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RESEARCH

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Management of large or giant Extracranial carotid artery aneurysms: a single-center experience

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Abstract

Background Extracranial carotid artery aneurysm (ECCA) is an infrequent disease with an incidence of less than 1%. However, our understanding is still incomplete, and the preferred method to treat ECAA remains unknown.

Methods To share our initial experience with treatment options for large ECCAs. We have retrospectively included 15 patients who underwent treatment at our institution from 2015 to 2022. The treatment modality, patient demography, aneurysm morphology, and clinical and radiographic follow-data were collected and analyzed in all patients.

Results During the study period, 15 patients (with 19 ECCAs) were diagnosed and treated, of whom 8 (53.3%) were male. The average age of the patients was 53.6 years. The primary presenting symptoms was pulsatile neck mass (10/15, 66.7%). The etiology of ECAAs included atherosclerotic (6/15, 40.0%), infectious (3/15, 20.0%), and dissecting (1/15, 6.7%), and iatrogenic due to acupuncture (1/15, 6.7%). The mean ± SD maximal diameter of the aneurysms was 23.8 ± 14.1 mm, with more than half of patients having aneurysms larger than 25 mm (52.6%). 79.0% aneurysm had intraluminal thrombus at admission. Six patients underwent successful neurosurgical resection for a total of seven ECCAs. Five patients received endovascular interventional treatment. The remaining four patients who presented with seven ECCAs were placed under observation. The mean follow-up period was 28.1 months. Out of the patients who received treatment, it was discovered that 10 of them had completely occluded at the latest imaging study. During the course of conservative observation, it was observed that in one patient, the aneurysm disappeared and the parent vessel became thinner following anti-infection treatment. Out of all the patients, only one who was treated with Willis stent experienced a large area of cerebral infarction after treatment, which ultimately resulted in their death.

Conclusions ECCA is rare and mostly present with mass effect. Neurosurgical treatment was more frequently feasible in large ECCAs, and endovascular surgery was the first choice for pseudoaneurysms and dissecting aneurysms. Anti-inflammation treatment was available for some infectious cases.

Keywords Aneurysm, Carotid artery disease, Endovascular treatment, Surgical treatment

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Background

Extracranial carotid artery aneurysm (ECCA) is an infrequent disease, with an incidence of less than 1% and was estimated to account for 0.4–4% of all peripheral arterial aneurysm [1, 2]. Early diagnosis of ECCA is challenging due to its clinical asymptomatic nature and occasional discovery during radiological examinations [3]. Additionally, distinguishing the underlying pathogenesis of ECCA further complicates its diagnosis. However, it is important to note that ECCA poses a significant risk for neurological thromboembolic events, with rates exceeding 50% [1].

Unlike intracranial saccular aneurysms, there have been past case reports of neurosurgical and endovascular treatments for ECCAs [4, 5]. Welleweerd and colleagues conducted a review of 39 articles encompassing 1239 patients with ECAAs. Their analysis revealed that 89% of patients received surgical treatment, while the remaining 11% were managed conservatively [6]. However, due to the lack of systematic study data, experience, and guidelines, as well as the presence of various etiologies, there is still a debate on the most effective treatment strategy. The characteristics of ECCAs play a significant role in determining the most suitable treatment strategies. As a result, neurosurgeons are presented with unique challenges specific to each individual case.

The purpose of this study is to provide a retrospective review of 15 patients who were diagnosed with 19 ECCAs at our center and underwent varying treatments such as conservative, neurosurgical, or endovascular treatments. By presenting our initial experience in treating ECCA, we aim to share valuable insights.

Methods

Study design

We have retrospectively included 15 ECCA' patients who underwent treatment at our single center from 2015 to 2022. The Ethics Committee of the local hospital has approved the study (Approval No. XJMU 20171018-01). Informed consent was obtained from each patient or their relatives during hospitalization.

ECCAs define and classification

All cases of ECCA were diagnosed using either CTA or DSA. ECCA was defined as a dilation of the carotid bulb that exceeded 1.5 times the diameter of the common carotid artery or over 2 times the diameter of the internal carotid artery (ICA). This definition takes into account that the normal carotid bifurcation is usually 40% larger in diameter than the more distal ICA [5]. In our study, we classified the location of aneurysms into five types according to the Attigah classification: type I, which refers to an aneurysm of the internal carotid artery distal to the carotid bifurcation; type II, aneurysm of the internal carotid artery; type III, aneurysm of the carotid bifurcation; type IV, aneurysm of both the internal carotid artery and the common carotid artery; and type V, aneurysm of the common carotid artery [4].

Data collection

We collected demographic and clinical data from the medical records of all patients. This data included age, gender, smoking habits, alcohol abuse history, family history, multiplicity, presence of symptoms, and any underlying conditions such as hypertension, hyperlipemia, diabetes mellitus, and coronary heart disease. We obtained anonymized DICOM image data and utilized them to measure parameters related to aneurysm morphology and classify ECCAs.

Treatment strategy

Three treatment methods were available for the management of these patients. The choice of treatment methods was based on pathological etiology, aneurysm location and morphology, as well as the parent artery and contralateral artery. The carotid artery was exposed, and the aneurysm was dissected and mobilized after both proximal and distal clamping. Standard protocols were followed throughout the treatment. The chosen method for reconstructing carotid vasculature depended on both the anatomic condition and the surgeon's discretion. Intraoperative indocyanine green video angiography was used to confirm the successful exclusion of the aneurysm while also preserving the branches. Additionally, aneurysm specimens were meticulously collected during the operation and pathological examination was performed.

Endovascular procedures were under general anesthesia. The treatment access was gained through percutaneous femoral artery puncture, with a intravenous bolus of heparin administered to achieve anticoagulation. Selection of treatment materials (such as coiling with or without stent and flow diverts) and procedure (including occlusion, covered stent, willis or pipeline) was based on the diameter of the parent artery and aneurysm characteristics. Patients who underwent stent deployment were discharged with oral antiplatelet therapy, consisting of long-term Aspirin (100 mg/day) and Clopidogrel (75 mg/day) for a minimum of 5 days before operation and six months after operation, along with long-term statins (20 mg/day).

Conservative treatment was administered to patients who presented with contralateral internal carotid artery occlusion, posterior circulation dissection aneurysm, and infected aneurysm. Those with infected aneurysms underwent biopsy and received anti-infection treatment.

In our study, we documented various complications, including those related to death, as well as cardiac and cerebral vascular events. We considered the treatment a success if the patient's symptoms improved, and there were no major complications during their hospital stay.

Follow-up

All patients were advised to undergo routinely (1, 3, 6, and every 12 months) follow-up after treatment using CTA or DSA. Additionally, their general condition was monitored through telephone communication after discharge. The study investigated the patency of the reconstructed arteries as well as any complications that arose from the intervention, such as bleeding, infection, nerve injury, stroke, and death. Stenosis is defined as a lumen stenosis greater than 50% [7]. Prior to treatment initiation, all patients were contacted by telephone for an interview.

Statistical analysis

In this study, we used means (SD) to summarize continuous variables, while frequency (percentages) were used to describe categorical variables. To carry out data management and statistical analyses, we employed SPSS V.25.0 (IBM Corp, Armonk, New Y, USA).

Results

Sample baseline and aneurysm characteristics

Table 1 provides a summary of the clinical features of all patients included in this study. There were 8 men and 7 women, and mean age was 53.6 years, ranging from 30 to 74 years (Table 2). The primary presenting symptoms was pulsatile neck mass (10/15, 66.7%).The etiology of ECAAs included atherosclerotic (6/15, 40.0%), infectious (3/15, 20.0%), and dissecting (1/15, 6.7%), and iatrogenic due to bacupuncture (1/15, 6.7%). Hyperlipidemia was the most prevalent comorbidity (53.3%), and 33.3% had hypertension. A positive smoking history (either previous or current smoker) was more frequent in patients with ECCA aneurysms (53.3%).

The mean ± SD maximal diameter of the aneurysms was 23.8 ± 14.1 mm, with more than half of patients having aneurysms larger than 25 mm (52.6%). According to Attigah classification, there were 7, 8, and 4 patients with types I, II, and III-VI, respectively. 79.0% aneurysm had intraluminal thrombus at admission.

Treatment details

Table 3 shows treatment details. Six patients underwent successful neurosurgical resection for a total of seven ECCAs. In one patient, bilateral giant aneurysms were treated in stages with neurosurgical resection and end-to-end anastomosis (refer to Typical Case 1 for further details). Of the six patients who underwent end-to-end anastomosis, one patient received internal-external

Table 1 Summary of clinical features of 15 patients with ECCA on admission

Case No.	Gender	Age (year)	Etiology	Site	Maximum diameter (mm)	Attigah classification	Presenting symptoms
1	Female	69	Atherosclerotic	Bilateral	Right 34.0 Left 40.0	II	Pulsatile neck mass
2	Male	71	Infectious	Right	21.0	III	Pulsatile neck mass
3	Male	40	Infectious	Left	19.0	III	Pulsatile neck mass
4	Male	31	Unkown	Left	21.0	I	Cerebral infarction
5	Female	40	Atherosclerotic	Left	50.0	II	Pulsatile neck mass
6	Female	30	Atherosclerotic	Left	27.0	II	Pulsatile neck mass
7	Female	63	Atherosclerotic	Right	36.0	II	Pulsatile neck mass
8	Male	69	Atherosclerotic	Right	25.0	II	Pulsatile neck mass
9	Male	69	Infectious	Left	34.0	III	Pulsatile neck mass with ulceration of skin an oozing blood
10	Male	38	Iatrogenic	Right	47.0	IV	Pulsatile neck mass
11	Female	59	Atherosclerotic	Right	35.0	II	Pulsatile neck mass
12	Female	51	Unkown	Bilateral	Right 6.0 and 2.0 Left 6.0	I	Asymptomatic
13	Female	53	Unkown	Right	19.0	I	TIA
14	Male	47	Dissection	Right	8.8 and 15.0	I	Asymptomatic
15	Male	74	Unkown	Left	6.0	II	Incident

Table 2 Baseline patient and aneurysm characteristics

Characteristic	Frequency
Patients	***n* = 15**
Age (mean ± SD)	53.6 ± 14.8
Female	7 (46.7%)
Presenting symptoms	
Pulsatile neck mass	10 (66.7%)
Asymptomatic	3 (20.0%)
Stroke or TIA	2 (13.3%)
Etiology	
Atherosclerotic	6 (40.0%)
Dissecting	1 (6.7%)
Iatrogenic	1 (6.7%)
Infectious	3 (20.0%)
Unkown	4 (26.6%)
Comorbidity	
Hypertension	5 (33.3%)
Diabetes	1 (6.7%)
Hyperlipidemia	8 (53.3%)
Cerebral infarction	1 (6.7%)
Coronary heart disease	1 (6.7%)
Smoking	
Never	8 (53.3%)
Previous	1 (6.7%)
Current	6 (40.0%)
Alcohol abuse	
Never	10 (66.7%)
Previous	1 (6.7%)
Current	4 (26.6%)
Multiple aneurysms	3 (20.0%)
Aneurysms	*n* = 19
Aneurysm size (max length, mm)	
Mean (SD)	23.8 ± 14.1
< 10 mm	4 (21.1%)
10–25 mm	5 (26.3%)
> 25 mm	10 (52.6%)
Parent artery diameter	4.7 ± 1.1
Attigah classification	
I	7 (36.8%)
II	8 (42.1%)
III-IV	4 (21.1%)
Intra-aneurysmal thrombus	15 (79.0%)

Data are *n* (%) or mean ± SD (range)

carotid anastomosis. Intraoperative indocyanine angiography confirmed that the aneurysm was obliterated and the bypass was patent.

Endovascular treatment proved successful for five patients in this study. Two of the patients had type I ECCA and were treated with a covered stent and Pipeline employment, respectively. Two other patients with type III ECCA received covered stent placement. The final patient had type IV iatrogenic ECCA and underwent internal carotid artery occlusion. Furthermore, two patients with infectious ECCAs received anti-infective treatment following implantation of the Willis stent (refer to Typical Case 2).

Table 3 Treatment details, clinical and angiographic outcome

Characteristic	Frequency (*n* = 19)
Treatment methods	
Neurosurgical treatment	
End to end anastomosis	6 (31.6%)
External carotid - internal carotid anastomosis	1 (5.3%)
Endovascular treatment	
Occlusion	1 (5.3%)
Pipeline Embolization Device	1 (5.3%)
Willis	3 (15.8%)
Conservative treatment	7 (36.8%)
Anti-inflammation therapy	3 (15.8%)
Angiographic outcomes	*n* = 19
Mean follow-up time (months)	28.1 ± 18.5
Satisfactory occlusion after operation at last follow-up	
Complete occlusion	10/11 (90.9%)
Neurosurgical treatment	6/7 (85.7%)
Endovascular treatment	4/4 (100%)
Incomplete occlusion (Neurosurgical treatment)	1/11 (9.1%)
Aneurysm after conservative treatment at last follow-up	
Disappeared	3/7 (42.9%)
No changes	4/7 (57.1%)
Satisfactory status of parent arteries at follow-up	
Patency	15 (88.2%)
Stenosis	1 (5.3%)
Occlusion	1 (5.3%)
Clinical outcomes	*n* = 15
Ischemic stroke	1 (6.7%)
Hemorrhage	0
Total complication	1 (6.7%)
Mortality	1 (6.7%)

Data is *n* (%)

The remaining four patients who presented with seven ECCAs were placed under observation. Among them, one patient with type III infective ECCA was administered antibiotics, and the aneurysm subsequently disappeared after an 18-month follow-up period (refer to Typical Case 3 for further details). In addition, one patient (case 12) had bilateral ECCAs, and the aneurysms at both sites were type I.

Clinical and angiographic outcomes

The mean follow-up period was 28.1 months (Table 3). Out of the patients who received treatment, it was discovered that 10 of them had completely occluded at the latest imaging study. Additionally, only one patient who was treated with a PED still had an aneurysm that could be seen during the 18-month follow-up period. During the course of conservative observation, it was observed that in one patient, the aneurysm disappeared and the parent vessel became thinner following anti-infection treatment. Furthermore, in two cases of dissecting aneurysms, the aneurysm disappeared during the follow-up period after receiving anti-platelet drug therapy. However, there were no significant changes observed in the other ECCAs during the follow-up period. Out of all the patients, only one who was treated with Willis stent experienced a large area of cerebral infarction after treatment, which ultimately resulted in their death.

Illustrative cases

Typical case 1

A 69-year-old female with a medical history of hypertension, diabetes, and hyperlipidemia presented with a 1-year history of a pulsatile neck mass and neck pain (Fig. 1). Upon examination, bilateral pulsatile neck masses were observed (Fig. 1A, B), but the patient was neurologically intact. Further investigation through a

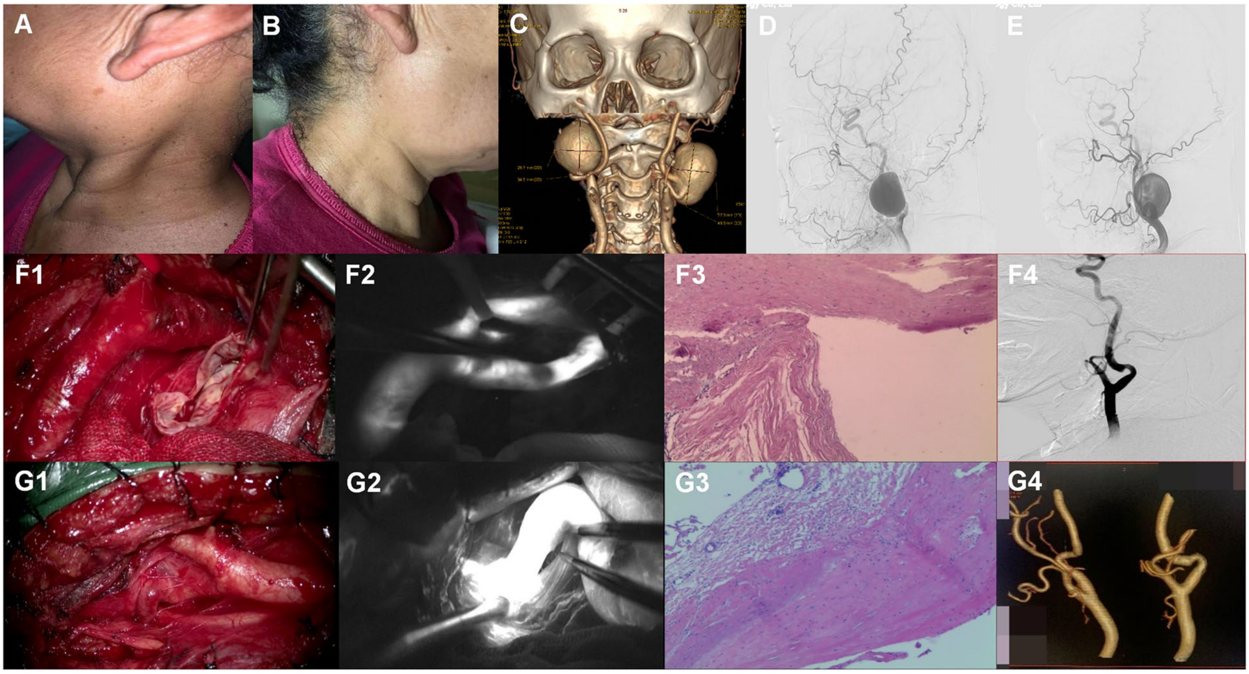

Fig. 1 Typical case 1; A 69-year-old female with a medical history of hypertension, diabetes, and hyperlipidemia presented with a 1-year history of a pulsatile neck mass and neck pain. Upon examination, bilateral pulsatile neck masses were observed (**A**-**B**), but the patient was neurologically intact. Further investigation through a CTA examination revealed an unruptured giant bilateral ECCA aneurysm (**C**). Cerebral angiography showed bilateral giant aneurysms of the extracranial internal carotid arteries (maximum diameter: 40.0 mm on the left and 34.0 mm on the right) (**D**-**E**). Firstly, the left ECCA aneurysm was surgically resected and underwent end-to-end anastomosis (**F1**). To ensure the patency of the parent artery, fluorescein angiography was performed during the operation (**F2**). The resected aneurysm specimen was also subjected to pathological examination (**F3**). Three days following the operation, angiography revealed complete disappearance of the aneurysm and smooth blood flow in the parent artery (**F4**). One year later, the patient underwent another neurosurgical resection and end-to-end anastomosis procedure to address the right aneurysm (**G1**). During this operation, fluorescein angiography was performed to verify the patency of the parent artery, and the resected aneurysm specimens were analyzed through pathological examination (**G2-3**). A year following the procedure, a CTA scan revealed that both aneurysms had vanished completely, and the blood flow in the parent artery was unobstructed (**G4**)

CTA examination revealed an unruptured giant bilateral ECCA aneurysm (Fig. 1C). Cerebral angiography showed bilateral giant aneurysms of the extracranial internal carotid arteries (maximum diameter: 40.0 mm on the left and 34.0 mm on the right) (Fig. 1D-E). For these patient, bilateral aneurysms were considered, and the aneurysms were large, close to the lower end of the carotid artery, and the vessels were tortuous. And if there are compression symptoms, we use end-to-end anastomosis. Firstly, the left ECCA aneurysm was surgically resected and underwent end-to-end anastomosis (Fig. 1F1). To ensure the patency of the parent artery, fluorescein angiography was performed during the operation (Fig. 1F2). The resected aneurysm specimen was also subjected to pathological examination (Fig. 1F3). Three days following the operation, angiography revealed complete disappearance of the aneurysm and smooth blood flow in the parent artery (Fig. 1F4). One year later, the patient underwent another neurosurgical resection and end-to-end anastomosis procedure to address the right aneurysm (Fig. 1G1). During this operation, fluorescein angiography was performed to verify the patency of the parent artery, and the resected aneurysm specimens were analyzed through pathological examination (Fig. 1G2-3). A year following the procedure, a CTA scan revealed that both aneurysms had vanished completely, and the blood flow in the parent artery was unobstructed (Fig. 1G4).

Typical case 2

A 71-year-old male with hyperlipidemia and a smoking habit of 1 pack per day presented with a pulastile neck mass that had been present for 4 months (Fig. 2). The mass was observed on the right side of the neck (Fig. 2A), and the patient was found to be neurologically intact upon examination. Further investigation through a CTA examination revealed an unruptured aneurysm in the right ECCA (Fig. 2B). The aneurysm was relatively small and the carotid artery was relatively straight, which was suitable for releasing the stent. After communicating with the patient's family, the Willis stent was used for treatment. Cerebral angiogram revealed a giant aneurysm (21.0 mm in maximum diameter) on the

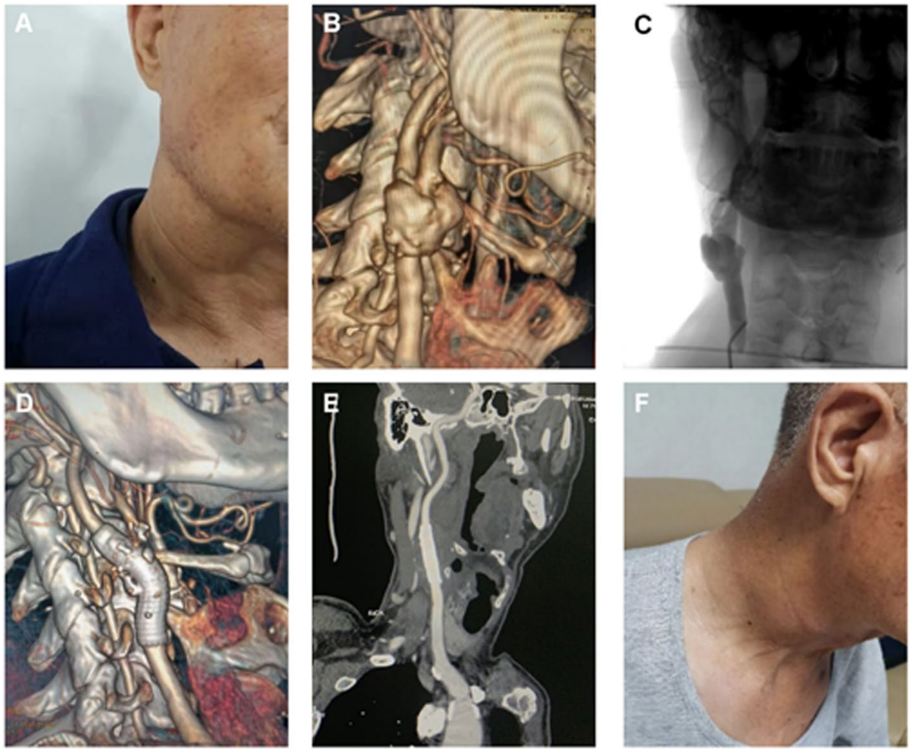

Fig. 2 Typical case 2; A 71-year-old male with hyperlipidemia and a smoking habit of 1 pack per day presented with a pulastile neck mass that had been present for 4 months. The mass was observed on the right side of the neck (**A**), and the patient was found to be neurologically intact upon examination. Further investigation through a CTA examination revealed an unruptured aneurysm in the right ECCA (**B**). Cerebral angiogram revealed a giant aneurysm (21.0 mm in maximum diameter) on the extracranial internal carotid artery (**C**), and was reconstructed with a 4.0·16 mm Willis (**D**). One month after the operation, the patient took oral anti-infective drugs. Upon re-examination angiography, it was found that the aneurysm had completely disappeared and the blood flow through the parent artery was smooth (**E**). Additionally, the pulsating mass in the neck had also disappeared (**F**)

extracranial internal carotid artery (Fig. 2C), and was reconstructed with a 4.0·16 mm Willis (Fig. 2D). One month after the operation, the patient took oral anti-infective drugs. Upon 4re-examination angiography, it was found that the aneurysm had completely disappeared and the blood flow through the parent artery was smooth (Fig. 2E). Additionally, the pulsating mass in the neck had also disappeared (Fig. 2F).

Typical case 3

A 40-year-old man who smokes one pack per day presented with a neck mass and low-grade fever that had persisted for a month (Fig. 3). Upon examination, the patient was found to be neurologically intact. A CTA examination was conducted, which revealed an unruptured aneurysm in the left ECCA (Fig. 3A-B). Further investigation through a cerebral angiogram revealed a large aneurysm (19.0 mm in maximum diameter) on the extracranial internal carotid artery (Fig. 3C). After an incisional neck biopsy and pathological examination, it was determined that the patient had an infected aneurysm (Fig. 3D-E). Anti-infection treatment was then administered, and a CTA re-examination was conducted one month later. The results showed that the aneurysm had disappeared and the parent artery had become thinner (Fig. 3F).

Discussion

ECCA is a rare disease with an unknown natural history. Previous reports have shown differences in incidence and demographics between men and women. Faggioli et al. reported that 65% of ECCA cases occurred in women, with a mean age of 54.8 ± 16.7 years [8]. On the other hand, Xu et al. found that incidence of EICA was three times higher in men than in women, with an average age of 61.9 years [9]. In this study, 53.3% of the participants were men, with an average age of 53.6 years. The most common presentation of ECCAs was a neck mass, along with arterial pseudoaneurysms that caused swelling and pain [10]. Neurological events, such as stroke or TIA,

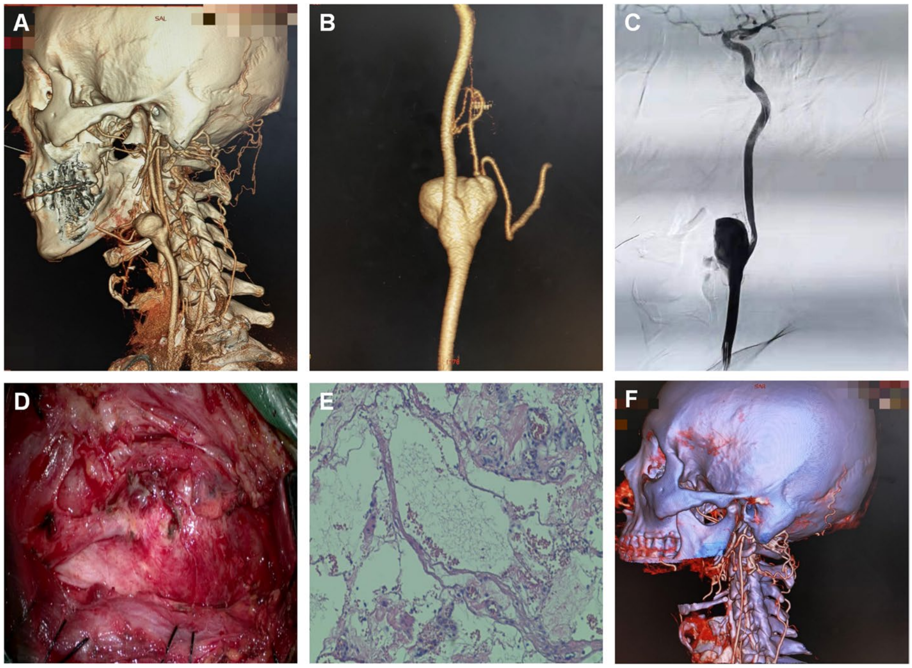

Fig. 3 Typical case 3; A 40-year-old man who smokes one pack per day presented with a neck mass and low-grade fever that had persisted for a month. Upon examination, the patient was found to be neurologically intact. A CTA examination was conducted, which revealed an unruptured aneurysm in the left ECCA (**A**-**B**). Further investigation through a cerebral angiogram revealed a large aneurysm (19.0 mm in maximum diameter) on the extracranial internal carotid artery (**C**). After an incisional neck biopsy and pathological examination, it was determined that the patient had an infected aneurysm (**D**-**E**). Anti-infection treatment was then administered, and a CTA re-examination was conducted one month later. The results showed that the aneurysm had disappeared and the parent artery had become thinner (**F**)

may be the first clinical findings caused by embolisms derived from thrombus in the aneurysm. If an aneurysm is located distally and is large, it can lead to dysfunction of postganglionic sympathetic nerve fibers and cranial nerves. This is due to compression or distension, which can cause swallowing difficulties, hoarseness, or even Horner syndrome [11–13].

Generally speaking, endovascular treatment is the preferred approach for infectious ECCAs in emergency situations. However, there are some experts who propose that patients with large or giant infectious carotid pseudoaneurysms that have been treated with stent placement may face heightened risks of persistent infection and eventual device failure [14–16]. In this study, three patients with infectious ECCAs were given antibiotics either after surgery or during the observation period. They were closely monitored and did not experience any recurrence of aneurysms or neck pain.

The management of ECAA can differ significantly due to its rarity and the lack of evidence-based recommendations for treatment [17]. Currently, management strategies consist of both conservative treatment and surgical intervention. Conservative treatment options primarily involve the use of anticoagulation and antiplatelet medications for true or dissection aneurysms, as well as antibiotic therapy for infectious aneurysms. A recent review found that conservative management of ECCAs results in a 4.67% mortality rate and 6.67% stroke rate [1]. However, favorable outcomes can also be achieved through nonsurgical treatment of patients with small, asymptomatic, dissecting aneurysms or pseudoaneurysms [18, 19]. In this study, four patients with true aneurysms, one with an infectious aneurysm, and two with a dissection aneurysm were observed, and no complications occurred during the follow-up period.

In 1805, Sir Astley Cooper successfully implemented the procedure of ECCA ligation, but unfortunately, the patient passed away 40 h after surgery [20]. As a result, ligation of the carotid artery became the only alternative approach for a significant period of time, despite a reported stroke risk of up to approximately 30% [10]. Today, surgical reconstructions and endovascular aneurysm exclusion are the preferred treatment options. Studies indicate that mortality and stroke rates can reach 50–71% if left untreated and occlusion or embolism occurs. While rupture is rare, it can still occur with small odds [21, 22]. Furthermore, Jin et al. have reported that even in the absence of clinical symptoms, surgical intervention should be considered if imaging examinations reveal any adverse impact on cerebral blood supply caused by ECCA [23]. Once a diagnosis has been made, it is important to consider active surgical intervention for ECCA. However, it is important to note that both strategies for ECCA carry certain risks and complexities. Previous reviews have reported that surgical management has a stroke rate of 0–1.91%, a death rate of 0–5.16% at 30 days, and a 7% rate of cranial nerve injuries [2, 6].

However, a recently reviewed endovascular treatment for ECAA reported a stroke rate of 1.8–2.5%, TIA rates of 4.3%, a cranial nerve injury rate of 0.5%, and an in-hospital mortality rate of 4.1% [24, 25]. These rates were lower than our results, which showed a mortality rate of 6.7% and a stroke rate of 6.7%. In terms of major outcomes, our findings indicate that open surgery is linked to lower rates of early mortality, stroke, and TIA. Interestingly, we observed similar rates for endovascular procedures.

Neurosurgical treatment for ECCAs has been in development for over 200 years, with the aim of preserving the brain from thromboembolism that could result in disabling stroke or even death [26]. Despite this long history, endovascular treatment is becoming more popular due to its minimally invasive nature, which can help avoid common complications like cranial nerve injury and hemorrhage that are often associated with open surgery. Altun G suggested that the Attigah classification can guide surgical options for ECCA patients [1]. However, some authors have argued that this classification is not appropriate for endovascular treatment [5]. It is worth noting that nerve injuries in ECCA patients are typically transient (11–22%) rather than permanent (3–13%). Additionally, type V ECCA may require special techniques such as sternotomy. Therefore, endovascular treatment is the best approach for managing type I and V ECCA.

A covered stent (Willis) or flow diverter (PED) would be a favorable option [27]. However, there are various treatment strategies for types II, III, and IV that remain controversial. Open surgical and endovascular approaches should be carefully selected for all three types, depending on the patient's specific condition, especially if the aneurysm is located near the bifurcation of the carotid artery, carotid artery bulb, or if the parent artery has tortuosity or is kinky with a large or giant ECCA. We concur with Davidovic et al.'s assertion that factors such as etiology, location, size, tortuosity, and morphology play a critical role in determining the most effective therapeutic approach [28]. In our own investigation, we conducted neurosurgery on six patients diagnosed with true ECCA type II. The results were overwhelmingly positive, with the exception of one patient who received a covered stent without proper consideration of tortuosity.

We also explored ligation as a neurosurgical strategy, particularly as a last resort in emergency situations such as aneurysm rupture or in the case of a particularly large aneurysm. Like ligation, carotid sacrifice through coiling can halt blood flow to the lesion, lowering the chances of recanalization and aneurysm recurrence. However, this method also raises the risk of ischemic injury. Therefore, it is crucial to perform temporary occlusion of the parent vessel to ensure collateral supply to vulnerable territories [29, 30].

Although this study offers valuable insights into the management of a rare disease, it is important to note its limitations. Firstly, the study is retrospective in nature and has a limited number of patients and events. Additionally, there is no control group. Despite these limitations, the study still provides valuable information on the current management of the disease.

Conclusions

ECCA is rare and mostly present with mass effect. Neurosurgical treatment was more frequently feasible in large ECCA, and endovascular surgery was the first choice for pseudoaneurysms and dissecting aneurysms. Anti-inflammation treatment was available for some infectious cases.

Supplementary Information

The online version contains supplementary material available at https://doi.org/10.1186/s12883-024-03970-z.

Supplementary Material 1

Supplementary Material 2

Acknowledgements

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Authors' contributions

AXiMuJiang AXiEr, Mirzat Turhon and Maimaitili Aisha conceived the research, drafted the manuscript, and statistical analysis. AXiMuJiang AXiEr, Mirzat Turhon, Xiaojiang Cheng, Zengliang Wang, and Maimaitili Aisha revised the manuscript and designed the study. Aierpati Maimaiti, Dilmurat Gheyret, Kaheerman Kadeer, Shihao JIang, Riqing Su, Nizamidingjiang Rexiati, Kai Wang performed the data collection. All authors read and approved the final manuscript.

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Data availability

No datasets were generated or analysed during the current study.

Declarations

Ethics approval and consent to participate

The Medical Ethics Committee of the First Affiliated Hospital of Xinjiang Medical University approved this study (approval number: XJMU 20171018-01). Written informed consent was obtained individually. We confirm that all methods in our study were performed in accordance with the relevant guidelines and regulations.

Consent for publication

Written informed consent was obtained from the patient for publication of this case report and any accompanying images.

Competing interests

The authors declare no competing interests.

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