

# Extracranial carotid artery aneurysm with myeloproliferative neoplastic cell invasion

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## ABSTRACT

The major causes of rare extracranial carotid artery aneurysms are arteriosclerosis, trauma, and radiation therapy. Here, we describe a patient with an extracranial carotid artery aneurysm caused by a myeloproliferative neoplasm. A 67-year-old woman underwent excision of an irregularly shaped aneurysm in the left common carotid artery and a saphenous vein graft without major complications. The pathologic findings revealed abscess formation and atypical megakaryocyte infiltration, which was also seen in her bone marrow, indicating that the aneurysm was caused by a myeloproliferative neoplasm. (*J Vasc Surg Cases and Innovative Techniques* 2020;6:243-6.)

**Keywords:** Extracranial carotid artery aneurysm; Myeloproliferative neoplasm; Blood disease; Aneurysmal excision; Bypass

Extracranial carotid artery aneurysms (ECAAs) are rare, accounting for <1% of all arterial aneurysms.<sup>1-3</sup> Development of an aneurysmal rupture in ECAAs is rare. When symptoms occur, they are mostly due to embolism from intra-aneurysm thrombus or nerve compression resulting in dysphagia and hoarseness.<sup>4-7</sup> The main causes of ECAAs have previously been local infection and syphilis,<sup>8</sup> whereas the rates of arteriosclerosis, trauma, and radiation therapy have recently increased.<sup>1,5,9</sup> Thus far, a relationship between ECAAs and blood disease, including myeloproliferative neoplasm (MPN), has not been reported. We describe a rare ECAA with MPN cell invasion that was surgically treated with a good outcome. The patient provided consent for publication of this case report.

## CASE REPORT

A 67-year-old woman presented with a 4-month history of swelling in the left side of her neck. A physical examination revealed an egg-sized pulsatile mass, and enhanced computed tomography revealed a large ECAA. The mass gradually grew without any infectious signs, such as redness on the skin or pain. She had no history of neck surgery, radiation therapy, hypertension, diabetes mellitus, or dyslipidemia. She did have a history of MPN that was incidentally diagnosed 27 years previously. Her MPN was asymptomatic and negative for the JAK2V617 F mutation, so she received no treatment. Other than the cervical mass, her condition was stable. Blood findings

at the time of admission were white blood cell count of  $29.8 \times 10^3/\mu\text{L}$ , platelet count of  $603 \times 10^3/\mu\text{L}$ , and hemoglobin level of 9.7 g/dL, indicating mild anemia. C-reactive protein concentration was 0.7 mg/dL. Echocardiography did not indicate infections endocarditis, and blood culture was negative even though she had no history of antibiotic treatment.

Cervical ultrasound revealed a mass that was consistent with a partially thrombosed carotid artery aneurysm. Angiography (three-dimensional rotational angiography) showed a large, irregularly shaped aneurysm at the left common carotid artery (CCA) bifurcation measuring  $47 \times 45 \times 35$  mm. The aneurysm was mainly positioned at the level of the C4 vertebra and extended to C2. The origins of the internal carotid artery (ICA) and the external carotid artery (ECA) were totally engulfed by the aneurysm. Brain magnetic resonance imaging did not find any evidence of an aneurysm-related ischemic lesion in the left hemisphere (Fig 1).

The aneurysm was basically asymptomatic without discomfort, but it had continued to grow, and intra-aneurysmal thrombus was considered a risk factor for artery-to-artery embolism. Moreover, the patient was worried about the pulsatile mass on her neck and finally agreed to undergo treatment. To check the pathologic features of the aneurysm and to relieve the mass effect on her neck, direct surgery was thought to be preferred to endovascular treatment in this case.

We therefore decided to excise the aneurysm and to reconstruct blood flow to the ICA using a great saphenous vein graft between the CCA and the ICA. Under general anesthesia with motor and sensory evoked potential monitoring, a curvilinear skin incision was placed just above the aneurysm and along the sternocleidomastoid muscle. The aneurysm and its branches (CCA, ICA, and ECA) were then fully exposed. The origin of the ICA from the CCA was not high, so no special procedures, such as mandibular subluxation, were required. The aneurysm was tightly adherent to surrounding tissues, but it was totally dissected and excised en bloc. After grafting of the CCA and ICA, the ECA was ligated at its origin. An internal shunt was considered unnecessary on the basis of motor and sensory evoked potential monitoring (Fig 2). Graft patency was confirmed using indocyanine green video angiography and

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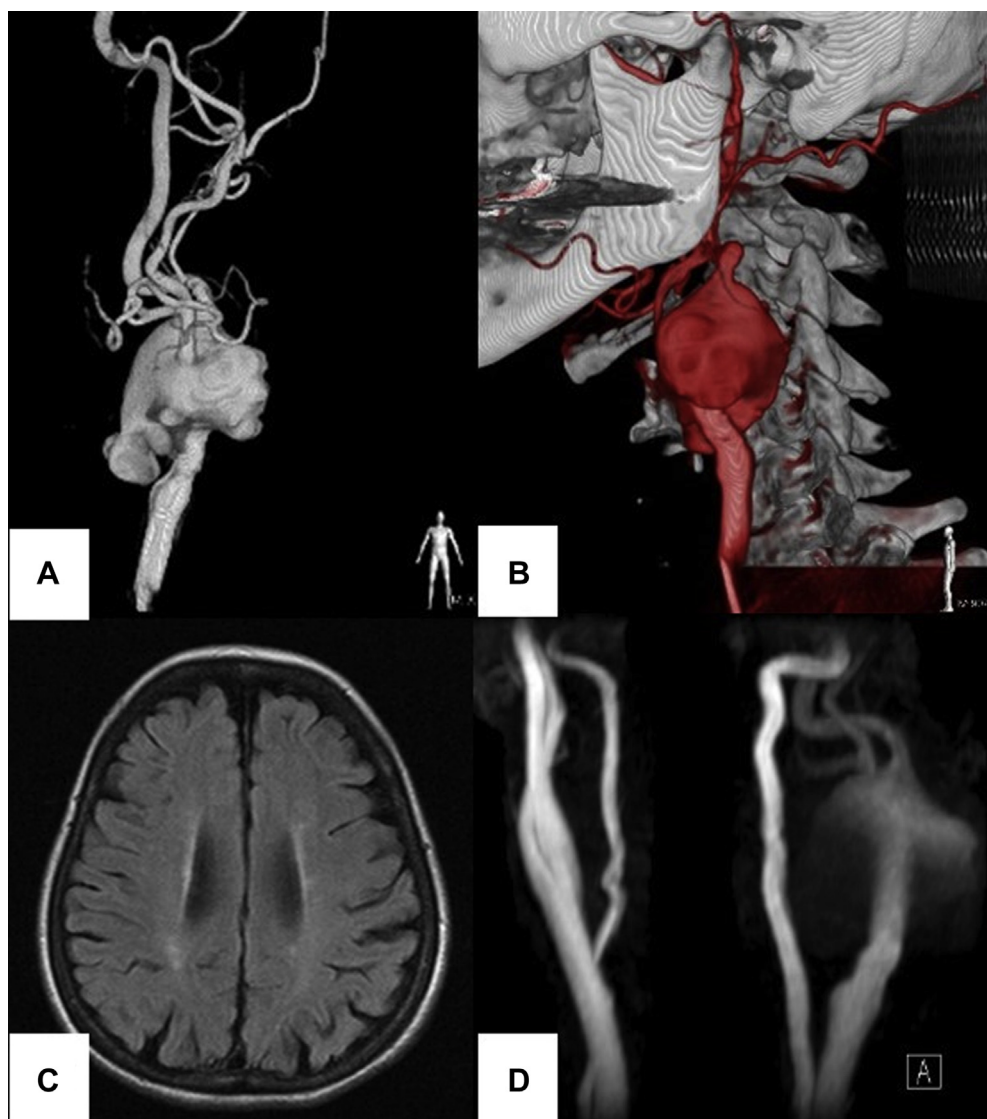
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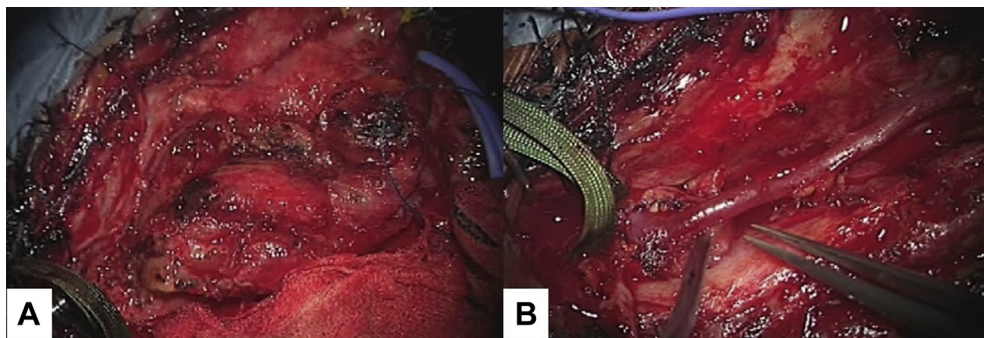


**Fig 1.** **A**, Angiography shows large, irregularly shaped aneurysm in the left common carotid artery (CCA) bifurcation, measuring  $47 \times 45 \times 35$  mm. The internal carotid artery (ICA) and external carotid artery (ECA) branch off from the top and frontal aspect of the aneurysmal dome. **B**, The aneurysm extends from C3 to C5, and its distal edge is 15 mm below the mandibular angle. **C**, Brain diffusion-weighted magnetic resonance image shows no evidence of prior embolic episodes caused by the aneurysm. **D**, Neck magnetic resonance angiography shows a balloon-shaped CCA.

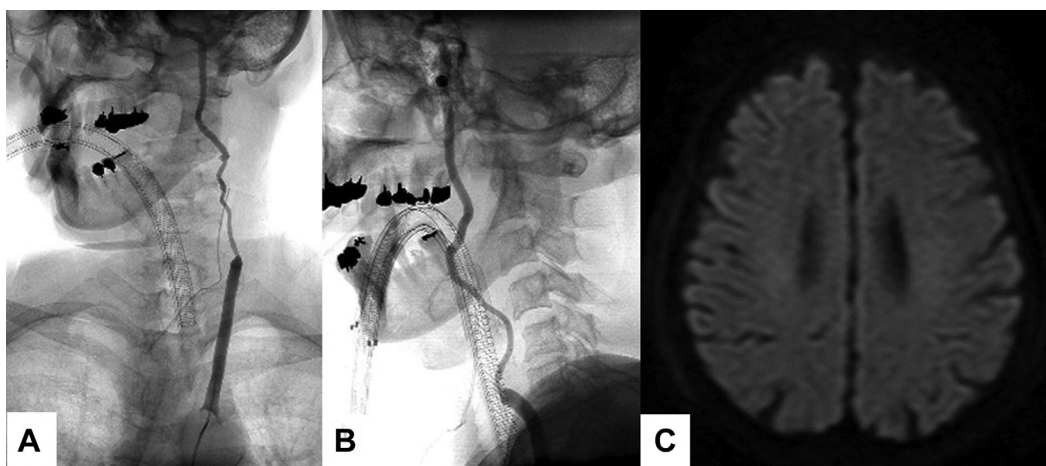
Doppler flowmetry. Postoperative angiography confirmed graft patency, and magnetic resonance imaging did not show any ischemic complications (Fig 3). The patient initially developed slight hoarseness and dysphagia but gradually recovered with rehabilitation, and she returned home 31 days after surgery with a modified Rankin scale score of 1. The pathologic diagnosis was an aneurysm with an abscess and atypical megakaryocyte infiltration. Granulation and ulceration were found within the aneurysm, and invasive megakaryocytes were morphologically abnormal. Similar megakaryocytes were found in her bone marrow (Fig 4). According to the negative margin of the specimens and no findings of systemic inflammation, we used antibiotics only for prophylaxis.

## DISCUSSION

ECAAs are rare and have been found in the ICA (49.1%), CCA (48.1%), and ECA (2.2%).<sup>10</sup> Asymptomatic aneurysms are being diagnosed more frequently because of the increasing use of cross-sectional imaging.<sup>1</sup> Most patients with ECAAs present with symptoms of central neurologic dysfunction, pulsatile masses, or local compression symptoms, such as dysphasia and hoarseness.<sup>7</sup> Moreau et al<sup>2</sup> reported that cerebral embolization was found in 74% of patients with ECAAs, and more recent reports indicate that 42% to 50.9% of patients with ECAAs also have neurologic symptoms, such as stroke and transient ischemic attack.<sup>4,7,11</sup> Neurologic accidents are typically



**Fig 2.** **A**, The left common carotid artery (CCA) is secured on the proximal side and the internal carotid artery (ICA) and external carotid artery (ECA) on the distal side of the aneurysm that is tightly adherent to the surrounding tissues, but abscess had not formed around the vessels. **B**, Carotid arteries are reconstructed with a saphenous vein graft and end-to-end ICA and CCA anastomosis on both sides and ligation of the ECA on the distal side of the aneurysm.



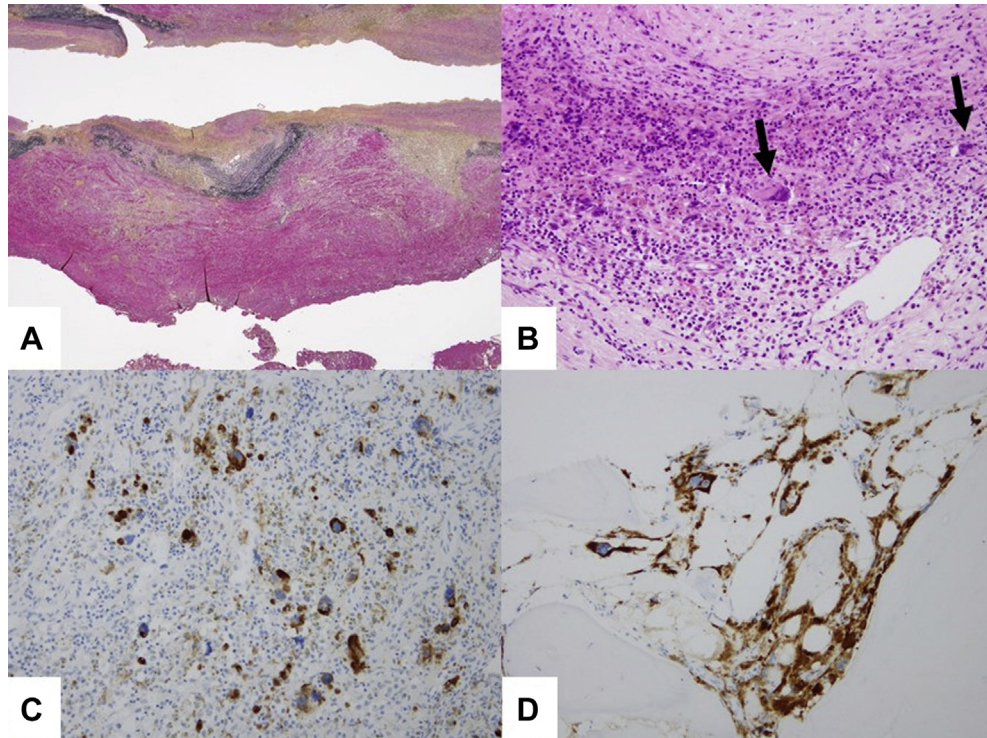
**Fig 3.** **A** and **B**, Angiography confirms graft patency under general anesthesia 1 day after surgery. **C**, Cerebral infarction is undetectable on magnetic resonance imaging 1 day after surgery.

due to thrombus embolization from aneurysms. According to Li et al,<sup>4</sup> thrombus formation was found in 18.4% of aneurysms in a series of 224 patients, and others have found local compression symptoms in 10% to 37% of patients.<sup>6,7</sup> Patients presenting with rupture or impending rupture accounted for <3% of all diagnosed patients.<sup>6</sup>

ECAAs can arise from many causes, but the most common is atherosclerosis in 26% to 50% of all patients, and it is also the major cause of ECAAs in older patients.<sup>5,12</sup> Trauma has accounted for 5% to 51% of aneurysms in the past,<sup>3,4</sup> whereas pseudoaneurysms after carotid endarterectomy have more recently accounted for 26% to 57%.<sup>6,7</sup> Less frequent causes include bacterial infection, fibromuscular dysplasia, collagen vascular diseases such as Ehlers-Danlos syndrome, and Marfan syndrome.<sup>13</sup> Treatment of ECAA includes medical treatment, surgery, and endovascular treatment, and it depends mainly on the cause of the ECAA. For the ECAA that is not suggestive of infection, endovascular treatment with covered stent is a promising treatment option; but in this case,

it was considered that MPN was associated with the cause of aneurysm formation because there were no findings suggestive of atherosclerosis, history of trauma, or cervical surgery. Therefore, direct surgery was thought to be preferred because pathologic diagnosis and mass reduction could be achieved at the same time.

We found some atypical megakaryocytes in a surgical specimen and in a bone marrow sample, indicating a relationship between the aneurysm and MPN. MPN represents several types of blood disorders, all characterized by the proliferation of cells of one or more myeloid lineages. Although ECAA, especially infectious aneurysm, can occur at a higher frequency in immunocompromised patients,<sup>8</sup> our patient did not show it in the clinical condition or in laboratory data. Furthermore, the aneurysm showed a gradual increase in size during 4 months without acute change, and a sign of infection, such as fever, redness, or local pain, was not observed with negative blood culture. Although surgical specimens included abscess and the possibility of infectious



**Fig 4.** **A**, Elastic fiber destruction in the arterial wall. Elastica-van Gieson stain (magnification  $\times 20$ ). **B**, Hematoxylin-eosin stain (magnification  $\times 200$ ) shows inflammatory cell infiltration and megakaryocytes with morphologic abnormalities (arrows). **C**, Abnormal megakaryocytes are CD42b positive. **D**, Similar CD42b-positive abnormal megakaryocytes in bone marrow biopsy specimen.

aneurysm as its origin could not be excluded, we suspected that the aneurysm was caused by MPN cell invasion rather than by local infection. Leukemia-induced aneurysm was reported in the lung arteries but not in ECAA.<sup>14</sup> We described the first instance of an ECAA that was closely associated with a blood disease.

## CONCLUSIONS

We described a rare ECAA with a megakaryocytic invasion in the context of MPN. As far as we can ascertain, this is the first demonstration of MPN-associated ECAA.

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