Spontaneous Regression of a Congenital Tectal Glioma

Alejandro Feria, MD

Philipp Aldana, MD, FAANS, FAAP

1American University of the Caribbean School of Medicine

2Chief of Neurosurgery, Wolfson Children’s Hospital, Division of Pediatric Neurosurgery, University of Florida College of Medicine, Jacksonville
Disclosures

- None
Introduction/Methods

- Tectal gliomas are a class of low grade astrocytomas that typically present with symptoms of increased intracranial pressure and make up ~5% of brain stem gliomas.
- Congenital brain tumors are also uncommon comprising 0.5-1.9% of all pediatric brain tumors.
- Regression of these lesions is notably rare with only 2 prior cases reported in the literature.
- Here we present an unusual case of spontaneous regression of a congenital tectal glioma.

- Methods: Chart review
Case Report

10 week old female referred for evaluation of a tectal lesion found on screening MRI

Term Baby, C Section (prolonged labor)

Prenatal ultrasound showed cardiac mass, confirmed at birth via ECHO

- 9X8 mm Atrial Mass
- Initially thought to be cardiac hemangioma
- Failed propranolol Tx prompting screening ABD US and Brain MRI

MRI Brain findings:

- 6 X 6 X 5 mm mass superior tectum
- Slightly hyper intense on T1 with heterogeneous enhancement
- No midline shift or evidence of transependymal flow
- Radiographically consistent with a Tectal Glioma
Case Report

- **Initial Neurosurgical Examination:**
  - Head Circumference 90\textsuperscript{th} percentile (increase from 60\textsuperscript{th} at birth) with soft flat fontanelles
  - Without extraocular movement or gaze impairment or any other focal neurological signs
  - Met all expected developmental milestones
  - Otherwise asymptomatic
  - ETV Success Score - 50
    - (10 points - age 1-6 months, 30 points - tectal lesion, 10 points - no prior shunt)
  - As the child was not acutely ill, developing appropriately, and there was no urgent indication for surgical intervention, after discussion with her parents the decision was made to follow the child closely with serial cranial ultrasounds and brain MRI to assess her ventricular size and monitor for progression of the lesion.

- **Pediatric Clinical Genetic evaluation for congenital lesions at 2 sites**
  - Did not mean any full criteria for known cancer predisposition syndromes
  - Nervous system/brain cancer screening panel all negative: ALK, APC, DICER1, EPCAM, HRAS, MEN1, MLH1, MSH2, MSH6, NF1, NF2, PHOX2B, PMS2, PRKAR1A, PTCH1, PTEN, RB1, SMARCA4, SMARCB1, SUFU, TP53, TSC1, TSC2, VHL, BAP1, BARD1, EZH2, GPC3, KIF1B, POT1, PTCH2.
Close Interval Follow up

- Head circumference to 99th percentile, but maintains steady growth curve
  - Macrocephaly felt to be due to benign extra axial fluid collections of infancy

6 Month Follow up

- Tectal lesion nearly resolved
- No enhancement
- Decreased ventricular size
- Increased Aqueduct caliber

12 Month Follow up

- No progression of ventriculomegaly
- Meeting all expected milestones
- No new symptoms

- Tectum grossly within normal limits
- No signal abnormality or contrast enhancement
- Further Improvement of ventricular size
- T2 CSF flow void through cerebral aqueduct
Composite MRI imaging of tectal glioma regression and improvement of hydrocephalus

A/B. Sagittal and axial contrasted T1 weighted MRI at presentation shows a heterogeneously enhancing lesion near the superior colliculus consistent with a tectal glioma with resultant aqueductal stenosis and dilation of the third and lateral ventricles. C/D. 6 month follow-up contrasted T1 MRI shows decrease in tectal tumor size with no contrast enhancement and decrease in third and lateral ventricle size. E/F. 12 month follow-up contrasted T1 MRI shows a tectum that is grossly within normal limits and further improvement in ventricular size.
Focused MRI findings of the tectal region

A. On presentation a T1 MRI with contrast revealed a 6 X 6 X 5 mm heterogeneously enhancing mass ventral to the superior colliculus with resultant aqueductal stenosis. B. 1 year follow up T1 MRI with contrast shows the tectum to be grossly within normal limits with no signal abnormality or contrast enhancement. C. 1 year follow-up T2 MRI shows a CSF flow void through the cerebral aqueduct.
2 Previously Reported Cases of Spontaneous Tectal Glioma Regression

**Case 1**

- 13 year old with prior Hx periauricular plexiform neurofibroma at age 3 weeks
  - Presented with visual blurring and papilledema, shunted
  - MRI - tri-ventricular hydrocephalus, multiple brain lesions, 2 enhanced (globus pallidus, tectum)
- 6 mo F/U - Tectal lesion enlarged, others static
- 12 mo F/U - lesions decreased size, less enhancement
- Age 16 - TX for shunt malfunction
  - Marked decrease in lesion size, no more enhancement
  - GP lesion minimal decrease

**Case 2**

- 24F prior Hx shunt for aqueductal stenosis at 8 weeks of age
  - Eval for chronic headaches - small non-enhancing tectal glioma that remained stable on follow-up MRI
  - Increased HA severity & frequency once she became pregnant, no imaging
  - 2 months post-partum MRI showed a larger (~2X), contrast-enhancing lesion, 1 mo F/U size further increased
  - During radiosurgery planning 4 months post-partum, symptoms improve and lesion size decreases.
  - At 5 months post-partum, lesion size continues to decrease and no longer enhances.
Discussion

- Although the case presented here is the third reported spontaneous tectal glioma regression in the literature, this case is quite unusual in the following respects:
  - First described case in a child
  - Only published radiographically complete regression
  - Seen in the very rare setting of a congenital tumor
- Similar to case 1: Possible underlying uncharacterized genetic syndrome (multiple congenital lesions), although none identified on genetic screening.
- Similar to case 2: Possible underlying hormonal influence on CNS lesions (as in glioma/meningioma) during pregnancy.

Conclusion

- Complex genetic and hormonal factors may influence the progression and resolution of the reported lesion. Spontaneous tumor regression remains poorly understood.

Limitations

- The diagnosis of tectal glioma was not histologically confirmed, however, for the majority of tumors of the tectal region, operative biopsy is not indicated due to the surgical morbidity and the diagnosis is made based on the imaging characteristics.
- We only report on the short term outcome for this tumor as long term follow up continues.