Myopericytoma of the axillary artery in a 4-year-old girl presenting with stroke and subsequent progression of micro-isolated lesion

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ABSTRACT

A 4-year-old girl presented with complaints of transient speech disorder and left-sided weakness. Magnetic resonance imaging revealed multiple strokes, computed tomography showed a right axillary artery aneurysm of 40 mm, and an angiogram indicated retrograde embolism. She underwent aneurysm resection and reconstruction with a saphenous vein graft. Two years after surgery, a mass lesion occurred in the native artery on the proximal side of the prior surgical anastomosis. Surgical resection and reconstruction were performed. An isolated 1-mm diameter lesion was found in the retrospectively reviewed completion angiogram from the initial surgery, which was the origin of the subsequent progressive lesion. The pathological examination, including after the initial surgery, revealed a myopericytoma. (J Vasc Surg Cases Innov Tech 2024;10:101493.)

Keywords: Axillary artery aneurysm; Myopericytoma; Stroke

Myopericytoma (MPC) is an uncommon benign tumor.[1] It is a distinctive perivascular myoid neoplasm that forms a morphological spectrum with myofibroma. MPC was first described in 1998 as a slow-growing, well-circumscribed nodule of ≤2 cm, affecting mostly the skin and superficial soft tissues of the distal extremities in adults. It was officially recognized by the World Health Organization in 2002.[2] MPCs occurring in the arterial wall are extremely rare.

We present a case of MPC in the right axillary artery of a 4-year-old girl who eventually required a second operation for progression of a micro-isolated lesion. The patient's parents provided written informed consent for the report of their daughter's case details and imaging studies.

CASE REPORT

A 4-year-old girl presented to a previous hospital with complaints of transient speech disorder and left-sided weakness of 3 hours' duration. The abnormal neurological findings had spontaneously resolved at the time of the visit. At her initial presentation, she was of standard build, 110 cm tall, and weighed 21 kg, with no pertinent medical history or history of trauma. Blood test results showed no evidence of sickle cell disease and no abnormalities of coagulation factors or thyroid function. Moreover, all tests for autoantibodies were negative, tests for metabolic diseases were negative, and genetic testing for Ehlers-Danlos syndrome was negative. An electrocardiogram showed no evidence of any arrhythmia, and an echocardiogram showed no intracardiac shunts, thrombus, or vegetation. Magnetic resonance imaging revealed multiple strokes, predominantly in the right hemisphere (Fig 1), and computed tomography (CT) revealed a multifocal tumor of 40 mm in the right axillary artery (Fig 2, *A*) and a bovine-type aortic arch. An angiogram indicated an axillary artery aneurysm with stagnation of contrast medium; retrograde blood flow from the axillary artery aneurysm flowed into the brachiocephalic artery and subsequently into the carotid artery during diastole (Supplementary Video, online only). The mechanism of the stroke was considered to be retrograde embolism, with thrombus originating from the aneurysm. In the acute phase of stroke, a neuroprotective agent was administered, and anticoagulation and antiplatelet therapy were given to prevent recurrence. No abnormal neurological findings were observed during conservative treatment and close examination of the cause of the stroke at the previous hospital, >4 months after the onset of symptoms. She was referred to our hospital to seek definitive treatment of the right axillary artery aneurysm, which was performed 6 months after the initial onset.

Preoperative findings showed that a multifocal aneurysm extended from the axillary artery to the brachial artery with little adhesion to the surrounding tissue (Fig 2, *B*). The patient underwent surgical resection of the aneurysm and interposition bypass using a great saphenous vein graft, connecting the axillary artery to the brachial artery via a transaxillary approach

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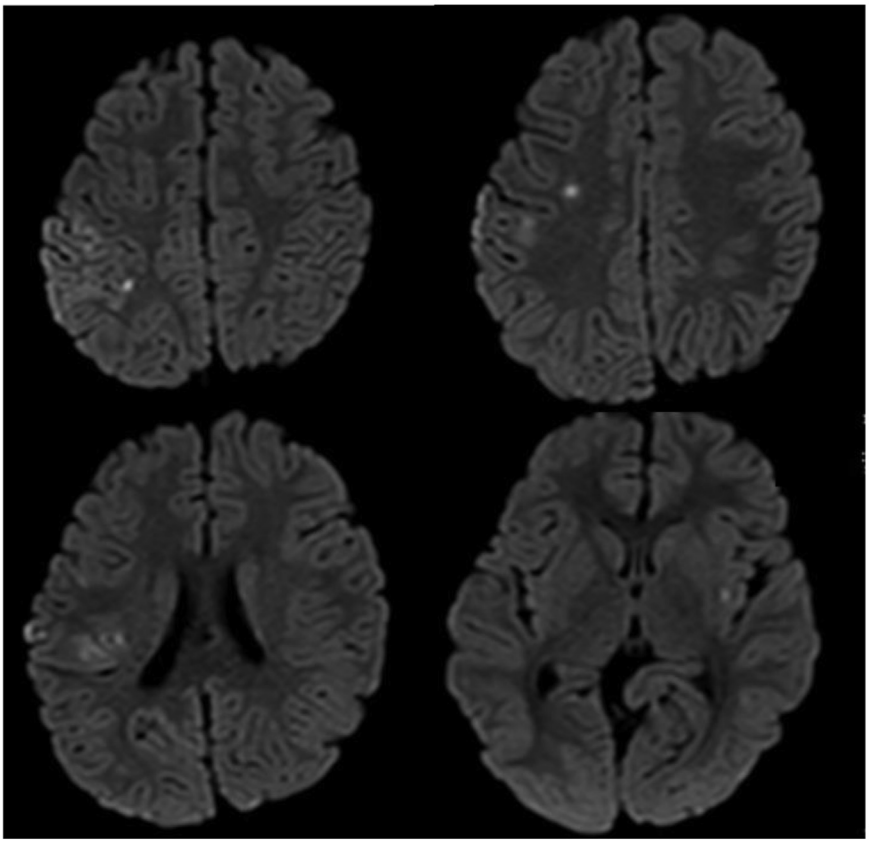

Fig 1. Magnetic resonance image showing multiple strokes (high signal) on diffusion-weighted imaging.

(Fig 2, *C* and *D*). The postoperative course was uneventful, and the patient was discharged from the hospital on postoperative day 6.

The resected specimen was submitted for pathological examination and measured 60 × 20 × 25 mm (Fig 3, *A*). Macroscopically, the artery was dilated at three locations in a beaded, saccular fashion. Histopathologically, the three-layer structure of the vessel wall (ie, intima, tunica media, and adventitia) was indistinct, revealing irregular wall thickening (Fig 3, *B*). In a part of the aneurysmal wall, dense proliferation of round and short spindle-shaped cells was observed, accompanied by slit-like capillaries (Fig 3, *C*). Some areas exhibited hemangiopericytoma-like morphology. The concentric perivascular growth of tumor cells presented only to a minimal extent. Mainly at the periphery of the lesion, the tumor cells were observed to proliferate, either around the small blood vessels or bulging into their lumens (Fig 3, *D*). There was no severe atypia suggestive of malignancy, although there were a few mitoses (Fig 3, *E*). The infiltration of these cells appeared to have disrupted the normal vascular wall structure, causing reactive intimal hyperplasia. Immunohistochemically, the tumor cells were positive for α-smooth muscle actin (Fig 3, *F*), partially positive for caldesmon, and negative for desmin, STAT6, MDM2, and CDK4. Consequently, the diagnosis was an MPC originating from the vascular wall and demonstrating destruction and aneurysmal dilatation of the muscular artery.

A follow-up ultrasound scan 2 years after the initial surgery confirmed a 36-mm multifocal aneurysm at the previous site. Initially, the aneurysm was thought to be secondary to great saphenous vein dilation; therefore, immediate surgical treatment was not performed. We elected to wait for the patient to grow until a prosthetic graft could be used. The aneurysm showed a tendency toward growth, measuring 43 mm at 3 years and 45 mm at 4 years after the initial surgery (Fig 4, *A*). At 4.5 years after the initial surgery, the patient presented with pain in the right axilla, and the diagnosis of impending rupture forced us to perform urgent surgery to treat the aneurysm. Using the same skin incision as for the initial surgery, an aneurysm was identified proximally at the prior anastomotic site that protruded on the dorsal side in a saccular shape (Fig 4, *B*). The patient underwent surgical resection of the aneurysm and reconstruction with an end-to-end anastomosis without the use of a graft (Fig 4, *C*). Pathological examination of the resected aneurysmal wall revealed the same diagnosis as that after the initial surgery. One year has passed after the reoperation without evidence of neuropathy, growth disorder of the right upper extremity, or recurrence.

DISCUSSION

We present a rare case of MPC located within the axillary artery wall. We report the histopathological characteristics and mechanisms of stroke and highlight the

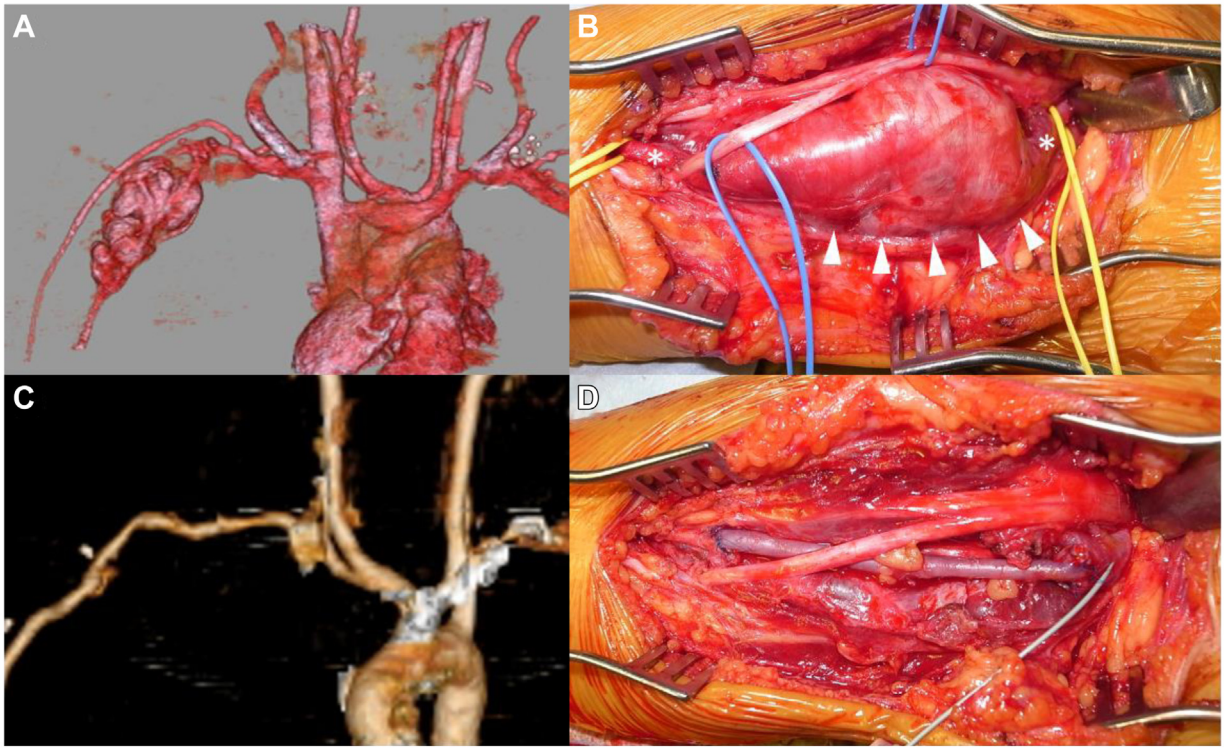

Fig 2. A, Three-dimensional reconstruction of computed tomography angiogram of a multifocal, 40-mm, right axillary artery aneurysm (AxAA). **B,** The ventral aspect of the AxAA was exposed (*arrowheads*), and each of the proximal and distal ends of the aneurysm were taped (*asterisks*). **C,** Three-dimensional reconstruction of follow-up computed tomography angiogram 1 month after surgery showing absence of the AxAA and favorable blood flow without anastomotic stenosis. **D,** Aneurysmectomy and reconstruction with a saphenous vein graft.

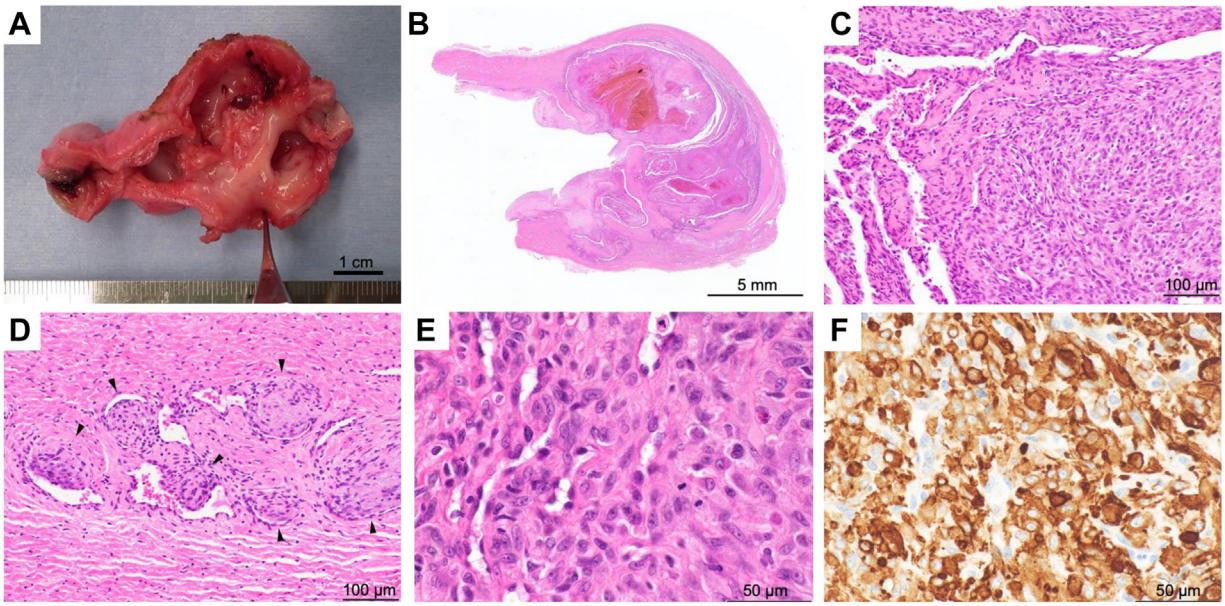

Fig 3. Pathological findings of the surgical specimen. **A,** Macroscopic image showing bead-shaped cystic dilation of the artery. **B,** Irregular aneurysmal wall thickening with vascular structure destruction at scanning magnification (hematoxylin and eosin [HE] stain). **C,** Dense proliferation of round to short spindle-shaped cells accompanied by slit-like capillaries (HE stain). **D,** Tumor cell proliferations bulging into the lumen of small vessels (*arrowheads*; (HE stain). **E,** A few mitoses can be observed without severe nuclear atypia (HE stain). **F,** Positive expression for α-smooth muscle actin of tumor cells.

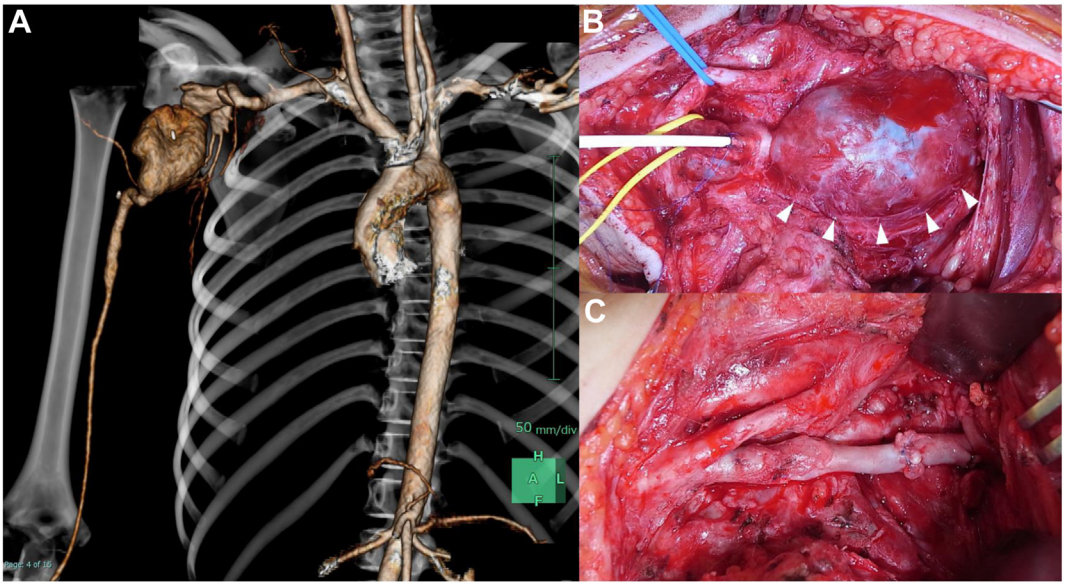

Fig 4. A, Three-dimensional reconstruction of follow-up computed tomography angiogram 4.5 years after initial surgery showing a multifocal 45-mm right axillary artery aneurysm (AxAA). **B,** The ventral aspect of the AxAA was exposed (*arrowheads*), and a sheath was inserted from the artery distal to the aneurysm. **C,** Aneurysmectomy and reconstruction with an end-to-end anastomosis of the proximal axillary artery and saphenous vein graft used in the initial surgery.

role of preoperative angiography in recognizing isolated lesions.

MPC is a rare tumor that affects individuals of all ages, with a peak after 50 years.[1] The most common location is in the lower extremities, particularly in the soft subcutaneous tissues, and rarely occurs in other sites. However, to the best of our knowledge, no studies have reported MPC occurring in the arterial wall, as observed in this case. A clinically similar pathology to MPC occurring in the vascular system is intravascular MPC (IVMP). IVMP, first described by McMenamin and Calonje[3] in 2002, is a rare subset of MPC in which the tumor is contained within the lumen of a vein, with or without invasion into surrounding structures. It has the same histological characteristics as MPC and presents as a distinct intravenous solid mass occupying a preexisting distended large vascular lumen.[4,5] Although most MPCs are rarely painful, IVMP typically manifests as a painful subcutaneous lump.[6] The intravascular position of the tumor and the formation of a thrombus might contribute to the pain.[7] The patient in this case, before the second surgery, experienced pain, which was thought to be associated with the impending rupture. The present case differs significantly from previously reported IVMPs in that it occurred in the arterial wall and exhibited intramural growth rather than luminal occupation.

The clinical course of this case was characterized by the onset of stroke, which also led to the diagnosis. There are several reports of stroke due to retrograde embolism caused by subclavian or axillary artery aneurysms or thoracic outlet syndrome, resulting in the release of a mural thrombus.[8-11] In our case, the preoperative angiogram clearly showed stagnation of contrast medium inside the aneurysm, the presence of a mural thrombus, and retrograde flow to the common carotid artery during the diastolic phase, which was thought to be the cause of the stroke. In addition, because of the bovine-type of aortic arch, a thrombus extending into the brachiocephalic artery caused stroke in both hemispheres. Fibromuscular dysplasia occurring in intracranial arteries is also included in the differential diagnosis as a cause of stroke in children.[12] In this case, both CT angiography and magnetic resonance angiography revealed no lesions suggestive of fibromuscular dysplasia in the carotid or intracranial arteries.

To the best of our knowledge, no studies have reported a micro-isolated lesion of MPC occurring in an arterial wall. Two years after our patient's initial surgery, a mass formation was observed in the native artery on the proximal side of the anastomosis, resulting in progression of a micro-isolated lesion. A retrospectively reviewed initial angiogram of the initial surgery revealed a 1-mm micro-isolated lesion approximately 3 cm proximal to the main lesion that remained slightly proximal to the anastomosis on completion angiography (Fig 5, *A* and *B*). The initial angiogram of the second surgery showed that the lesion was located at the same site as this residual isolated lesion identified on the final

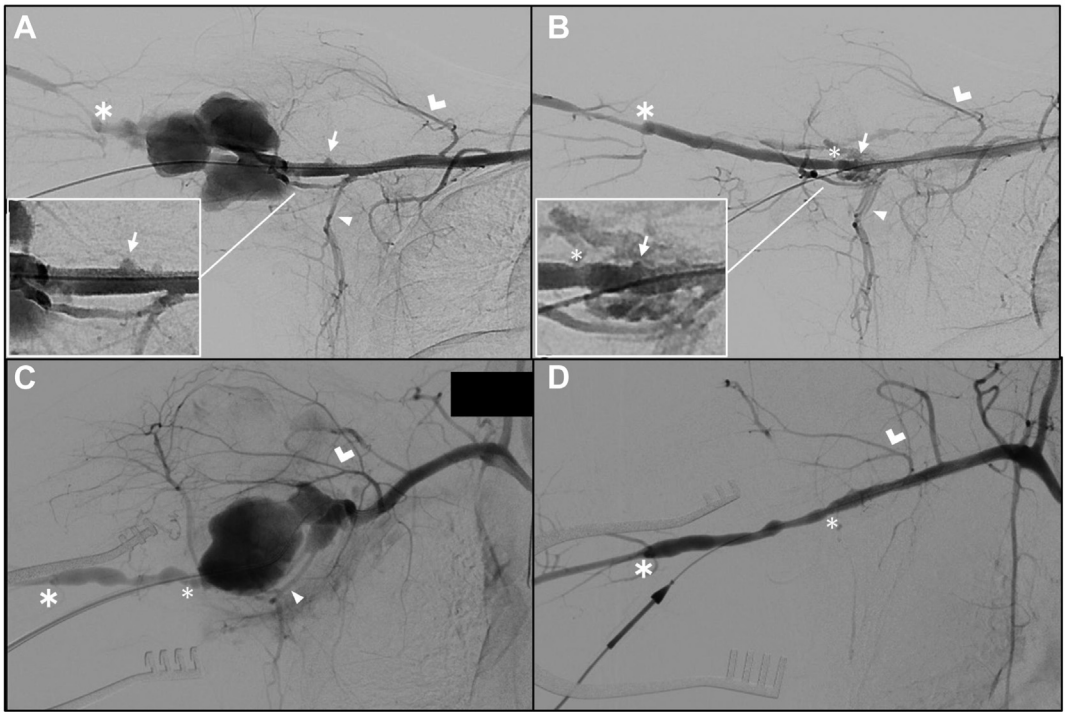

Fig 5. Location of the proximal anastomosis (*asterisks*), distal anastomosis (*large asterisks*), lateral thoracic artery (*arrowheads*), and deltoid branch (*large arrowheads*) indicated in each image. **A,** Preoperative angiogram of the initial surgery showing an isolated lesion (*arrow*) away from the main lesion. **B,** Postoperative angiogram of the initial surgery showing a residual isolated lesion (*arrow*) on the proximal side of the anastomosis (*arrowhead*). **C,** Preoperative angiogram of the secondary surgery showing a multifocal right axillary artery aneurysm on the proximal side of the prior anastomosis (*asterisk*). **D,** Postoperative angiogram of the secondary surgery with no visible residual isolated lesion on the proximal side of the new anastomosis (*asterisk*).

angiogram of the initial surgery (Fig 5, *C* and *D*). Also, the pathological findings were compatible with progression of a micro-isolated lesion. This isolated lesion could not be recognized on ultrasound or CT before to the initial surgery, underscoring the importance of a cautious assessment of the preoperative angiogram with the possibility of isolated MPC lesions. We assert that angiography is more effective than CT as a modality for detecting microscopic lesions in cases of nonatherosclerotic arteries, such as in the present case. In addition to its utility in the diagnosis of MPC, preoperative angiography is recommended for diagnosing and evaluating the localization of lesions in other peripheral aneurysms.[13-15]

CONCLUSIONS

MPC occurring in the arterial wall is extremely rare. Preoperative angiography is recommended for peripheral aneurysms; our case especially underscores the importance of careful interpretation of the preoperative angiogram and other imaging studies because of the possibility of isolated MPC lesions.

DISCLOSURES

None.

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