Cervical intramedullary spinal cord tumors typically present with progressive neck pain, sensory complaints, weakness, or gait disturbance. Magnetic resonance imaging is the imaging modality of choice for patients with complaints suggestive of a process involving the cervical cord. If an enhancing intramedullary lesion is found, no additional imaging is performed in most cases.

Os odontoideum is a common cause of atlantoaxial instability in the pediatric population. The authors present the cases of 2 patients whose initial clinical presentation and MR imaging findings were suggestive of an intramedullary neoplasm, but whose ultimate diagnosis was determined to be cervical spine instability and cord injury due to os odontoideum. (DOI: 10.3171/2011.7.PEDS1186)

Key Words • os odontoideum • cervical instability • spine

Case Reports

Case 1

History and Examination. This previously healthy 13-year-old girl presented with progressive neck pain and myelopathy lasting approximately 1 month. There was no history of trauma or previous medical illness. On physical examination, she was grossly myelopathic with mild left hemiparesis, 3+ biceps reflexes, and a positive Hoffman sign bilaterally. Magnetic resonance imaging demonstrated focal intramedullary enhancement at C-1 with surrounding T2 hyperintensity (Figs. 1A and 2A); no dynamic imaging or CT scan was obtained, as the intramedullary lesion was presumed to be neoplastic.

First Operation. The patient underwent an uncomplicated C-1 laminectomy for resection of a presumed neoplasm. No overt instability was observed intraoperatively. Postoperative MR imaging demonstrated a gross-total resection of the enhancing lesion, but final pathology was consistent with gliosis and inflammation, without clear evidence of tumor.

First Postoperative Course. The patient’s neurologi-
cal examination remained unchanged during the immediate postoperative period and at short-term follow-up visits. Serial surveillance MR images demonstrated decreased T2 signal and no new enhancement. However, 1 year later she returned with progressive sensory loss. Additional MR imaging demonstrated persistent T2 signal at the cervicomedullary junction and a small area of contrast enhancement (Figs. 1B and 2B). At this point, a previously unseen os odontoideum was identified on MR imaging and CT scanning (Fig. 3).

**Second Operation.** The patient subsequently underwent posterior occiput–C2 instrumentation and fusion with iliac crest autograft. Although there was no evidence of occipital–C1 instability, instrumentation and fusion were extended up to the occiput because of the prior C-1 laminectomy. Despite preoperative traction, complete reduction of C1–2 could not be achieved.

**Second Postoperative Course.** The patient had an uncomplicated postoperative course and was discharged home on postoperative Day 3 with a hard collar for 3 months. She returned for 1-year follow-up with complete resolution of her myelopathy and radiographic resolution of intramedullary T2 hyperintensity and contrast enhancement (Figs. 1C and 2C).

**Case 2**

**History and Examination.** After receiving an outside diagnosis of unresectable cervicomedullary spinal cord tumor, this 8-year-old girl with cerebral palsy presented wheelchair-bound with progressive spastic quadriplegia. On physical examination, she had significantly increased tone in her bilateral lower extremities, with hyperreflexia, clonus, and bilaterally upgoing toes. She was unable to ambulate.

Magnetic resonance imaging demonstrated focal intramedullary enhancement and T2 hyperintensity from the cervicomedullary junction to C-3 (Figs. 4 left and 5 left). An os odontoideum was suspected and confirmed by CT scanning, and flexion-extension radiographs demonstrated instability (Fig. 6).

**Operation.** The patient underwent posterior C1–2 instrumented fusion with iliac crest autograft. She was discharged home after a brief, uncomplicated postoperative course and remained in a hard collar for 3 months.

**Postoperative Course.** The patient returned for her 3-month follow-up with significant clinical improvement. On physical examination, she had improvement in tone, hyperreflexia, and strength in the lower extremities bilaterally. She was able to ambulate short distances with assistance. Magnetic resonance imaging demonstrated marked reduction in edema at the cervicomedullary junction, with reduced T2 signal and cord thickening (Fig. 5 left). Contrast-administered T1-weighted sequences demonstrated a reduction in contrast enhancement (Fig. 4 left).
Os odontoideum mimicking intramedullary spinal cord tumor

Discussion

Atlantoaxial instability in the pediatric population is commonly associated with trauma or congenital disorders such as Down syndrome, osteogenesis imperfecta, neurofibromatosis, and Klippel-Feil syndrome. These children can develop instability because of hypoplasia of the odontoid process, laxity of the transverse ligament, or other bony abnormalities. The development of neurological symptoms in these at-risk populations, which range from neck pain and dysesthesias to tetraplegia, often leads to evaluation for atlantoaxial or craniocervical instability early in the course of decline. Timely imaging thus reflects acute to subacute cord injury, demonstrating little or no T2 cord signal change.8,13

The present cases demonstrate the consequences of chronic atlantoaxial instability and cord injury in non-syndromic children whose os odontoideum remained undiagnosed. In these cases, T2 hyperintensity and contrast enhancement at the cervicomedullary junction were mistaken for an intramedullary tumor, and the os odontoideum was overlooked. Because MR imaging is the imaging modality of choice in the setting of myelopathy, the initial identification of an intramedullary enhancing lesion may lead to a presumed diagnosis of intramedullary tumor. However, instability should always be considered in the differential diagnosis of a lesion at the cervicomedullary junction. Flexion-extension plain radiography and CT scanning of the cervical spine are the ideal modalities to assess for os odontoideum, other fractures, or instability.14 In retrospect, review of the initial MR images in Case 1 demonstrated an increased clivus-canal angle (approximately 115°), which may be useful as a clue to prompt additional investigations including dynamic imaging.

Perhaps most critical to our patients’ misdiagnoses of spinal cord tumor is the understanding that MR imaging findings in chronic cord compression can mimic those of other pathological neoplastic and inflammatory processes, despite little to no data in the pediatric population. Lee et al.9 reported 6 cases of chronic cervical spondylosis in an adult population causing both extensive T2 hyperintensity and contrast enhancement at the site of maximal cord compression; notably, T2 signal and cord thickening resolved postoperatively in 4 of the 6 patients. Although no intramedullary biopsy was performed in the patient in Case 2, the resolution of cord signal on MR imaging and the patient’s marked clinical improvement following instrumented fusion indicate that her progressive decline on presentation was entirely attributable to os odontoideum–related C1–2 instability.

Conclusions

Children who present with progressive myelopathy
should be carefully investigated for os odontoideum, because chronic trauma due to C1–2 instability can produce focal intramedullary enhancement and cord edema that may mimic an intramedullary neoplasm. If instability is found, C1–2 stabilization should be considered before other intradural procedures are pursued.

Disclosure

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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