Intracranial dermoid cysts are rare congenital lesions that typically occur in the cisternal spaces. However, exceptionally rare cases of intraaxial involvement have been reported, with only 8 cases having been described in the literature. The authors report the first case of an intraaxial dermoid cyst located in the medulla and the first occurrence in an elderly patient. They also review the literature of the existing intraparenchymal cases and provide treatment guidelines. A 66-year-old man presented with slowly progressive dysphagia, left lower-extremity numbness, nausea, and hyperhidrosis. Neurological examination revealed decreased pinprick sensation of the left side of his face and body, and decreased vibratory sensation in his left lower extremity. Additionally, he had an unusual extraocular movement in which abduction of the eye resulted in closure of the contralateral eye. Magnetic resonance imaging revealed a nonenhancing cystic lesion centered in the medulla. The patient underwent a suboccipital craniotomy with laminectomy of C1–2 for excision of the cyst, with subtotal resection due to adherence of the cyst wall to the brainstem. At follow-up 7.5 years after surgery, the patient’s neurological examination was stable. Magnetic resonance imaging did not reveal any progression or recurrence of the cyst. As the cyst wall is typically adherent to surrounding structures, resection is usually subtotal due to the risk of neurological deficits. As there have been no cases of progression after subtotal resection, gross-total resection is not warranted for the treatment of these lesions.

Case Report

**History and Presentation.** This 66-year-old man presented with dysphagia, left lower-extremity numbness, nausea, and hyperhidrosis around his eyes that slowly progressed over a period of months to years. Neurological examination revealed decreased pinprick sensation on the left side of his face and body and decreased vibratory sensation in his left lower extremity. Additionally, he had an unusual extraocular movement in which abduction of the eye resulted in closure of the contralateral eye. Magnetic resonance imaging revealed a 1.7 × 1.7 × 2.0-cm nonenhancing cystic lesion centered in the medulla. The lesion was homogeneously hypointense on T1-weighted images and homogeneously hyperintense on T2-weighted images and demonstrated no solid enhancing components. It was also hyperintense on DWI (Fig. 1). There was an associ-

**Key Words**  
- brainstem  
- cyst  
- dermoid  
- medulla  
- resection  
- oncology

Abbreviation used in this paper: DWI = diffusion-weighted imaging.
Dermoid cyst of the medulla

ated tiny cervical spinal cord syrinx, and perilesional and perisyrrinx edema in the medulla.

Operation. The patient underwent a suboccipital craniotomy with C1–2 laminectomy for excision of the cyst. Once the medulla was exposed, the cyst was readily apparent. To preserve all medullary structures that could be stuck to the capsule, the capsule was incised and all cyst contents were carefully removed without spillage in a piecemeal fashion. The contents were seen to contain squamous material and hair strands. Since the capsule could not be easily dissected off the medulla, pieces of the cyst wall were left within the medulla to prevent worsening of the patient’s neurological deficits. The dura was sealed with a pericardial graft, and the bone flap was replaced.

Pathological Findings. Histopathological examination showed dense fibrous connective tissue and reactive pilocytic astrocytes that enclosed the squamous epithelial cyst (Fig. 2). The cyst contained keratin debris, hair follicles, and hair strands.

Postoperative Course. The patient had an uneventful postoperative course. Neurological examination revealed stable extraocular movements and left-sided facial numbness. He also had mild lower-extremity weakness and decreased vibratory sensation below the knees bilaterally in addition to hyperreflexia. Nine months after surgery, his dysphagia and lower-extremity weakness had improved. At follow-up 7.5 years after surgery, the patient’s neurological examination was only remarkable for his abnormal extraocular movements. He also reported persistent and stable swallowing difficulties. Magnetic resonance imaging at this time did not reveal any progression or recurrence of the cyst.

Discussion

Dermoid cysts are rare, congenital lesions that are thought to arise from the inclusion of ectodermal cells within the developing neural tube during primary neurulation.1,2,5,18 Although this is consistent with many dermoid cysts preferentially localizing to the midline, rare lesions have been reported in lateral and intraaxial positions.3 Two hypotheses have been proposed to explain this distribution: 1) the differentiation of ectodermal cells included in the neuroectoderm and proliferation of multipotent stem cells, and 2) the migration of ectodermal remnants into brain parenchyma along neurovasculature after the development of Virchow-Robin spaces.24 Our case of an intraaxial location of a dermoid cyst in the medulla supports the second hypothesis, adding to a growing body of literature suggesting that ectodermal remnants can originate in an extraaxial location but migrate along Virchow-Robin spaces to invade the brain parenchyma.2,7,10,23

Differentiating dermoid cysts from other intraaxial cystic masses can be difficult, and MRI remains the best investigative tool for distinguishing dermoid cysts from other brain lesions. Dermoid cysts most commonly show a hyperintense signal on T1-weighted imaging and hypointense signal on T2-weighted imaging.15 The lesion in our case was homogeneously hypointense on T1-weighted images and homogeneously hyperintense on T2-weighted images, consistent with findings of 2 reported cases of cerebellar dermoid cysts.16,17 Fat suppression imaging and DWI may aid in identifying dermoid cysts due to the frequent presence of fatty and keratinoid components.21 The imaging approach to intraaxial cystic lesions should begin

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**Fig. 1.** Magnetic resonance images revealing a cystic lesion within the medulla, consistent with an intraaxial dermoid/epidermoid cyst. **A:** Contrast-enhanced T1-weighted axial image demonstrating a homogeneously hypointense lesion. **B:** Axial T2-weighted image showing a homogeneously hyperintense lesion. **C:** Diffusion-weighted image demonstrating the lesion to be hyperintense, suggesting an epidermoid or dermoid cyst. **D:** Contrast-enhanced T1-weighted sagittal image demonstrating a cyst centered in the medulla.

**Fig. 2.** Photomicrograph showing a squamous epithelial cyst with adherent keratin debris, consistent with a dermoid cyst. H & E, original magnification ×40.
with careful inspection for evidence of contrast enhancement of the wall of the lesion. While some nonneoplastic cystic lesions can occasionally show rim enhancement, the presence of a solid enhancing mural nodule should raise suspicion for a neoplasm. When enhancement is not present, however, purely cystic intraaxial lesions are most commonly nonneoplastic and may include parasitic cysts, enlarged Virchow-Robin spaces, neuroglial cysts, lacunar infarcts, porencephalic cysts, ependymal cysts, or neur-enteric cysts.11,13

Dermoid cysts progress slowly and generally do not produce clinical symptoms until the 3rd decade of life.6 Our 66-year-old patient is the oldest reported for an intraxial dermoid, suggesting unusually slow growth of the cyst or late onset of clinical sequelae. If clinical symptoms arise, or if there is a risk for cyst rupture, the recommended approach is resection of the cyst with removal of the capsule.12 For intraaxial lesions, the capsule may be difficult to remove if it adheres tightly to the surrounding parenchyma and vasculature.19 An aggressive surgical approach can result in high morbidity and mortality, which represents an extreme risk for the resection of a benign mass. In our case, we left a portion of the cyst wall intact in the medulla to prevent the development of postoperative complications.

Review of the Literature

Review of all intraaxial dermoid cysts (n = 9) revealed the median age to be 27 years (range 4–66 years), with 55.6% of cases occurring in females (Table 1). Of these lesions, 33.3% were found in the brainstem, with the remaining cases being located in the cerebellum and frontal lobes. Presenting symptoms were all dependent on cyst location: those in the brainstem presented with cranial nerve palsies while those in the cerebellum presented with signs of obstructive hydrocephalus, possibly due to cerebellar dermoid cysts compressing the fourth ventricle and obstructing CSF flow. Imaging characteristics were varied, with 42.9% of cases appearing homogeneously hypointense on T1-weighted imaging and 57.1% appearing hyperintense on T2-weighted imaging. Additionally 28.6% of cases had heterogeneous signal characteristics due to both fluid and fat components. Contrast enhancement was only seen in one case, presenting difficulty in distinguishing the dermoid radiologically from malignant cyst-associated lesions that also exhibit contrast enhance-

<table>
<thead>
<tr>
<th>Authors &amp; Year</th>
<th>Age (yrs), Sex</th>
<th>Location</th>
<th>Presenting Symptoms</th>
<th>MRI Characteristics</th>
<th>Treatment</th>
<th>Recurrence (length of FU in mos)</th>
</tr>
</thead>
<tbody>
<tr>
<td>van Calenbergh et al., 2005</td>
<td>4, F</td>
<td>pontomedullary junction</td>
<td>CN VI &amp; VII palsy</td>
<td>hypo/hyper</td>
<td>hyper</td>
<td>NA</td>
</tr>
<tr>
<td>Sener et al., 2004</td>
<td>27, M</td>
<td>basal frontal lobes</td>
<td>headache</td>
<td>hypo/hyper</td>
<td>hyper</td>
<td>no</td>
</tr>
<tr>
<td>Caldarelli et al., 2001</td>
<td>16, F</td>
<td>pons</td>
<td>CN VI &amp; VII palsy, nuchal pain</td>
<td>hyper/hyper</td>
<td>hypo</td>
<td>no</td>
</tr>
<tr>
<td>Pant &amp; Joshi, 2008</td>
<td>12, F</td>
<td>cerebellar midline</td>
<td>headache, emesis</td>
<td>hypo</td>
<td>hyper</td>
<td>no</td>
</tr>
<tr>
<td>Masri et al., 2009</td>
<td>4, F</td>
<td>cerebellar vermis</td>
<td>headache, emesis, recurrent meningitis, fever</td>
<td>hypo</td>
<td>hyper</td>
<td>yes†</td>
</tr>
<tr>
<td>Fernandez-Miranda et al., 2010</td>
<td>49, M</td>
<td>mesial frontal lobe</td>
<td>unilat numbness, paresthesia, personality changes</td>
<td>hyper</td>
<td>hypo</td>
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</tr>
<tr>
<td>Fenstermaker et al., 1989</td>
<td>43, F</td>
<td>lt frontal lobe</td>
<td>generalized seizures, aphasia, rt central facial paresis</td>
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<td>NA</td>
<td>NA</td>
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<tr>
<td>Uglietta et al., 1989‡</td>
<td>46, M</td>
<td>bilat frontal lobes, nasal sinus</td>
<td>confusion, memory loss</td>
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<td>NA</td>
<td>NA</td>
</tr>
<tr>
<td>current case</td>
<td>66, M</td>
<td>medulla</td>
<td>swallowing difficulty, nausea, LLE numbness, periorbital sweating</td>
<td>hypo</td>
<td>hyper</td>
<td>no</td>
</tr>
</tbody>
</table>

summary: median age 27 yrs, 55.6% female; 33.3% in brainstem; 33.3% headache; 42.9% hypo; 57.1% hyper; 16.7% enhancing; 88.9% subtotal; no recurrence (36 mos).

* CN = cranial nerve; FU = follow-up; hyper = hyperintense; hypo = hypointense; LLE = left lower extremity; NA = not available; STR = subtotal resection.
† Small enhancing nodule.
‡ Extension of nasal dermoid sinus cyst into the frontal lobes.
Dermoid cyst of the medulla

Adjuvant therapy. Progression of the cyst after subtotal resection has not been reported, with median follow-up of 36 months (range 1–90 months). Also, there have been no reported cases of chemical meningitis or leptomeningeal spread after resection.

Conclusions

Intraaxial dermoid cysts are exceptionally rare lesions that may mimic the appearance of cystic neoplasms. They typically occur in younger patients and may occur throughout the brain. Thus far only 2 previous cases have reported dermoid cysts within the brainstem, both involving the pons. We report the first case of an intraaxial dermoid cyst occurring in an elderly patient and the first case occurring within the medulla. Additionally, the reported case has the longest follow-up of all previously reported cases, demonstrating long-term outcome. Careful examination of the cyst wall for contrast enhancement and the use of imaging techniques such as fat suppression and DWI may aid in preoperative diagnosis. As the cyst wall is typically adherent to surrounding structures, resection is usually subtotal for these benign lesions because of the risk of neurological deficits. Resection may be aided by neuromonitoring and neuronavigation. As there have been no cases of progression after subtotal resection, gross-total resection is not warranted for the treatment of these lesions.

Disclosure

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

Author contributions to the study and manuscript preparation include the following. Conception and design: Adamson, Park, Babu. Acquisition of data: Park, Babu, Kranz, McLendon. Analysis and interpretation of data: Park, Babu, Kranz, McLendon. Drafting the article: Park, Babu. Critically revising the article: all authors. Reviewed submitted version of manuscript: all authors. Approved the final version of the manuscript on behalf of all authors: Adamson. Statistical analysis: Babu. Study supervision: Adamson.

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