Self-mutilation in a child with a tethered spinal cord

Case report

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Self-mutilation after deafferentation injuries has rarely been reported in humans. The authors report on a 16-year-old girl who was born with a myelomeningocele. In adolescence it was noted that concurrent with her spinal cord becoming symptomatically tethered she began to self-mutilate her digits. A rare manifestation of the tethered spinal cord may be dysesthesias that led to self-mutilation.

KEY WORDS • dysesthesia • meylomeningocele • spina bifida aperta • spinal dysraphism • paresthesia • pain • self mutilation

SELF-MUTILATION, or autotomy, after deafferentation injuries such as brachial plexus trauma has rarely been reported in humans; however, this finding is more common with pathological entities such as congenital analgesia.

Case Report

History. This 16-year-old girl was born with an in utero–diagnosed myelomeningocele. She was the product of a 40-week gestation and was delivered by cesarean section. Her Apgar scores at birth were 8 at 1 minute and 9 at 5 minutes. A large thoracolumbar myelomeningocele was associated with a midline area of focal hirsutism at its superior aspect. The child developed hydrocephalus and subsequently underwent placement of a ventriculoperitoneal shunt. Perinatally, she was noted to have a left pelvic kidney and a normally positioned right kidney with a duplicated right ureter. At the age of 12 years the patient complained of severe radicular chest and back pain that were unresponsive to narcotic agents. These dysesthesias were so significant that chronic use of a heating pad caused severe burns over her anterior chest wall. Also during this time period, it was noted that she began chewing on her finger tips and eventually autoamputated the distal third of the distal phalanx of the right third through fifth digits. The distal phalanx of the right first digit was completely removed by biting. Subsequently, she developed osteomyelitis of the right first digit that required intravenous antibiotic treatments. The results of self-mutilation were also evident on the distal phalanx of the left second and third digits. Magnetic resonance imaging of the lumbosacral cord demonstrated a dorsally adherent distal tethered spinal cord (Fig. 1 left). The patient attends mainstream classes in school and has passing grades. She has no history of neuropsychological disturbances.

Initial Operations. Following a detethering procedure of her spinal cord both her radicular chest and back pain resolved, and her self-mutilation behavior ceased. Three months following a ventriculoperitoneal shunt revision for headache she developed a Staphylococcus aureus shunt infection.

Subsequent Operations and Postoperative Course. Two years later, this patient again developed severe radicular chest pain and was again noted to begin gnawing her fingertips. A second detethering procedure was performed and soon after her symptoms abated. At the second operation for spinal cord detethering it was noted that the cord was significantly adherent to it surroundings, and the long-term success of this procedure was questioned. One year following her second detethering procedure, she is without the symptoms of radicular chest pain and does not engage self-mutilation behaviors. Similarly, Myles, et al., have reported on three patients who self-mutilated following spinal cord procedures (Fig. 2).

SWEET9 has stated that self-mutilation is not uncommon in certain young children who suffer congenital analgesia. These patients typically gnaw their fingers, lips, and tongue. LEVITT has theorized that in these cases self-mutilation may be treated successfully with anticonvulsant medications. This study was based on three patients. TARVIN and Prata10 have shown that self-mutilation of the hindlimbs, tail, perineum, anal region, and genitals is seen in certain young children who suffer congenital analgesia. Albe-Fessard, et al., have reported on an adult patient with a myelomeningocele who mutilated her fingers. At operation, one hemicord was found to be tethered. Dahlin, et al., have reported on an adult patient with ophthalmic herpes zoster that autoamputated her right first digit when cured of his pain by a dorsal horn lesion, never engaging in self-mutilatory behavior again. Procacci and Myles6 have reported on two cases of autotomy in ventilator-dependent children with myelomeningocele. These two cases of autotomy were attributed to the splitting of the transverse processes following anterolateral cordotomy, and the self-mutilation behaviors (Fig. 1 right) remained small, with no change following both detethering procedures (Fig. 2).
Self-mutilation by a child with tethered cord

Our patient presented with tethered spinal cord and self-mutilatory behaviors. Similarly, Myles, et al.,6 have reported on a 12-month-old girl with a cervical split cord malformation who mutilated her fingers. At operation, one hemicord was found to be tethered. Dahlin, et al., 4 have reported on two adult men with complete spinal cord injury at C-4 who exhibited multiple finger amputations in the absence of psychosis. Colville and Mok3 have reviewed two cases of lip biting in ventilator-dependent children with spinal cord injury. These two cases of autotomy were attributed to psychological stresses. Vogel and Anderson11 concluded that patients who self-mutilate following spinal cord injury may be treated successfully with anticonvulsant medications. This study was based on three patients. Tarvin and Prata10 have shown that self-mutilation of the hindlimbs, tail, perineum, anal region, and genitals is seen in some dogs with severe lumbosacral stenosis. Decompression of the stenotic canal alleviated the clinical signs in all cases. In monkeys, Levitt5 has theorized that self-mutilation following anterolateral cordotomy is due to dysesthesias produced by this procedure.

Conclusions

Although seemingly quite rare in humans with tethered spinal cord, self-mutilation may be found. Patients with this rare symptom should be evaluated for tethered cord syndrome.

References

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SEUDOANEURYSMS involving the intracranial VA, an increasingly recognized cause of SAH, are complex lesions with a threatening natural history. Because the vessel injury is both transmural and circumferential in nature, these lesions do not typically lend themselves to conventional clip-assisted arterial reconstruction. Moreover, despite numerous recent reports in which successful endoluminal stenting for complex posterior circulation aneurysms has been documented, long-term follow-up data are not yet available and limitations of this therapy have not been fully elucidated. As such, current treatment approaches for these lesions often rely on hunterian strategies with or without cerebral revascularization. Segmental isolation of the lesion through trapping, either surgically or by endovascular means, is the treatment of choice because it definitively uncouples the aneurysm from the intracranial circulation, thereby preventing further growth and the risk of future hemorrhage.

The treatment of dissecting VA pseudoaneurysms involving the origin of the PICA with a trapping procedure poses particular challenges, because PICA revascularization is generally required to avoid brainstem and cerebellar ischemia. Several different approaches to PICA revascularization have been described in the neurosurgical literature, the least common of which is PICA reimplantation (Table 1). In this paper, we report on the case of a pediatric patient with a ruptured traumatic VA pseudoaneurysm from which the PICA emerged. The patient was treated with surgical trapping of the aneurysm and direct reimplantation of the PICA origin distal to the diseased vessel segment, resulting in an excellent clinical and angiographically documented outcome. Based on a review of the literature, we are aware of only two other previously documented cases of direct PICA reimplantation in the treatment of a complex VA aneurysm: only one of which involved a VA–PICA anastomosis and neither of which occurred in a child. In addition to reporting this extremely rare case and discussing features of commonly used PICA revascularization strategies, we briefly review the existing literature in which PICA reimplantation has been performed as an adjunctive measure in the treatment of complex VA aneurysms.

**Case Report**

**Clinical Presentation.** This 13-year-old boy with no significant medical history sustained an accidental gunshot wound through the mouth. He was transported to our institution on the paediatric intensive care unit.

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