy should be more strongly considered in those cases.

Declaration of patient consent

Patient's consent not required as patients identity is not disclosed or compromised.

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Conflicts of interest

There are no conflicts of interest.

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APPENDIX

Appendix Table 1: A table summary of the systematic review performed in this article.

Report			Case								
N.	First author	Year	Patient age/gender	Presentation	ICA anomaly	Lie classification (collateral flow)	Acom aneurysm			Outcome	Notes
							Site	Ruptured	Mgmt.		
1	Cohen and Kristiansen[12]	1957	-	-	Lt. agenesis	-	-	-	-	-	-
2	Lagarde *et al.*,[26]	1957	42 y/M	-	Rt. hypoplasia	Type B	-	Yes	-	-	-
3	Burmester and Stender[7]	1961	43 y/M	Sudden "raging" headache, projectile vomiting, LOC	Rt. agenesis	Type A	Lt. junc.	Yes	Muscle wrapping	Good	Rt. frontal craniotomy, rt. frontal pole resection, wrapping with musclePatient improved and went on living healthily.
			32 y/F	Sudden "violent" headache	Lt. agenesis	Type B	Rt. junc.	Yes	Muscle wrapping	Good	Rt. frontal craniotomy, partial rt. frontal pole resection, wrapping with muscle Patient improved and went on living healthily
4	Lhermitte[28]	1968	63 y/F	Sudden severe headache, vomiting	Lt. hypoplasia	-	-	Yes	-	-	-
5	Teal *et al.*,[52]	1973	66 y/F	Incidental, admitted after FFH	Lt. agenesis	Type A	Lt. junc.	No	None	Poor	+ Multiple small aneurysms of rt. ICA+ Absent rt. ECA Patient died 3 days post subacute SHD evacuation
6	Naito *et al.*,[36]	1977	48 y/M	Lt-sided numbness and LL weakness	Lt. agenesis	Type A	-	Yes	Clipping	Good	+ Absent supraoptic rt. ACA Improvement postoperative
7	Waga *et al.*,[55]	1978	60 y/F	Severe frontal headache, N/V	Lt. aplasia	Type A	Rt. junc.	Yes	Clipping	-	+ Absent lt. CCA and ECA Uneventful clipping: Fronto-temporal craniotomy
8	Bernini *et al.*,[4]	1980	38 y/F	Sudden severe headache, vomiting, left hemiparesis	Rt. agenesis	Type A	Lt. junc.	Yes	Conservative	Good	+ Lt. vertebral artery agenesis Pt. refused surgery, improved afterwards and is well
9	Huber[19]	1980	26 y/-	Sudden severe headache, DLoC	Lt. aplasia	Type D	Rt. junc.	Yes	Clipping	Poor	+ Autosomal dominant polycystic kidney disease Pt. died on day 2 postoperative after severe secondary hemorrhage and respiratory failure

(*Contd...*)

Appendix Table 1: *(Continued).*

Report			Case								
N.	First author	Year	Patient age/gender	Presentation	ICA anomaly	Lie classification (collateral flow)	Acom aneurysm			Outcome	Notes
							Site	Ruptured	Mgmt.		
10	Shigemori *et al.*,[47]	1980	48 y/F	Sudden severe headache, vomiting	Lt. agenesis	Type A	Lt. junc.	Yes	Clipping	Good	Uneventful clipping: Fronto-baso-lateral (pterional) approach Full recovery
11	Afifi *et al.*,[1]*	1987	4 m/F	Microcephaly, seizures	Lt. hypoplasia	-	Rt. junc.	No	Clipping	Good	Coexisting cerebral hemiatrophy Transient DI, improvement at 7m follow-up
12	Kunishio *et al.*,[25]	1987	70 y/M	Sudden severe headache	Lt. agenesis	Type A	Rt. junc.	No	None	-	+ Unruptured basilar aneurysm + Ruptured MCA aneurysm + Lt. CCA and ECA agenesis Lt. fronto-temporal craniotomy for (for MCA aneurysm clipping)
13	Petrela *et al.*,[41]	1987	53 y/F	Sudden severe headache, vomiting	Rt. agenesis	Type A	Lt. junc.	Yes	Clipping	Good	2 Acom aneurysms + "cross-over" duplication of MCA ((interhemispheric aneurysm was probably a thrombosed lesion of Acom)) Uneventful clipping: Rt. pterional craniotomy
							-	Yes	Clipping		
14	Tracy[53]	1987	34 y/M	Acute-onset dizziness, brief LOC, intermittent headaches	Rt. aplasia	Type D	Lt. junc.	Yes	Clipping	Good	Uneventful clipping: lt. fronto-temporal craniotomy
15	Yoshida *et al.*,[58]	1988	67 y/F	Sudden severe headache, LOC	Lt. agenesis	Type B	-	Yes	Conservative	Good	Pt. died after 15 days of hospitalization
16	Quint *et al.*,[43]	1989	60 y/F	Lt.-sided headache and decreased visual acuity	Rt. aplasia	Type D	Lt. junc.	No	Conservative	Poor	+ giant (3cm) supraclinoid lt. ICA aneurysm + rt. MCA trifurcation aneurysm Pt. died after 3wks, 6 days following an episode of SAH
			65 y/F	Sudden severe headache	Lt. agenesis	Type D	Lt. junc.	Yes	Clipping	Good	Uneventful clipping
17	Nakai *et al.*,[37]	1992	27 y/F	Sudden severe headache, vomiting	Rt. Agenesis	Type B	-	Yes	-	-	-

(Contd...)

Appendix Table 1: *(Continued).*

Report			Case								
N.	First author	Year	Patient age/gender	Presentation	ICA anomaly	Lie classification (collateral flow)	Acom aneurysm			Outcome	Notes
							Site	Ruptured	Mgmt.		
18	Armand *et al.,*[2]	1996	-	Sudden severe headache	Lt. agenesis	Type B	-	Yes	-	-	-
19	Tanaka *et al.,*[50]	1996	45 y/M	Sudden severe headache, DLoC	Lt. hypoplasia	Type D	Rt. junc.	Yes	-	-	+ Lt. rete carotidis
20	Czarnecki *et al.,*[13]	1998	45 y/M	Progressive hearing loss	Lt. hypoplasia	Type B	Rt. junc.	No	Clipping	Good	2 Acom aneurysms + Coexisting absent supraclinoid ICA Uneventful clipping: rt.-sided approach
21	Lee *et al.,*[27]	2003	55 y/M	Sudden severe headache, DLoC	Rt. agenesis	Type A	Rt. junc.	Yes	Clipping	Good	Uneventful clipping: Pterional craniotomy
			19 y/F	Sudden severe headache, DLoC	Lt. agenesis	Type A	Lt. junc.	Yes	Clipping	Good	+ lt. CCA hypoplasia + lt. temporal arachnoid cyst Uneventful clipping: Pterional craniotomy
			51 y/M	Sudden severe headache, DLoC	Rt. agenesis	Type A	Rt. junc.	Yes	Clipping	Good	Uneventful clipping: Pterional craniotomy
			57 y/F	Headache, general weakness	Lt. agenesis	Type B	Lt. junc.	No	Clipping	Good	Uneventful clipping: Pterional craniotomy
22	Wong *et al.,*[57]	2006	81 y/F	Sudden LOC	Lt. agenesis	Type A	Rt. junc.	Yes	Conservative	Poor	+ Rt. MCA moyamoya phenomenon Not suitable for embolization or microsurgery, pt. died 15 days later of pneumonia
23	Orakdöğen *et al.,*[40]	2007	43 y/F	Sudden severe headache, vomiting, DLoC	Lt. agenesis	Type A	Rt. junc.	Yes	Clipping	Good	Uneventful clipping: rt. pterional trans-sylvian approach

(Contd...)

Appendix Table 1: *(Continued).*

Report			Case							
N.	First author	Year	Patient age/gender	Presentation	ICA anomaly	Lie classification (collateral flow)	Acom aneurysm			Outcome	Notes
							Site	Ruptured	Mgmt.		
24	Baek *et al.*,[3]	2007	58 y/M	Dysarthria, general weakness	Lt. hypoplasia	Type B	Rt. junc.	No	Clipping	Good	+ Lt. A1 ACA aneurysm (rt. projection) Direct clipping of Acom aneurysm: rt. pterional approach Direct clipping of A1 aneurysm via lt. pterional approach 2wks later, weak nearby arterial walls reinforced with cotton sheets and fibrin glue. Good recovery
			38 y/F	Sudden severe headache	Lt. hypoplasia	Type A	Rt. junc.	Yes	Clipping	Good	Direct clipping + weak nearby arterial walls reinforced with cotton sheets and fibrin glue.Good recovery
25	Demirgil *et al.*,[14]	2007	18 y/F	Sudden severe headache, vomiting	Lt. agenesis	Type A	Rt. junc.	Yes	Clipping	Good	Uneventful clipping: rt. pterional craniotomy 3m follow-up good
26	Chen *et al.*,[9]	2008	53 y/F	Sudden severe headache, vomiting	Rt. agenesis	Type A	Lt. junc.	Yes	None	Poor	+ Absent rt. CCA and ECA Pt. died 10 days after refusing surgery
27	Horie *et al.*,[18]	2008	55 y/F	None, incidental	Lt. agenesis	Type D	Rt. junc.	No	Clipping	Good	Uneventful clipping: lt. fronto-temporal craniotomy discharged 1wk later
28	Itokawa *et al.*,[21]*	2009	62 y/M	Continual severe headache	Lt. hypoplasia	Type A	Rt. junc.	No	Coiling	Good	Irregular large aneurysm 27 coils, 419cm total2 caths used for framing, 18 microcath for intrasellar, 10 microcatheter for suprasellar portion no complications at 3y follow-up
29	Suyama *et al.*,[49]	2009	69 y/F	Sudden severe headache	Lt. agenesis	Type D	Rt. junc.	Yes	Clipping	Good	+ Anomalouos lt. MCA from the contralateral ICA Uneventful clipping: rt. fronto-temporal craniotomy

(Contd...)

Appendix Table 1: *(Continued).*

Report			Case								
N.	**First author**	**Year**	**Patient age/gender**	**Presentation**	**ICA anomaly**	**Lie classification (collateral flow)**	**Acom aneurysm**			**Outcome**	**Notes**
							Site	**Ruptured**	**Mgmt.**		
30	Wani *et al.*,[56]	2011	48 y/M	Sudden severe headache, vomiting, DLoC	Lt. agenesis	Type A	Rt. junc.	No	Clipping	Good	+ ruptured rt. paraclinoid aneurysm Uneventful clipping: rt. pterional transsylvian approachFor paraclinoid aneurysm: ant. clinoid drilling, ICA mobilization, clipping with fenestrated clip
31	Bhaskar *et al.*,[5]	2012	32 y/M	Sudden severe headache, brief LOC	Lt. agenesis	Type A	Rt. junc.	Yes	Clipping	Good	2 Acom aneurysms Clipping by rt. pterional craniotomy, transsylvian approach, intraoperative rupture, temporary clipping of rt. A1 for 5 min. Patient is well at 7m follow-up
32	Kumagai *et al.*,[24]	2017	47 y/M	Lt.-sided numbness	Lt. agenesis	Type D	Rt. junc.	No	Clipping	Good	Uneventful clipping: frontal craniotomy, interhemispheric approach
							Rt. junc.	No	Clipping		
33	Shukla *et al.*,[48]	2018	60 y/M	Chronic headache	Lt. agenesis	Type A	Rt. junc.	No	Follow-up	Good	No complaints, under follow-up
34	Al-Khafaji	2020	51 y/M	Sudden severe headache, nausea and violent	Rt. agenesis	Type B	Lt. junc.	Yes	Cautery and Muscle wrapping	Good	Lt. pterional transsylvian approach; vasospasm and wide-neck made clipping difficult without proximal control by temporary clipping; resorted to cautery of bleeding part and muscle wrapping instead

*The only two cases of large (more than 10 mm largest diameter) Acom aneurysms associated with ICA anomaly reported in the literature