Pediatric Septal Dysembryoplastic Neuroepithelial Tumor (sDNT): Current Trends

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Introduction

• Dysembryoplastic Neuroepithelial Tumors (DNTs) are a rare disease process with an overall incidence of 0.033 per 100,000 person-years (95% CI, 0.030-0.037) within the subset of the United States population\(^4\).

• Septal DNT (sDNT) histopathology may reveal infiltrative growth with entrapped neurons and axons\(^2\). sDNT are distinct from DNT as the pathology originates in septal nuclei.

• sDNT commonly have a high concentration of PDGFRA, FGFR1, and NF1 mutations\(^2\).

• Most common clinical presentation is complex partial seizures with secondary generalization\(^8\).

• On MRI, a lobular interventricular mass involving the anterior-inferior septum pellucidum is seen

• Treatment is usually gross or subtotal surgical resection
Methods

• A case reported from our institution is compared to the seven cases currently reported on the literature.

• We conducted a retrospective review of patients with sDNT on the basis of age, gender, symptoms, imaging findings, mechanism of therapy, outcome, length of outcome, and presence of metastatic disease. Seven patients were identified and compared to the case present at our institution.
# Results (Table 1)

<table>
<thead>
<tr>
<th>Case</th>
<th>Age</th>
<th>Gender</th>
<th>Presenting Symptoms</th>
<th>Imaging Findings</th>
<th>Mechanism of Therapy</th>
<th>Overall Outcome</th>
<th>Length of Follow-up</th>
<th>Presence of Metastatic Disease</th>
</tr>
</thead>
<tbody>
<tr>
<td>1$^2$</td>
<td>6</td>
<td>F</td>
<td>Personality Change + Seizures</td>
<td>Non-enhancing mass of interior septum extending through foramen of Monro bilaterally into anterior third ventricle</td>
<td>GTR</td>
<td>No further disease post-resection</td>
<td>16 mo</td>
<td>No</td>
</tr>
<tr>
<td>2$^2$</td>
<td>13</td>
<td>M</td>
<td>Headaches + Seizures</td>
<td>Lobulated non-enhancing mass of inferior septum extending through both foramen of Monro into Third ventricle; non-enhancing cortical mass of left temporal lobe with multiple small subependymal lesions</td>
<td>GTR of septal mass; lesions at other locations not biopsied</td>
<td>No further disease post-resection of septal mass; temporal and subependymal masses have stable disease processes</td>
<td>20 mo</td>
<td>Yes</td>
</tr>
<tr>
<td>3$^2$</td>
<td>17</td>
<td>M</td>
<td>Headaches with Nausea and Vomiting</td>
<td>Non-enhancing intraventricular mass of anterior septum located in close proximity to foramen of Monro</td>
<td>STR</td>
<td>Stable residual disease, currently asymptomatic</td>
<td>6 mo</td>
<td>No</td>
</tr>
<tr>
<td>4$^2$</td>
<td>19</td>
<td>M</td>
<td>Obstructive hydrocephalus</td>
<td>Non-enhancing mass of inferior septum extending into anterior third ventricle via foramen of Monro</td>
<td>GTR</td>
<td>N/A</td>
<td>N/A</td>
<td>N/A</td>
</tr>
</tbody>
</table>
### Results (table 1 continued)

<table>
<thead>
<tr>
<th>Case</th>
<th>Age</th>
<th>Sex</th>
<th>Symptoms</th>
<th>Radiological Findings</th>
<th>Treatment</th>
<th>Diagnoses</th>
<th>Follow-up</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>500</td>
<td>12</td>
<td>F</td>
<td>Headache with bilateral optic disk edema, transient visual loss, intracranial noises</td>
<td>Non-enhancing intraventricular mass with obstructive hydrocephalus</td>
<td>GTR with fenestration of septum pellucidum</td>
<td>Possible residual tumor vs. postoperative encephalomalacia</td>
<td>N/A</td>
<td>No</td>
</tr>
<tr>
<td>61</td>
<td>15</td>
<td>M</td>
<td>Seizure</td>
<td>Non-enhancing intraventricular mass involving right leaflet of cavum septum, superior portion of corpus callosum, left frontal horn of lateral ventricle, and extension towards foramen of Monro</td>
<td>GTR</td>
<td>N/A</td>
<td>N/A</td>
<td>N/A</td>
</tr>
<tr>
<td>73</td>
<td>15</td>
<td>F</td>
<td>Loss of consciousness following head trauma</td>
<td>Non-enhancing mass in foramen of Monro and septum pellucidum with hydrocephalus</td>
<td>GTR</td>
<td>No residual tumor</td>
<td>N/A</td>
<td>N/A</td>
</tr>
<tr>
<td>Our patient</td>
<td>16</td>
<td>F</td>
<td>Trouble focusing, generalized weakness, and occasional parasthesias</td>
<td>Non-enhancing lesion involving the anterior aspect of the septum pellucidum and fornix. Subtle findings of leptomeningeal spread</td>
<td>Endoscopic Biopsy</td>
<td>Residual tumor (stable)</td>
<td>3 Mo</td>
<td>No</td>
</tr>
</tbody>
</table>
Results (our patient)

Figure 1 (a-d)

Figure 2
Results

Literature Review Results
• average age at presentation: 14.1 years
• Population: 50% male, 50% female
• Most common presentation: Seizure (n=3), headache (n=3)
• All patients received T1-weighted MRI scans
• GTR in 6 of 8 patients
• Metastatic disease seen in 1 patient
• Average follow up time 11.2 months

Patient Case
• 16 y/o F w/ dysautonomia and Postural Orthostatic Tachycardia Syndrome
• Dx/ with low-grade glioma of the septum
• Patient underwent biopsy 1 month later
• Tolerated procedure well, no post-operative complications
• Patient underwent repeat MRI 3 months later with stable clinical and radiographic presentation
Discussion

• sDNT is a rare pediatric disease and can commonly present with headache or epilepsy.

• Previous recommendation has been gross resection

• Our case + 7 others in literature demonstrate biopsy as a reasonable alternative

• Patients should undergo serial imaging and followed clinically to ensure therapeutic benefit

• Our case demonstrated post-contrast FLAIR being less conspicuous on repeat exam

• Histopathology will likely reveal high concentration of PDGFRA mutation in addition to GFGR1/NF1
References


