Focal drug-resistant temporal lobe epilepsy associated with an ipsilateral anterior choroidal artery aneurysm: illustrative case

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BACKGROUND The occurrence of both an intracranial aneurysm and epilepsy, especially drug-resistant epilepsy (DRE), is rare. Although the overall incidence of aneurysms associated with DRE is unclear, it is thought to be particularly infrequent in the pediatric population. Surgical ligation of the offending aneurysm has been reported in conjunction with resolving seizure activity, although few cases have cited a combined approach of aneurysm ligation and resection of an epileptogenic focus.

OBSERVATIONS We present the case of a 14-year-old female patient with drug-resistant temporal lobe epilepsy and an ipsilateral supraclinoid internal carotid artery aneurysm. Seizure semiology, electroencephalography monitoring, and magnetic resonance imaging all indicated a left temporal epileptogenic focus, in addition to an incidental aneurysm. The authors recommended a combined surgery involving resection of the temporal lesion and surgical clip ligation of the aneurysm. Near-total resection and successful ligation were achieved, and the patient has remained seizure free since surgery at 1 year postoperatively.

LESSONS In patients with focal DRE and an adjacent intracranial aneurysm, a combined surgical approach involving both resection and surgical ligation can be used. Several surgical timing and neuroanesthetic considerations should be made to ensure the overall safety and efficacy of this procedure.

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KEYWORDS drug-resistant epilepsy; focal epilepsy; ipsilateral cerebral aneurysm; pediatric epilepsy
resolved following surgical ligation of the offending aneurysm. All but 2 cases report on adult patients, underscoring the paucity of literature on this entity in pediatric patients. Furthermore, only a few studies have reported a combined treatment approach of surgical ligation of the unruptured aneurysm and resection of an epileptogenic focus.

Given the dearth of literature outlining the surgical decision making, planning, and technical nuances associated with DRE coupled with an intracranial aneurysm, we present the case of a 14-year-old female patient with drug-resistant temporal lobe epilepsy noted to have an ipsilateral supraclinoid internal carotid artery (ICA) aneurysm.

**Illustrative Case**

A 14-year-old, right-handed female patient presented to Boston Children’s Hospital Epilepsy Center with DRE associated with a left temporal lesion. Seizure onset was at approximately 4 years of age, with episodes of behavioral change and a scared look with her right arm shaking that could progress to bilateral tonic-clonic movements. These episodes occurred approximately 1 to 2 times per month. On neuropsychological examination, her IQ was in the extremely low range, with stronger visual-spatial processing compared with reasoning and verbal skills, as well as attention and executive function difficulties, all implicating dysfunction of the dominant frontotemporal systems. Her visual fields were intact.

Because of persistent seizures despite adequate trials of 3 anti-seizure medications (ASMs), she was referred for epilepsy surgery evaluation. Long-term electroencephalography monitoring demonstrated left temporal slowing interictally and left hemispheric ictal onset maximum in the left temporal region. Magnetic resonance imaging (MRI) revealed a faintly T2-weighted hyperintense lesion with a hypointense medial rim in the left anterior temporal lobe with extension into the mesial structures and subtle contrast enhancement along the medial and superomedial uncus (Fig. 1A and B). Notably, a left supraclinoid ICA aneurysm was also identified medial to the uncal enhancement. Magnetic resonance angiography (MRA) confirmed a 4-mm saccular aneurysm arising posteriorly from the left anterior choroidal artery (AChA) (Fig. 1C and D). Functional MRI (fMRI) revealed left-dominant language and working memory, while visual and motor function demonstrated bilateral representation. Consensus was reached at a multidisciplinary epilepsy surgery conference to proceed with left frontotemporal craniotomy for electrocorticography (ECOG)-guided resection of the left temporal epileptic foci and microsurgical clipping of the left AChA aneurysm.

**Operative Course**

After standard preoperative assessment and counseling, the patient was admitted to our institution on the day of surgery. General anesthesia was modified to permit intraoperative neuromonitoring, somatosensory evoked potentials, and ECOG. After the induction of anesthesia and tracheal intubation, general anesthesia was maintained with dexmedetomidine (0.5 µg/kg/hr), sufentanil (0.1 µg/kg/hr), and isoflurane (0.5%), and neuromuscular blockade with vecuronium (0.1 mg/kg/kg). To mitigate excessive blood loss, tranexamic acid (10 mg/kg loading dose and 5 mg/kg/hr) was administered to provide antifibrinolysis. Upon opening the dura, pre-resection ECOG was performed using a 4 × 8 subdural grid over the left anterolateral temporal lobe and a 2 × 8 grid extending posteriorly on the temporal lobe (Fig. 2A). Of note, mesial strip placement was...
deferred out of concern for aneurysm disruption. Initial ECOG data demonstrated left temporal interictal spiking predominantly at the anterior temporal tip. Less prominent spiking was noted in the posterior superior temporal gyrus, corresponding to receptive language activation identified on preoperative fMRI. An anterior temporal lobectomy, extending 3 cm posterior to the temporal tip on the superior temporal gyrus and 4 cm on the middle temporal gyrus, was planned.

Before temporal resection commenced, proximal control of the left ICA was obtained using a left lateral supraorbital trajectory. This helped identify the supraoptic arachnoid adhesions, free the ipsilateral optic nerve, and circumferentially release peri-carotid arachnoid bands that would allow for temporary clip occlusion in the event of an intraoperative rupture. A 20-mm straight temporary clip was prepared, and a 0.5 x 3 cottonoid was placed on the basal frontal surface to define the trajectory of the approach. Once proximal access to the supracoiloid ICA was obtained, the anterolateral temporal lobectomy was performed using a standard subpial technique. The lateral temporal cortex was resected en bloc with preservation of the mesial structures to minimize traction on the aneurysm and associated vasculature. Abnormal-appearing fibrous tissue was identified on the lateral margin of the remaining mesial structures. A small specimen was obtained and sent to pathology for frozen section analysis (Fig. 2B), which was suspicious for a low-grade glioneuronal neoplasm.

Following the lateral temporal resection, our attention was directed to further exposure of the ICA given the newly optimized access to the carotid cistern. Throughout this aneurysm exploration, the neuroanesthesia team transitioned to administering propofol, allowing for burst suppression as needed. The ICA was dissected and exposed distally. Notably, the arachnoid was abnormally thick, with hemosiderin staining concerning for a previous subclinical subarachnoid hemorrhage. Further release in the subfrontal chiasmatic and supracarotid regions demonstrated the PCA, the left third nerve, the AChA, and the posteriorly projecting AChA aneurysm. Given the aneurysm's size, morphology, and suspicion of a remote sentinel hemorrhage, the neurovascular and epilepsy surgical teams decided that further dissection may require temporary clip placement, necessitating the patient to be placed in burst suppression. However, since burst suppression may alter the ability to perform post-resection ECOG and pilot clipping may obstruct the remaining resection, further dissection and clip ligation of the aneurysm were deferred to allow for continuation of the mesial temporal resection.

The mesial structures were then removed in the usual subpial fashion. The uncus and amygdala were quite fibrous and difficult to dissect, resulting in a careful, piecemeal resection; this tissue was also sent to pathology. Following resection of the anterior 1 cm of the head of the hippocampus, post-resection ECOG was performed using a 1 x 8 strip on the inferior surface of the temporal lobe along the hippocampus posteriorly and another 4 x 8 grid that was placed over the lateral temporal lobe extending posteriorly (Fig. 2C). Prominent interictal spiking was noted posterior to the resection line near the middle and inferior temporal gyri, with no abnormal activity from the remaining posterior hippocampus. A safe margin for an extended resection was planned using the vein of Labbé as the posterior margin.

Following ECOG, our attention was returned to the ICA aneurysm, where propofol delivery recommenced. Further sharp microdissection was performed, the patient was placed in burst suppression, and the aneurysm neck and dome were released (Fig. 2D). An initial intraoperative indocyanine green fluorescence angiography (ICG-FA) was performed, demonstrating that the AChA derived from the shoulder of the aneurysm, precluding complete clipping of the aneurysm at its origin.
from the left ICA (Fig. 3A). A 10-mm straight temporary clip was placed on the proximal ICA for less than 10 minutes, allowing for the placement of a 6.5-mm, slightly curved permanent clip without visual evidence of parent vessel occlusion. During clip placement, a small site of aneurysm wall tear was noted on the inferior medial aspect of the aneurysm dome, which was secured on final clip positioning. A 5-mm mini-permanent clip occluded a small amount of neck residual at the distal end of the aneurysm. Proximal and distal temporary occlusion was initiated for 6 minutes for further inspection of the clip placement to confirm the exclusion of a small adjacent perforator (Fig. 2E). Subsequent visual inspection and an additional ICG-FA were performed to ensure complete aneurysm occlusion and continued perfusion of the parent artery and surrounding perforators without signs of vasospasm or occlusion (Fig. 3B).

With the aneurysm secured, the posterior resection of the middle and inferior temporal gyri and basal temporal lobe was completed in a subpial fashion using the previously planned posterior resection margin (Fig. 2F). Intraoperative MRI demonstrated complete resection of the enhancing lesion and a small infarct centered on the posteriorinferior putamen. Closure then commenced in the standard fashion. After 11.5 hours, the patient emerged from general anesthesia and neuromuscular blockade. She responded to commands, tolerated tracheal extubation, and was admitted to the surgical intensive care unit for hemodynamic monitoring and serial neurological examinations.

**Postoperative Course**

Postoperatively, the patient resumed her home ASMs (levetiracetam, oxcarbazepine, zonisamide) and experienced no lasting neurological deficits, except for a mild right superior quadrantanopia. Her speech function has remained at baseline, although she noted subtle cognitive improvements enabling increased productivity at school; however, she continues to endorse reading and speech comprehension challenges. The patient has remained on ASMs following surgery per the recommendation of our epileptologists. Notably, one of her ASMs (zonisamide) was successfully weaned. Pathology was consistent with gliosis and possible meningoangiomatosis, which is rare but generally considered hamartomaticous and nonneoplastic. The tissue samples were negative for both BRAF p. V600E and clinically significant fusions, although somatic alterations in the NF2 gene (NF2 c.1540A > G [p.M514V]) were noted on molecular Oncomap analysis.

**Patient Informed Consent**

The necessary patient informed consent was obtained in this study.

**Discussion**

Intracranial aneurysms are rare in the pediatric population, and their association with focal epilepsy is not well understood. To our knowledge, this case involving a 14-year-old female patient with drug-resistant temporal lobe epilepsy associated with an intracranial aneurysm is one of the youngest involving DRE and an ipsilateral ICA aneurysm. Given the dearth of literature on this association, we herein document our treatment approach and surgical decision-making process. In doing so, we highlight critical technical nuances for neurosurgeons to consider when attempting surgical clip ligation of an aneurysm and resection of a seizure focus during a single procedure.

**Observations**

While the prevalence of drug-resistant focal epilepsies associated with pediatric aneurysms remains unclear, focal epilepsy is one of the predominant forms of epilepsy in children.9 According to a recent meta-analysis by Sultana et al.,20 the cumulative incidence of DRE in the pediatric population is reported to be as high as 25%, with focal epilepsy being a significant predictor of DRE. As a result, patients with DRE are often considered for neurosurgical intervention to reduce seizure frequency and/or achieve seizure freedom, particularly when a lesional epileptogenic focus is identified, as seizure freedom rates as high as 60%–80% have been reported in pediatric series.21–26 In addition, improved cognition, mood, and quality of life can be achieved.27–29

Pediatric aneurysms are rare12,13 and can be more difficult to treat than adult aneurysms because of differences in size, intracranial locations, clinical presentations, and outcomes.13,30–32 In children with intracranial aneurysms, subarachnoid hemorrhage is a common presentation,17 in addition to clinical symptoms related to rupture, such as headache, vomiting, seizures, and loss of consciousness.33 Standard treatment modalities typically involve endovascular or microsurgical approaches,34 the latter of which was chosen in our case because of the simultaneous resection of the left temporal epileptogenic lesion. To maximize postoperative seizure control, resection was also combined with intraoperative ECOG, which has been shown to improve seizure outcomes in patients with low-grade lesions and focal cortical dysplasia.35–38

When deciding to perform both procedures concurrently, there were several important factors to consider. Notably, it was necessary to discuss whether clipping the aneurysm should occur during the same surgery as resection of the aforementioned lesion, and if done concurrently, what sequence would be most appropriate while also balancing neuroprotective strategies with the desire to perform ECOG. Another critical consideration involved the risk of removing brain structures adjacent to the aneurysm to allow direct exposure.
of the aneurysm, which could also lead to potential traction on the unsecured aneurysm and associated vasculature. Proximal control was achieved before resection to mitigate this risk.

Anecdotally, tumor progression and vessel wall involvement have been associated with the eventual rupture of an intracranial aneurysm.\textsuperscript{38} In addition, many reports on intracranial aneurysms and brain tumors emphasize treating the aneurysm before or concurrent with the tumor resection to safeguard against perioperative rupture.\textsuperscript{40–43} Although the pathology of our patient’s lesion did not definitively confirm neoplastic tissue, these findings suggest that treatment of the aneurysm at or before resection of an associated, possibly neoplastic epileptic lesion is likely warranted. Moreover, despite limited literature on the co-management of aneurysms and epilepsy, a few series have recommended a combined surgical approach when the aneurysm is positioned ipsilateral to the lesion.\textsuperscript{11,16,17} In our patient, the aneurysm’s ipsilateral location, coupled with the meningeal thickening and hemosiderin staining of the aneurysm dome observed during surgery, pointed toward the value of simultaneous management of the aneurysm and lesion. We do not believe there was a clinically detected remote hemorrhage event in the patient’s presentation; however, evidence of rupture would further justify the rationale for clipping the aneurysm in the same surgery as the lesion resection.

During surgery, several technical nuances were essential for ensuring the success of the resection and surgical ligation. Because mesial temporal/uncal dissection posed a significant risk of aneurysm rupture during surgery, careful and deliberate decisions were made in the surgical sequencing. We began the surgery by establishing pre-resection ECOG to measure baseline seizure activity. Before the temporal resection, we obtained proximal control of the aneurysm, allowing for temporary clip placement in the event of an intraoperative rupture. Then, we began an anterior temporal lobe resection, which improved visualization and the working corridor for potential intraoperative rupture. During this time, we began obtaining proximal control of the aneurysm in the same surgery as the lesion resection.

The timing of aneurysm exploration and clip ligation is particularly important to this single-surgery approach. Early in the procedure, once the initial craniotomy and baseline ECOG are established, it is essential to obtain proximal exposure of the aneurysm in case of intraoperative rupture during later resection of the seizure focus. To minimize interference with post-resection ECOG monitoring and hindrance of the clip within the surgical cavity, clip ligation and subsequent burst suppression should be done after the majority of the epileptogenic lesion has been resected. Throughout the procedure, careful and constant communication between the surgical and anesthesia teams should occur, as the different surgical stages may necessitate different neuroanesthetic agents. In our case, the patient has done well postoperatively and remains seizure free at her neurological baseline since surgery.

Ultimately, focal DRE epilepsy associated with an ipsilateral intracranial aneurysm is rare. However, using a multidisciplinary approach and careful technical considerations enables the safe and effective treatment of both pathologies in a single operation.

References

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Lessons

Our case highlights the rare co-occurring entity of lesional DRE and ipsilateral intracranial aneurysm in a pediatric patient and the feasibility of a combined treatment approach. In these cases, we recommend the patient and family be counseled on the benefits of a combined surgical approach, involving both resection of the epileptogenic lesion and surgical ligation of the aneurysm. This procedure has several critical steps that require deliberation by a multidisciplinary surgical team, as each stage of the resection and ligation comes with its own risks and benefits.

Disclosures
The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

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Conception and design: Stone, Phillips, Rangwala, Segar, Pinto, See. Acquisition of data: Phillips, Rangwala, Papadakis, Segar, Tsuboyama, Pinto, Harmon, Soriano. See. Analysis and interpretation of data: Stone, Phillips, Rangwala, Papadakis, Tsuboyama, Pinto, Harmon, Soriano. Drafting of the article: Stone, Phillips, Rangwala, Papadakis, Segar, Pinto, Soriano. Critically revising the article: Stone, Phillips, Rangwala, Papadakis, Segar, Tsuboyama, Pinto, Harmon, Soriano. Madsen, See. Reviewed submitted version of the manuscript: Stone, Phillips, Papadakis, Segar, Tsuboyama, Pinto, Soriano, Madsen. See. Approved the final version of the manuscript on behalf of all authors: Stone. Administrative/technical/material support: Stone. Study supervision: Stone. Patient care: Munoz.

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