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Neuroimaging Highlight

Angiography and Pre-operative Embolization of an Extra-axial, Supratentorial Atypical Teratoid Rhabdoid Tumor

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Atypical teratoid rhabdoid tumor (ATRT) is a rare and malignant central nervous system tumor commonly seen in children less than 3 years old and rarely in older children or adults.[1] It usually presents as an intra-axial infratentorial tumor, with only three extra-axial cases reported so far.[1,4,5]

We report a rare case of an adolescent female with an extra-axial, dural-based, supratentorial ATRT, who underwent catheter angiography, pre-operative embolization, and staged surgical resection. To our knowledge, this is the first reported case of the use of catheter angiography and pre-operative embolization for ATRT.

An 18-year-old female presented with a large recurrent frontotemporoparietal tumor that was subtotally excised at another institution 10 months prior, with a histopathologic diagnosis of meningioma. On examination, she was well with no focal neurologic deficits. There was a large, exophytic, firm mass measuring 13 × 12 × 8 cm at the left frontotemporal area.

Contrast cranial MRI showed a 12.0 × 12.7 × 12.6 cm avidly enhancing, extra-axial mass in the left frontotemporoparietal area, with lobulated borders and intratumoral hemorrhage, extending extracranially through a craniectomy defect. (Figure 1)

The patient underwent pre-operative embolization. Catheter angiography showed an extremely hypervascular exophytic mass in the left frontotemporal region with arterial supply predominantly arising from multiple branches of the left middle cerebral artery (MCA), frontal branches of the left anterior cerebral artery (ACA), and the middle meningeal (MMA), deep temporal, and sphenopalatine branches of the left internal maxillary artery (IMAX). There was also marginal contribution from the frontal and parietal branches of the left superficial temporal artery (STA) and distal branches of the right MMA and STA. No supply was observed from the posterior circulation. The blood vessels were tortuous and exhibited significant tumor blush.

Superselective catheterization and embolization was performed using a Progreat microcatheter (2.7 Fr) and polyvinyl alcohol (355-500 microns) and gelfoam particles as embolic agents. The embolization was successful for the left IMAX, bilateral STA, and right MMA feeders, with complete cessation of flow from the feeding vessels. However, the large feeders from the left MCA and ACA could not be superselectively catheterized and were left untouched. (Figure 2)

She underwent tumor resection one day post-embolization. The tumor was extra-axial with a good arachnoid plane but was extremely vascular despite embolization. Only a subtotal excision (approximately 90%) was achieved because of the 3.7-liter blood loss. She underwent another operation 3 days later for complete tumor resection.

Histopathology showed a malignant round cell neoplasm with rhabdoid and epithelioid features. Immunohistochemistry showed GFAP, vimentin, and EMA positivity, and loss of INI-1 nuclear expression in tumor cells, consistent with ATRT.

Since ATRTs are common in very young patients, angiography and pre-operative embolization are not routinely used due to concerns with radiation-associated complications. This age group has decreased tolerance to the high radiation doses employed in angiographic procedures; furthermore, it is technically difficult to catheterize small-caliber pediatric blood vessels.[3] This patient was an adolescent; hence, we were able to perform tumor embolization with the aim of decreasing blood loss and operative time.[2]

Statement of Authorship. J.G.P.: Conceptualization; resource; data curation; writing – original draft preparation; writing – review and editing.

J.A.S.: Data curation; writing – original draft preparation; writing – review and editing.

M.C.S.: Writing – review and editing.

V.G.T.: Writing – review and editing.

E.B.C.: Resource; writing – review and editing.

K.O.K: Conceptualization; writing – original draft preparation; writing – review and editing; supervision; project administration.

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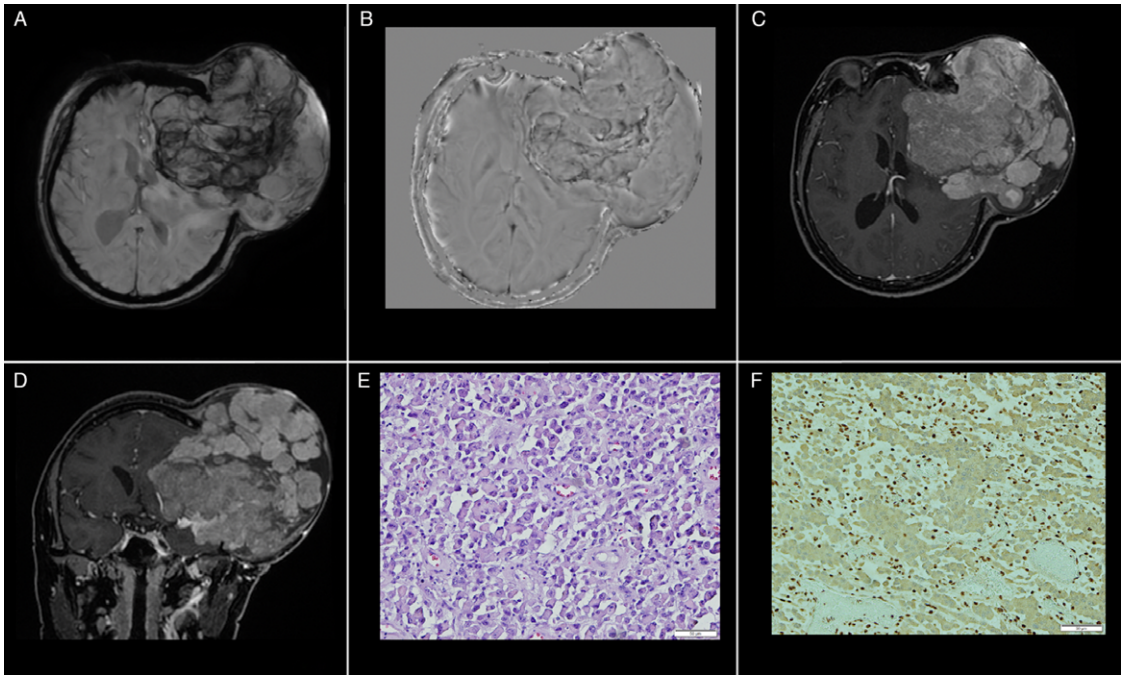

Figure 1: Contrast cranial MRI and histopathology photomicrographs. (A, B) Susceptibility-weighted imaging with corresponding filtered-phase image revealed extensive magnetic susceptibility artifacts consistent with marked intratumoral hemorrhage; (C, D) Axial and coronal cuts of the contrast-enhanced cranial MRI, T1W sequence with fat suppression, showing an avidly enhancing, dural-based, extra-axial mass in the left frontotemporal area measuring 12.0 x 12.7 x 12.6 cm, exhibiting extracalvarial extension beyond the left frontotemporal craniectomy defect. The tumor had lobulated borders and multiple feeding vessels at the area of the ACA and MCA; (E) Hematoxylin-eosin stain of the tumor specimen showing neoplastic cells with well-defined cell borders, abundant cytoplasm, and eccentrically located nuclei with vesicular chromatin and prominent eosinophilic nucleoli; (F) Immunohistochemistry studies showing loss of nuclear expression of INI-1 in tumor cells.

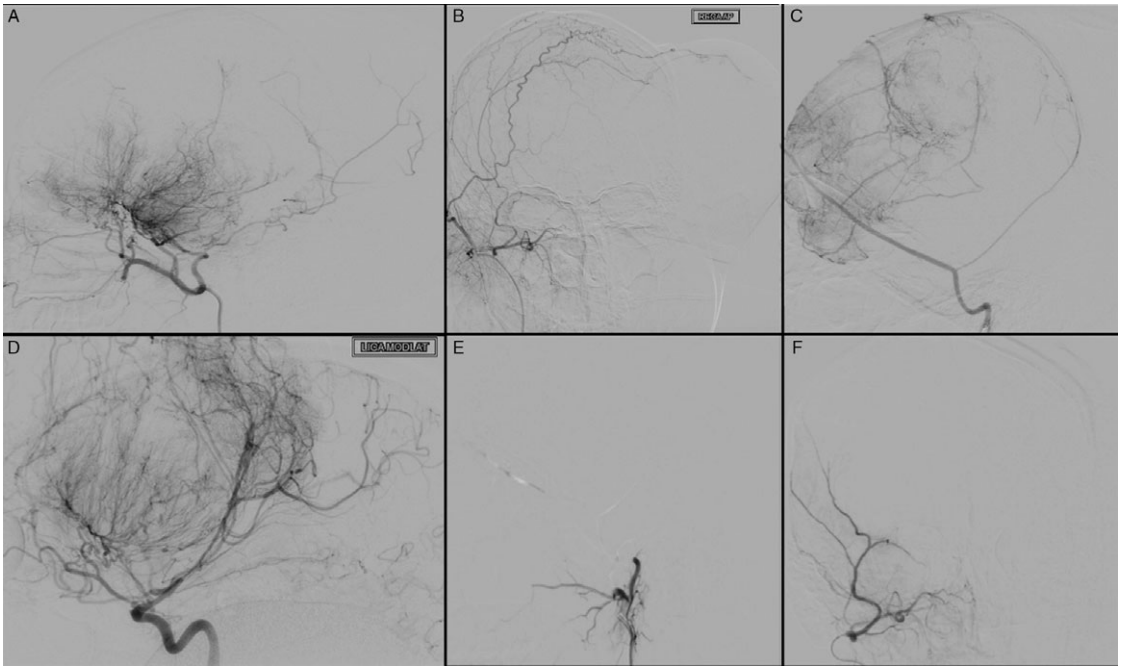

Figure 2: Diagnostic and post-embolization catheter angiography images. Pre-embolization catheter angiography images showing the following: (A) lateral view of left IMAX feeders involving the MMA, deep temporal, and sphenopalatine branches; (B) Right distal ECA injection (Anteroposterior view) showing feeders from the right MMA and STA; (C) Selective left STA injection (Modified Lateral view) showing tumor feeders arising from the frontal and temporal branches; and (D) Left ICA injection (Modified Lateral view) demonstrating the predominant feeders from the left MCA and ACA. Post-embolization catheter angiography images of the (E) Left distal ECA branches (Left IMAX and Left STA) and (F) Right MMA, showing successful embolization of these feeders. The large feeders from the left ACA and left MCA were not embolized.

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Conflicts of interest. The authors declare no conflicts of interest.

Consent to participate. Informed consent was secured from the patient.

Consent for publication. Informed consent for publication was secured from the patient.

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