Parry-Romberg Syndrome and an Associated Complex Vascular Lesion Managed With Hybrid Strategies: A Case Report

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BACKGROUND AND IMPORTANCE: Parry-Romberg syndrome (PRS) is a condition of unknown etiology, most commonly characterized by progressive facial hemi-atrophy due to breakdown of the underlying skin and soft tissues. PRS has been associated with neurologic disease such as intracranial aneurysms and vascular malformations. However, its exact role in the incidence of neurovascular disease remains unknown.

CLINICAL PRESENTATION: We present a case of a complex fusiform aneurysm involving the A1 segment of the left anterior cerebral artery in a 21-year-old man with PRS. The patient initially presented with a left giant fusiform/dissecting internal carotid artery aneurysm initially managed with overlapping flow diverters. His post-flow diverter treatment course was confounded by progressive growth of the aneurysm with worsening mass effect on the optic chiasm leading to complete visual loss in the left eye and a right hemifield cut. To manage the aneurysm recurrence, retreatment with surgical bypass-supported carotid occlusion was performed. Thereafter, visual symptoms improved, but complete normalization of vision was not achieved. The patient remained clinically stable for several years until follow-up imaging demonstrated a novel adjacent recurrence involving the ipsilateral A1 segment, previously noted to be angiographically normal. The patient underwent superselective angiography followed by endovascular occlusion of the new fusiform lesion.

DISCUSSION: The case presented here illustrates the hybrid roles of endovascular and surgical treatment modalities in treating complex vascular lesions. The recurrence of the lesion following initial treatment strategies introduces questions about how the nature and development of the lesion affected the success of treatment at each stage. Considerations for the efficacy of each stage of treatment in this case include the patients underlying connective tissue disorder, the pediatric etiology of the lesion, and the timing of each treatment option given these factors.

CONCLUSION: In addition to understanding the biological behavior of flow diversion for complex intracranial aneurysms, the role of PRS in the incidence of neurovascular disease should also be considered when applicable. More research into the mechanisms of PRS in neurovascular disease is needed.

KEY WORDS: Bypass, Case report, Endovascular coiling, Flow diversion, Intracranial aneurysm, Parry-Romberg syndrome, Vascular disorders

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Intracranial aneurysms in pediatric patients are rare compared with the adult population. Aneurysms in children tend to be larger at the time of presentation, may be associated with an underlying genetic condition, and compared with adults, aneurysms in children are seen predominantly in male instead of female individuals.[1-4] In general, the etiology of cerebral aneurysms in adults is associated with intrinsic factors, such as genetic conditions, and extrinsic factors and acquired characteristics, such as smoking and hypertension. In children, extrinsic factors such as head trauma and infectious disease are more pertinent.[1,2,5] Many authors suggest that etiological differences in children are seen in the greater significance intrinsic factor contributions to aneurysm formation

ABBREVIATIONS: MRA, magnetic resonance angiography; **PED,** pipeline embolization device; **PRS,** Parry-Romberg syndrome.

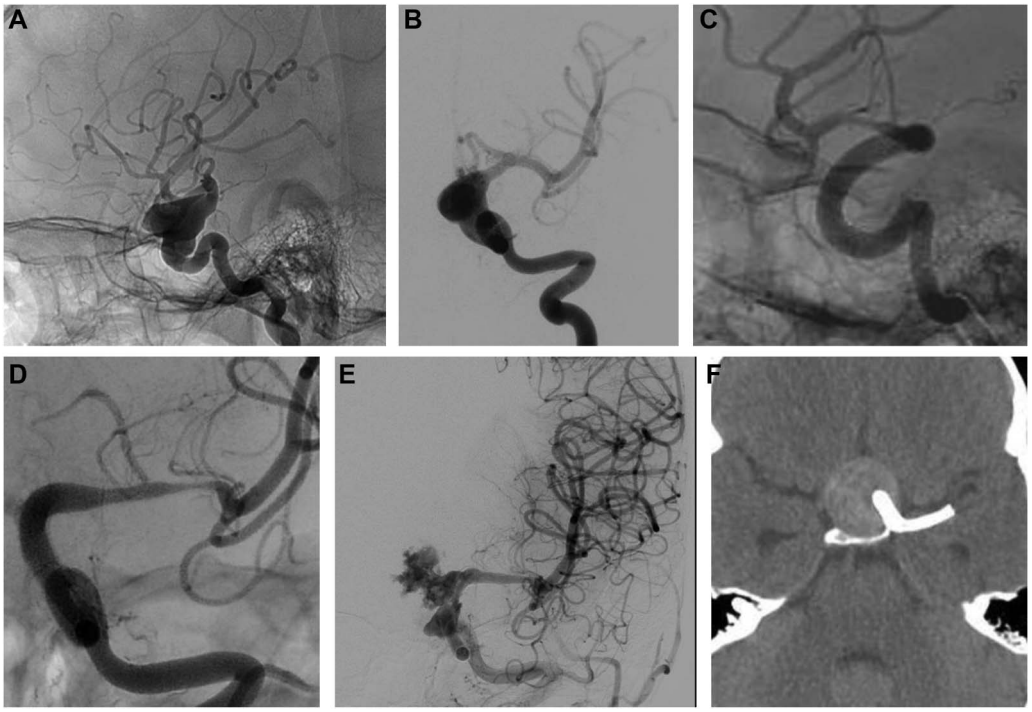

FIGURE 1. *The patient at age 10 years with giant fusiform/dissecting ICA aneurysm.* **A**, *Lateral and* **B**, *anteroposterior preprocedural cerebral angiograms demonstrate giant fusiform/dissecting ICA aneurysm.* **C** *and* **D**, *Follow-up cerebral angiograms 4 months after PED placement are notable for absence of flow into the previously visualized aneurysm.* **E**, *Follow-up cerebral angiogram 9 months after PED insertion at outside hospital demonstrate recurrent filling of the previously remodeled ICA aneurysm.* **F**, *Follow-up computed tomography angiography further demonstrates recurrence of the giant ICA aneurysm. ICA, internal carotid artery; PED, pipeline embolization device.*

such as genetic conditions including but not limited to polycystic kidney disease, moyamoya disease, Ehler-Danlos syndrome, and Marfan syndrome and potentially Parry-Romberg syndrome (PRS) as illustrated by our case example.[1-3,5,6]

We present the case of a patient with PRS and a history of giant fusiform/dissecting internal carotid artery (ICA) aneurysm that required surgical bypass after failed treatment with multiple flow diverting stents at age 10 years and later fusiform aneurysmal dilation of the A1 segment of the anterior cerebral artery (ACA).[7] PRS is a rare, acquired, connective tissue disease that most commonly causes progressive hemifacial atrophy.[7-11] It usually appears in the first decade of life and has a higher incidence in women.[8,12] The

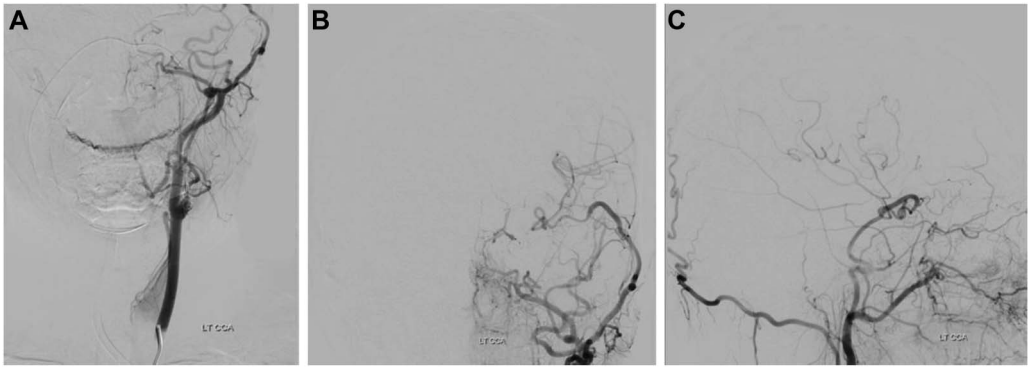

FIGURE 2. **A**, *Carotid occlusion and* **B** *and* **C**, *superficial temporal artery-middle cerebral artery bypass at age 12 years.*

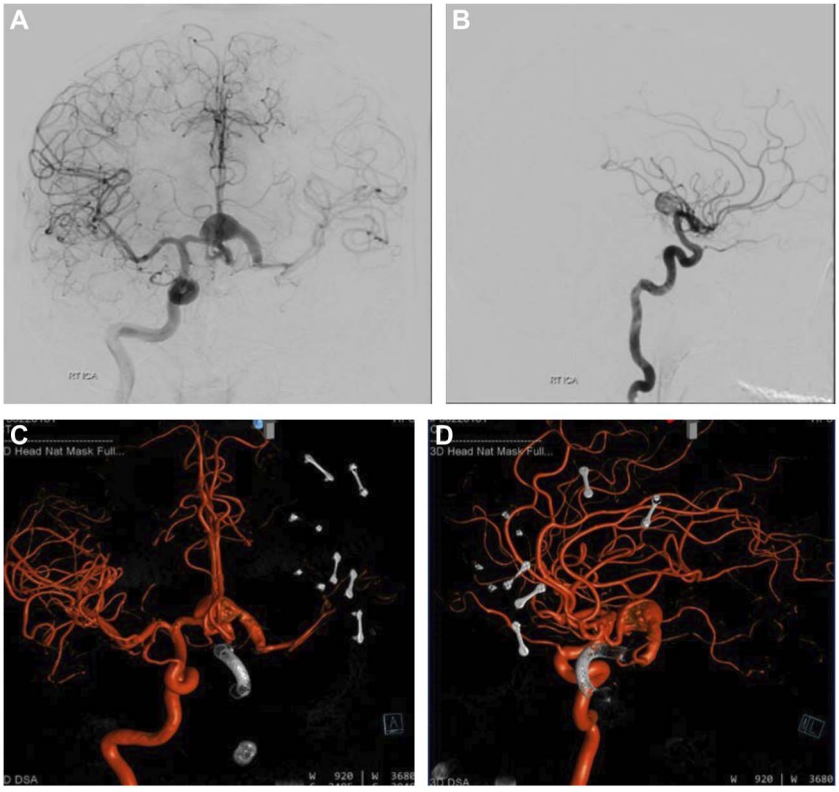

FIGURE 3. *The patient at age 21 years with aneurysmal dilation with progression toward the A1–A2 junction of the left ACA.* **A**, *Anterior and* **B**, *preoperative lateral cerebral angiograms demonstrate an approximately 9.0 mm fusiform aneurysmal dilation along the A1 segment of the left ACA.* **C** *and* **D**, *3D imaging confirm the presence of a 9.0 mm aneurysmal dilation of the left ACA. With permission from ©New York University. All rights reserved. ACA, anterior cerebral artery.*

origin of PRS is unknown, but proposed hypotheses include trauma, immune-mediated processes, and sympathetic dysfunction.[8] As a connective tissue disorder, its potential effects on vascular endothelia may contribute to the diseased segments of the patient's intracranial vessels.[5] In fact, the most frequent systemic manifestations associated with PRS are neurologic, including seizures, intracranial aneurysms, and vascular malformations.[8-12] This case report has been reported in line with the Surgical CAse REport (SCARE) criteria.[13]

CLINICAL PRESENTATION

A 21-year-old male patient with PRS and a history of a giant fusiform/dissecting ICA aneurysm with a large saccular component (Figure 1A and 1B) was treated with 7 overlapping pipeline embolization devices (PEDs) (ev3 Inc.), extending from the left M1 segment proximally into the cavernous segment of the left ICA across the fusiform aneurysm, reconstructing the affected ICA. His post-PED procedure clinical course was notable for persistent loss of vision in the left eye and temporal hemifield deficit in the right eye presumably because of mass effect on the optic chiasm owing to progressive thrombosis and incomplete aneurysm occlusion.[7] On imaging follow-up, after initial remodeling of the left internal carotid artery (Figure 1C and 1D), the aneurysm ultimately recurred (Figure 1E and 1F) and was subsequently treated with low-flow superficial temporal artery-middle cerebral artery (MCA) bypass and carotid occlusion in 2012 (Figure 2A-2C).[7] High-flow bypass from the right common carotid artery to the M2 segment of the right MCA with clip ligation of the parent vessel was initially recommended. At the time of surgery, it was recognized that the proposed clip would have disrupted sufficient perfusion of the anterior choroidal artery. In addition, the high-flow bypass was not performed because of the small size and short length of the radial artery.[7] The required institutional review board approval was obtained to prepare this case report.

At the latest presentation in 2022, the most recent follow-up angiogram from 2021 demonstrated development of a new fusiform aneurysm involving the A1 segment of the left ACA extending from the margin of the initial flow diverter construct at

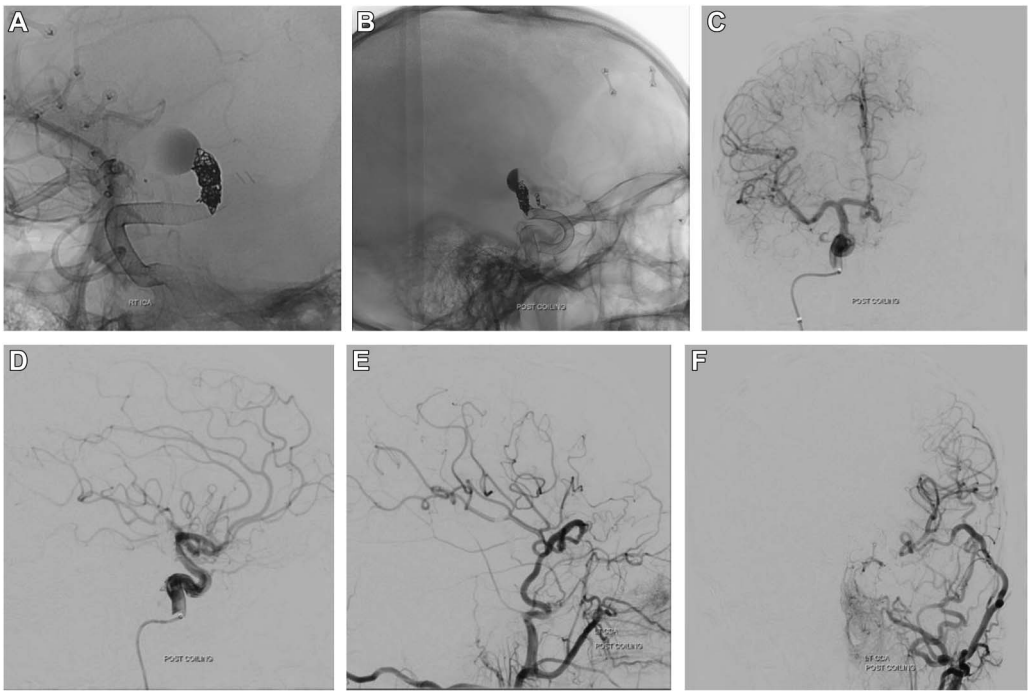

FIGURE 4. **A** *and* **B**, *Intraoperative cerebral angiogram demonstrates stagnation of contrast in the aneurysmal dilation immediately after trapping with detachable coils.* **C** *and* **D**, *Postoperative angiogram notable for no flow into the previously noted left anterior cerebral artery aneurysmal dilation.* **E** *and* **F**, *Postoperative cerebral angiogram demonstrates fully functioning bypass. With permission from* ©*New York University. All rights reserved.*

the level of the ICA bifurcation. Corresponding serial computed tomography angiography and magnetic resonance angiography (MRA) suggested progression toward the A1–A2 junction of the left ACA and growth of the dilation to approximately 9.0 mm cross-sectional diameter. Diagnostic angiography at the time of most recent retreatment demonstrates the latest aneurysmal development just before treatment (Figure 3A-3D).

With informed consent, diagnostic and superselective angiography +/− balloon test occlusion were planned to determine the best course of treatment. Ultimately, balloon test occlusion of the A1 dilation was deemed unnecessary after flow with the left A1 segment was found to be reversed; physiologically (filling from the right ICA through the anterior communicating artery [AComA]), superselective angiography did not reveal evidence of patent

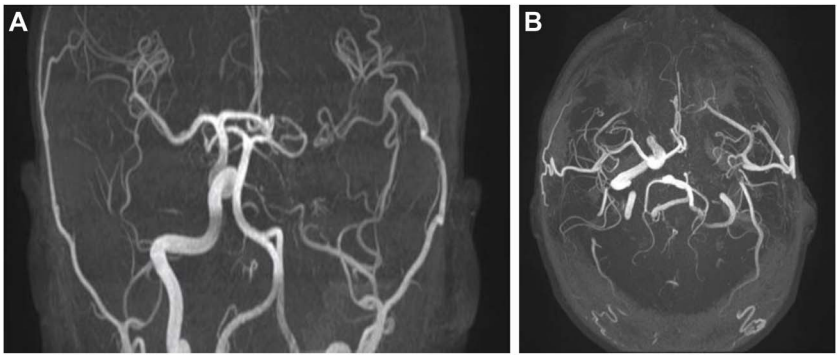

FIGURE 5. **A** *and* **B**, *Magnetic resonance angiography at 6 months postoperation demonstrates no evidence of residual or recurrent fusiform aneurysmal dilation and a fully functioning bypass.*

medial lenticulostriate vessels arising from the diseased left ACA segment, and the filling of the posterior communicating artery was found to be retrogradely supplied. Parenthetically, the left MCA combined contribution: directly through the still patent bypass and indirectly through the AComA. Considering these angiographic rearrangements in cerebrovascular support, the patient underwent endovascular occlusion of the diseased left A1 segment to the level of the flow diverter construct at the ICA bifurcation. Stagnation of contrast in the dome of the fusiform aneurysm was observed immediately after the placement of coils (Stryker Neurovascular) (Figure 4A and 4B). The distal left A2 was supplied through the AComA and the left MCA through the bypass (Figure 4C and 4D). There were no intraoperative complications, and the patient was neurologically unchanged postprocedure and at hospital discharge.

MRA at 6 months post-treatment showed no evidence of residual or recurrent aneurysm and a fully functioning bypass (Figures 4E and 4F and 5A and 5B).

DISCUSSION

The fusiform aneurysm of the left A1 segment was successfully and uneventfully treated deconstructively by detachable coil occlusion—rearranging the cerebral vasculature to definitively arrest flow in all segments of the aneurysmal ICA-ACA continuum and essentially completing the deconstructive occlusion envisioned at the time of the bypass. Stable occlusion of the complex evolving aneurysm was confirmed at 6-month noninvasive follow-up MRA. The original case study outlining the PED placements and surgical retreatment examined the potential limitations of flow diversion as a solo treatment for complex evolving intracranial aneurysms in specific situations such as the setting of unusual underlying conditions.[7] Although much enthusiasm for treating complex large and giant ICA aneurysms followed publication of the initial PUFS trial[14] and its follow-up,[15] subsequent consideration of failed treatments indicated specific conditions prognostic of treatment failure.[16] Abla et al,[7] in evaluating the initial failure with flow diverters therapy in this case, extended that analysis: hypothesizing that changes in intraaneurysmal flow dynamics after flow diverters placement in complex aneurysms can, under certain conditions, cause mural destabilization of the aneurysm and disrupt thrombosis, contributing to failed treatment with possible exacerbation of mass effect or ultimate rupture. Although some of this speculation may be true, it seems equally likely in this case, given the continued evolution of the lesion after attempted bypass-supported ICA occlusion, that treatment failure was predicated on premature intervention on an aneurysm early in its morphologic development: destined to involve the ICA bifurcation and left A1 segment, possibly related to an intrinsic vessel vulnerability confounding the patient's PRS.[5] For instance, histopathological examination of PRS skin has revealed atrophy of vessels, and electron microscopy has identified degenerative alterations of vascular endothelia.[8,17] These modifications have potentially important implications for flow diverter endothelialization. From this perspective, it is instructive to consider the potential role of PRS in the temporal expression of a patient's aneurysmal disease and how this knowledge might contribute to determining the optimum time for treatment: for instance, if serial imaging, with vessel wall interrogation, has a role in estimating the morphologic maturation of such lesions—guiding treatment initiation and indications for potential staged approaches. Alternatively, given the pediatric nature of the lesion, it is likely there was no true optimal time for intervention because pediatric aneurysms are well documented to have higher rates of recurrence and de novo aneurysm formation. As such, it may be pertinent to emphasize the role of continued, frequent follow-up and intervention as necessary[1-6,18,19]

Limitations

Limitations of this study include those inherent to case reports and the retrospective study design.

CONCLUSION

While important to consider the biological behavior of flow diversion for complex and giant intracranial aneurysms, it is also pertinent to consider the potential role of PRS in the incidence of neurovascular disease when applicable. More research into the effect of PRS on cerebral vasculature and subsequently aneurysm formation is needed. A better understanding of these mechanisms may elucidate the nature of aneurysms in the pediatric population, improve treatment, and determine the clinical importance of intracranial aneurysms in differential diagnoses and continued long-term follow-up in PRS cases.

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The authors have no personal, financial, or institutional interest in any of the drugs, materials, or devices described in this article.

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COMMENTS

This is a very interesting case involving a fusiform internal carotid artery aneurysm, initially treated with a flow diversion stent, which failed to cure the aneurysm, subsequent performance of a low flow bypass, and a later occurrence of a fusiform ACA aneurysm, which required endovascular occlusion. Although the initial treatment was appropriate, when the aneurysm recurred, a high flow bypass using a radial artery or saphenous vein should have been done, with aneurysm occlusion. This would have been safer for the patient and avoided the later development of an ACA aneurysm which was due to increased collateral flow.

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The authors present an interesting case of de-novo aneurysm formation as an unintended consequence in a pediatric patient with Parry-Romberg (progressive hemifacial atrophy) syndrome following aneurysm treatment. In this case, a giant fusiform aneurysm of the left internal carotid artery recurred after flow diversion. It was treated with low-flow bypass and internal carotid artery sacrifice with successful obliteration of the index lesion. Due to the combination of the patient's underlying connective tissue disease and iatrogenic flow reversal, a de-novo fusiform lesion later developed in the adjacent A1 vessel, and the patient then underwent successful treatment for this new lesion via coil embolization of the diseased segment without neurological sequelae.

In this case, the emphasis is on the dynamic and progressive nature of aneurysm formation in this unique subset of patients. Although an endovascular solution may be initially appropriate in similar patients, particularly when considering the potential need for future microvascular facial reconstructive plastic surgery, open and combined answers should remain in the cerebrovascular armamentarium. Flow diversion may have limitations as an isolated modality for treating complex evolving intracranial aneurysms, especially in the context of unpredictable vessel pathologies. This case highlights the complexity of decision-making in pediatric patients with underlying vascular disease and the need for continued close observation in vulnerable patients with rare and acquired aneurysms, even after an initial angiographic "cure" is achieved. Therapy success in this group of patients has to be measured against the lifetime ahead of them, which is arguably a high bar.

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