CSF goes astray no matter what is in the way: occipital intradiploic pseudomeningocele and de novo syringomyelia following foramen magnum decompression for Chiari I malformation in a pediatric patient

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Nontraumatic intradiploic pseudomeningoceles and de novo syringomyelia formation are very rare entities. The authors have previously reported the case of a 4-year-old girl who underwent foramen magnum decompression without dural closure for Chiari I malformation. Three years after the operation an intradiploic pseudomeningocele was documented, but the patient was lost to follow-up without undergoing revision surgery. Four years later, at the age of 11 years, the patient returned for treatment of intensifying symptoms. Radiological imaging then showed an increase in the size of the intradiploic pseudomeningocele and a new cervical syrinx. The patient underwent a first revision surgery in which a part of the internal layer of the occipital bone was removed and arachnoid scar lysis was performed. Two months later the syrinx had worsened, and in a second revision surgery a pseudomeningocele-peritoneal shunt was placed. Here, the authors describe what is to their knowledge the first case of an intradiploic pseudomeningocele and de novo syringomyelia formation following Chiari I decompressive surgery.

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MOST cases of the rarely occurring occipital intradiploic pseudomeningoceles are of traumatic or idiopathic origin,^{1,5,17,27} and only a few reported cases have occurred postoperatively.^{3,11,23,24,28,31} Postoperative development of syringomyelia has been mainly described following spinal CSF drainage,^{7,14,26,30} and anecdotally after pleural effusion aspiration³² and after surgery for spontaneous subdural hematoma.² There is to our knowledge only one previous study of de novo syrinx formation, which included 7 pediatric patients presenting with de novo syrinx after decompressive surgery for Chiari I malformation.²⁵ We have previously reported the case of a 4-year-old girl who had developed an occipital intradiploic pseudomeningocele following decompressive surgery for Chiari I malformation without dural closure. After under-

going radiological imaging, this patient was lost to follow-up without having revision surgery.¹⁹ Then, after 4 years of loss to follow-up, she presented with an increase in the size of her intradiploic pseudomeningocele and a new cervical syrinx. We herein report the surgical revision performed in this patient and discuss possible underlying causes of both complications. To our knowledge no case of iatrogenic intradiploic pseudomeningocele and de novo formation of a syrinx after surgical treatment for Chiari I malformation has been described previously.

Case Report

History and Examination

This female patient first presented at the age of 3 years

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FIG. 1. Sagittal T2-weighted MR image showing the patient's Chiari I malformation (preoperative).

with complaints of headache, pins and needles predominantly in her upper limbs, and hypersensitivity to heat. One year later, at the age of 4 years, she underwent an MRI examination that showed a Chiari I malformation with the cerebellar tonsils descending to C2 but without a syrinx (Fig. 1). The MRI did not show signs of hydrocephalus. The patient's neurological status was normal. Foramen magnum decompression with a limited suboccipital craniectomy of 2.5×1.5 cm and partial resection of the posterior arch of C1 was performed, followed by a wide durotomy and opening of the arachnoid. While the dura was left open, meticulous closure of the muscles, fascia, subcutaneous tissue, and skin was done. Three years later, when the patient was 7 years old, she returned for treatment due to the reappearance of her preoperative symptoms. MRI was performed that showed an occipital intradiploic pseudomeningocele (Fig. 2). After this imaging was performed, the patient was lost to follow-up without undergoing revision surgery. She returned for treatment 4 years later because of the intensification of her preoperative symptoms, pins and needles with altered sensation in her limbs, which were suggestive of spinal cord dysfunction. MRI was performed that showed increased size of the intradiploic pseudomeningocele and a new cervical syrinx extending from C2 to C3 (Fig. 3). At this time the patient's neurological examination was again normal.

FIG. 2. Sagittal T1-weighted MR image obtained 3 years after the first operation demonstrating a hypointense occipital collection with mass effect on the cerebellum.

Revision Surgeries

The patient underwent a first revision surgery when she was 11 years old (7 years after undergoing foramen magnum decompression without dural closure). We exposed the small defect of the occipital cortical bone that had been created during the first operation. The bony defect lay directly over the small opening of the foramen of Magendie. We partially excised the inner layer of the occipital bone housing the intradiploic pseudomeningocele to restore space in the posterior fossa, and then performed arachnoid scar lysis. Meticulous wound closure in 4 layers was performed without dural closure, as in the patient's first operation.

An MRI examination performed 2 months after the first revision surgery showed worsening of the syrinx to involve the whole cervical spinal cord and spinal cord edema extending to the conus (Fig. 4). In a second revision surgery we placed a pseudomeningocele-peritoneal shunt with an adjustable pressure valve (Strata, Medtronic; pressure setting at performance level 2.5).

Postoperative Course

The patient's postoperative course was uneventful, and she was discharged home with a normal neurological examination. Her preoperative symptoms had resolved and

FIG. 3. Sagittal T2-weighted MR image obtained 7 years after the first operation. The hyperintense occipital fluid mass has increased and a new syrinx extending from C2 to C3 is visible.

FIG. 4. Sagittal T2-weighted MR image obtained 2 months after revision surgery. The syrinx spans the whole cervical spine and edema in the spinal cord extends to the conus.

her sensory symptoms had improved. The MRI performed 1 year after the shunt placement showed a significant decrease of the syrinx (Fig. 5).

Discussion

Multiple methods exist for decompressive surgery for symptomatic patients with Chiari I malformation. Foramen magnum decompression with or without partial removal of the posterior arch of C1 is generally accepted. However, there is still controversy as to whether for decompression a durotomy or a durotomy with subsequent duraplasty has to be performed in these cases.^{4,6,8,12,13,15,20–22,36,37} Yet another option is decompressive surgery without dural repair.^{10,16} The bony decompression is followed by durotomy, with opening of the arachnoid and resection of visible adhesion, and the dura is then intentionally left wide open. The reason for not closing the dura is to ameliorate the CSF flow by creating a pseudomeningocele that enlarges the cisterna magna.^{34,35} The use of alternative methods, such as primary dural closure, duraplasty, or a patch that is subject to failure may partly neutralize the maximal decompressive effect of the surgical procedure.

We have previously reported that in addition to the child in the present case, 6 patients with Chiari I malformation have been described thus far who postoperatively devel-

oped an occipital intradiploic pseudomeningocele.^{18,19} Three of these patients had undergone surgery with duraplasty,^{3,11,23} and 3 without dural closure.^{18,19} To the best of our knowledge we present here the first report of a patient who initially developed an occipital intradiploic pseudomeningocele and then a de novo cervical syrinx following decompressive surgery for Chiari I malformation.

The formation and the size of an intradiploic pseudomeningocele might depend on the composition of the diploe at the place where it is opened on the occasion of the primary foramen magnum decompression. In this context the participation of the diploe in the draining of CSF into the venous system^{15,29,33} should be considered. The opened diploe comes into contact with the pulsating force of the CSF with its water hammer effect. When the diploe is opened where there is little or no CSF drainage, the internal and external layers of the occipital bone might splay, leading to the buildup of an intradiploid fluid collection. However, if the diploe is opened where there is adequate CSF drainage the diploe may not widen or may widen just a little. This mechanism might explain why the patients with a postsurgical occipital intradiploic CSF collection reported by Bulleid et al.³ and another patient we previously reported¹⁹ complained of low-pressure headaches and showed only slight splaying of the diploe.^{3,19}

FIG. 5. Sagittal T2-weighted MR image demonstrates the diminution of the syrinx 1 year after placement of a pseudomeningocele-peritoneal shunt.

The formation of an intradiploic pseudomeningocele might be averted by meticulous sealing of the opened diploe when it is violated during the suboccipital craniectomy. But does closure of an opened diploe years later at revision surgery always resolve the clinical symptoms?

To our knowledge, only 2 patients with intravertebral CSF collections have been reported so far.^{9,18} Gandhoke et al.⁹ first reported a patient with Chiari I malformation who developed a collection of CSF in C2 after foramen magnum decompression with expansile duraplasty. This patient had been asymptomatic for 14 years. At revision surgery a dural opening that communicated with the right lamina of C2 was found below the former duraplasty. The surgeons closed the dural defect and reinforced the inner and outer layers of the bony cavity with demineralized bone matrix. One week after surgery the patient presented with symptoms of elevated intracranial pressure (headache, nausea, and vomiting). Imaging showed a small pseudomeningocele without ventricular enlargement. The patient's symptoms were only relieved with CSF diversion, first performed with lumbar drainage and then with a ventriculoperitoneal shunt. Gandhoke et al.⁹ concluded that over the years the communication between the dural opening and the intraosseous fistula had built up an accessory CSF drainage pathway that was obstructed

at revision surgery. We reported elsewhere a female patient who had developed a significant intraosseous C2 CSF collection and a very small suboccipital intradiploic CSF collection.¹⁸ She complained of headaches and vomiting 5 years after Chiari I decompressive surgery. At revision surgery we filled the hollow space of C2 with a bioactive bone graft substitute, closed the bony defects of the suboccipital bone and C2 with bone wax, and resected the arachnoid adhesions. After revision surgery our patient remained asymptomatic and did not require further treatment despite the blockage of 2 potential additional sites of CSF absorption. The difference between our previously reported patient¹⁸ and the patient reported by Gandhoke et al.⁹ may have been that before the foramen magnum decompression Gandhoke et al.'s patient had an underlying compensated hydrocephalus that became clinically relevant after the revision surgery and led to an obstruction of the additional CSF absorption pathway that had developed over the years.

Only a few cases of iatrogenically induced syringes have been described so far. In 2013 Naftel et al. reported 7 pediatric patients with de novo syrinx after decompressive surgery for Chiari I malformation.²⁵ The syringes occurred between 0.2 and 4.3 years after the first operation. All patients had undergone suboccipital craniectomy and C1 laminectomy, 4 of them had undergone tonsil resection or coagulation, and all underwent duraplasty. Operative findings at the revision surgery were described as scarred arachnoid, arachnoid webs, intradural scarring, and fibrous bands at previous C1-laminectomy sites.

The patient we present here developed a large occipital intradiploic pseudomeningocele in the first 3 years after foramen magnum decompression and then also developed a syrinx extending from C2 to C3 during the 4th and 7th years after the first operation. The formation of a large occipital intradiploic CSF collection may be a sign that the diploe was violated at a place where there is no or only a small amount of CSF drainage. The late induction of the syrinx over the years may have been due to postoperative arachnoid scarring after the first surgery. Another reason for the formation of the syrinx might have been an underlying component of hydrocephalus that was not sufficiently drained by the additional intradiploic CSF draining site.

In case of hydrocephalus or elevated intracranial pressure, sealing of the opened suboccipital diploe at the primary surgery or at the first revision surgery might not have prevented the formation or the aggravation of the syrinx. The increase of the syrinx after the first revision surgery might have been due to progressive arachnoid scarring provoked by arachnoid scar lysis or the persistence of underlying hydrocephalus or elevated intracranial pressure. In this case the lysis of arachnoid adhesions might not have addressed the filling pressure of the syrinx without CSF diversion.

Although the preoperative MRI did not show radiological signs of hydrocephalus in this case, we cannot formally rule out compensated hydrocephalus. The patient did not undergo intracranial pressure monitoring. Subsequent pseudomeningocele-peritoneal shunting seems to have addressed the elevated pressure or hydrocephalus, resulting

in the resolution of preoperative symptoms and a significant decrease of the syrinx.

Conclusions

The appearance of an intradiploic pseudomeningocele or low-pressure headache after foramen magnum decompression might be avoided by meticulous sealing of the opened diploe. The formation of a syrinx after decompressive surgery for Chiari I malformation might be an indicator of preexisting hydrocephalus or elevated intracranial pressure. Intracranial pressure monitoring in Chiari I malformation patients prior to foramen magnum decompression might detect patients who need CSF shunting.

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Disclosures

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Author Contributions

Conception and design: both authors. Acquisition of data: both authors. Drafting the article: Kurzbuch. Critically revising the article: Magdum. Reviewed submitted version of manuscript: both authors. Approved the final version of the manuscript on behalf of both authors: Kurzbuch.

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