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# *Adult-onset Sacral Meningocele Causing a Specific Headache Triggered by Compression or Adoption of a Sitting or Supine Posture*

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## Abstract

**We report a rare case of adult-onset sacral meningocele where compression triggered a specific headache. A 46-year-old woman presented with a headache, which worsened when she was in a sitting or supine position. A subcutaneous mass was observed on her left buttock, the compression of which also induced headache. No neurological deficits were evident. Lumbar and sacral magnetic resonance imaging demonstrated a meningocele in the left dorsal buttock, connecting to the sacral cerebrospinal fluid (CSF) space, and spinal computed tomography revealed sacral dysplasia. Initial meningocele resection improved the patient's headache, but the cyst recurred 2 years later. Following repeated surgery to reinforce the meningocele orifice, the headache was relieved and has been absent for more than 6 years. The headache was due to intracranial pressure fluctuations due to CSF influx into and drainage from the meningocele. Meningocele development in adulthood can be owing to a spinal bone defect and pressure load on the spinal dura. Surgical resection can improve symptoms resulting from meningocele, and reinforcement of the orifice using an artificial surgical membrane effectively prevents recurrence.**

Keywords: adult-onset, meningocele, headache, supine posture

## Introduction

Latent spina bifida is commonly diagnosed before puberty, exhibiting neurological symptoms caused by tethering,[1,2] and has a reported prevalence of 15%-23%.[3,4] Meningocele diagnosed in adulthood is rare but may develop due to defects in the bony spine, probably caused by cerebrospinal fluid (CSF) pressure loading. Here, we report an adult case of meningocele causing a characteristic, orthostatic headache.

## Case Report

A 46-year-old woman was referred to our hospital because of a 1-month history of stabbing headaches triggered by adopting a sitting or supine posture. The headache also occurred when standing up in the morning. She had no particular medical history or developmental abnormalities. Concurrently, she also noticed a soft mass on her left buttock, which when pressed triggered the headache. She had no neurological deficits or problems with urination. Deep tendon reflexes in her extremities were normal. A soft subcutaneous mass of 4 cm diameter was confirmed in her left upper gluteal cleft, but the skin surface was normal without abnormal hair or pigmentation. Manually compression of the mass or adopting a supine or sitting position triggered the headache. Magnetic resonance imaging (MRI) of the sacral region revealed a 4-cm-diameter mass with a homogeneous high-intensity signal on T2-weighted images located slightly to the left of the dorsal sacral median (Fig. 1A and B). The anteroposterior diameter of the mass appeared smaller on MRI than measured via physical palpation, probably due to the patient being in a supine position. The margin was clear, and no neural tissue or neoplastic lesion was evident in the mass. MRI also showed a tethered spinal cord but without intramedullary syrinx or lipoma. Computed tomography (CT) of the lumbar and sacral spine showed spina bifida and dysplasia of the bone in the sacral region (Fig. 1C). CT and MRI of the head showed no abnormality. Based on these findings, the

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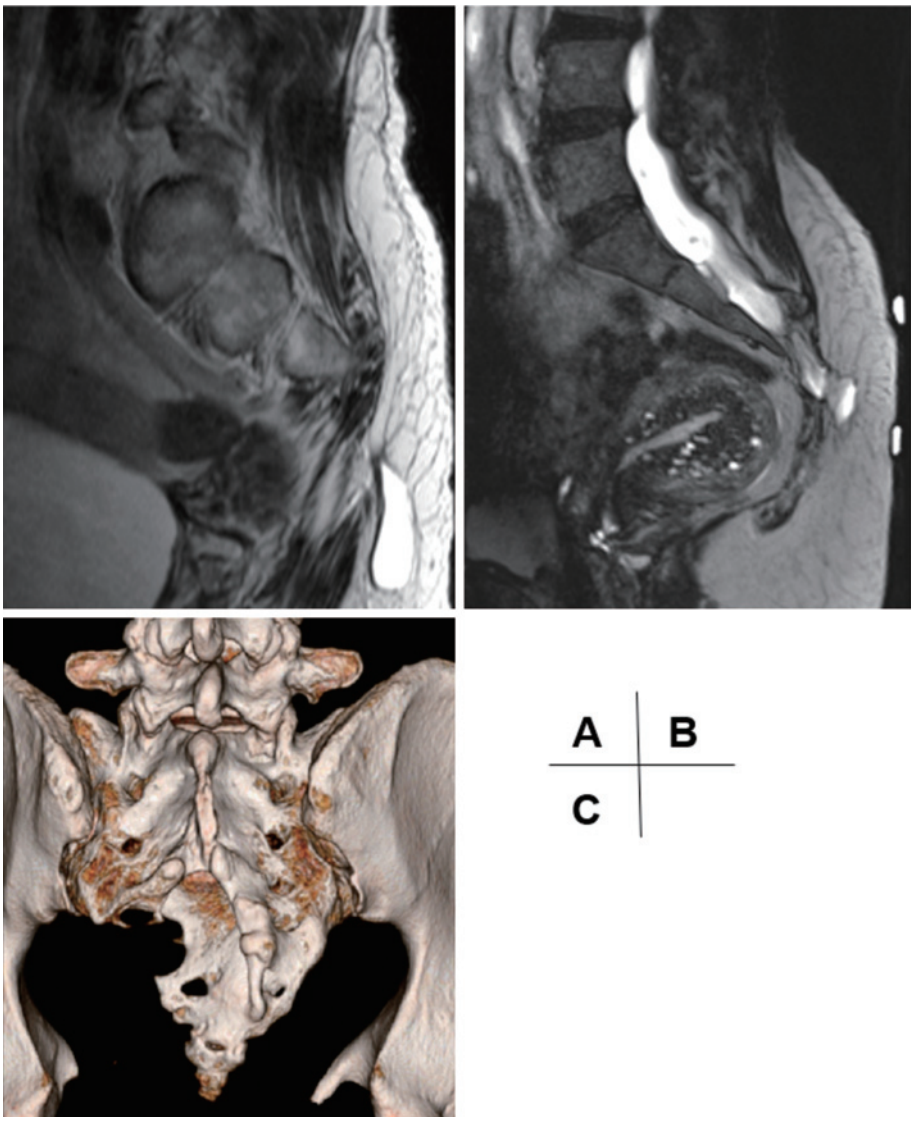


**Fig. 1 A: Sagittal lumbosacral magnetic resonance imaging T2-weighted image showing a homogeneous, high-signal subcutaneous mass of 4 cm in diameter in the left upper gluteal cleft. The anteroposterior diameter of the mass appeared smaller than that measured with palpation, probably due to the patient being supine. B: The mass is contiguous from the dural sac of the sacral spine without any internal neural tissues, suggesting a diagnosis of meningocele. C: Three-dimensional computed tomography scan of the pelvis showing a defect of the left vertebral arch at the S4 and S5 levels and slight rightward deviation of the vertebral body.**

patient was diagnosed with meningocele accompanying occult spina bifida.

Upon initial surgery, ligation and excision of the meningocele were performed via a 4-cm skin incision. Intraoperative observation revealed a pulsatile mass covered with dura mater protruding from the sacral defect without internal neural structures. Pathological examination of the resected nodular structures demonstrated that they comprised collagen fibers, blood vessels, and a fatty stroma, consistent with the preoperative diagnosis of meningocele. The patient's postoperative course was uneventful, and her headache was relieved, regardless of whether she was sitting or lying supine (Fig. 2A).

However, 2 years after the initial surgery, the headache recurred when the patient was supine, and a subcutaneous mass also reappeared in the left buttock. Lumbar and sacral MRI revealed meningocele recurrence (Fig. 2B), and a second operation was performed. This time, the skin incision was widened to 7 cm, and the recurrent meningocele was similar to that observed in the first operation. The orifice of the meningocele was closed and sutured, and then the lateral and medial walls of the meningocele capsule (dura mater) were overlaid and further sutured for reinforcement. In addition, the sutured orifice and exposed dural canal were covered with a piece of Gore-Tex surgical membrane (W.L. Gore and Associates Inc., Newark, USA),

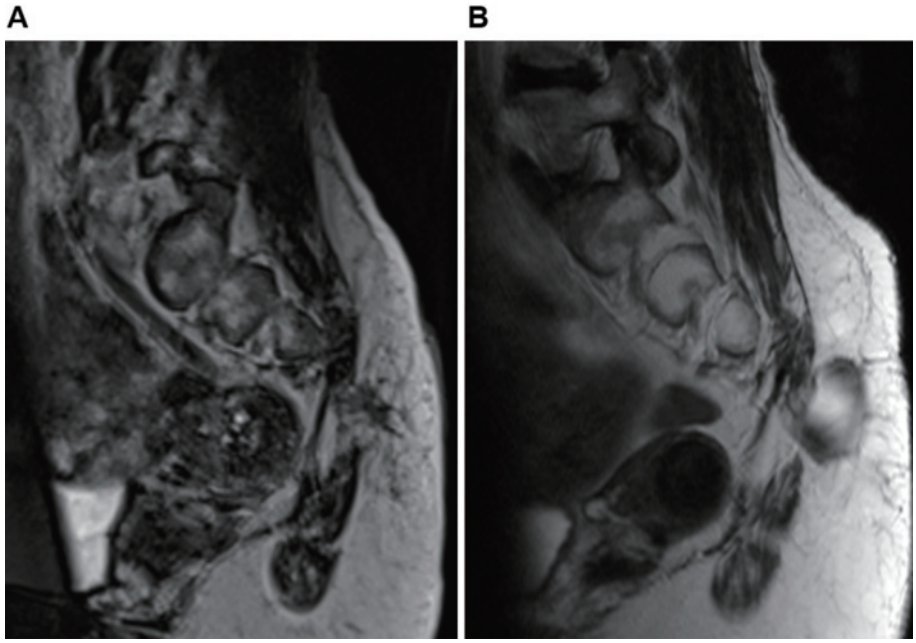


**Fig. 2 A: Lumbosacral magnetic resonance imaging (MRI) T2-weighted sagittal image after the first surgery. The meningocele was completely resected. B: Sagittal lumbosacral MRI T2-weighted image 2 years after the first surgery showing recurrence of the meningocele protruding from the sacral spine.**

the edge of which was sutured tightly to the surrounding fascia and periosteum for additional reinforcement. The patient's headache immediately disappeared following the repeated surgery. Thereafter, her postoperative course was uneventful. During follow-up for more than 6 years, the headache has not recurred, and MRI has not shown any recurrence of the meningocele.

Written informed consent for publication of the clinical details was obtained from the patient.

## Discussion

In our reported case, the headache was triggered by pressure on the meningocele located in the lateral sacral region when the patient was sitting or supine, or when direct manual compression was applied, including upon standing in the morning. Temporary increased intracranial pressure due to the influx of CSF within the mass into the subarachnoid space seemed to cause the headache. Alternatively, CSF flowed into the meningocele when the patient stood up, leading to CSF hypovolemia symptoms. Due to the occult spina bifida in the sacral region, the dura would have lacked any supporting bony structure, and thus, would have been subjected to a relatively heavy pressure load from the orthostatic CSF. Under these circumstances, the meningocele would have gradually manifested in middle age. Accordingly, reinforced repair of the meningocele was required, as performed in the second operation.

As the patient's headache caused by compression of the meningocele repeatedly appeared with reliable reproducibility, further examinations were not performed in this case. For objective examination, a lumbar puncture to determine intracranial pressure can be useful, with or without compression of the meningocele. As previously reported, a method for determining intracranial compliance with MRI[5] would have helped confirm the diagnosis and pathophysiology when performed with and without decompression of the meningocele by applying cushions around it.

The overall reported prevalence of occult spina bifida is 15%-23%.[3,4] As reflected in the name, it is usually an asymptomatic entity diagnosed only occasionally. Meningocele acquired or manifested in adulthood due to spina bifida is rare, and only a few cases have been reported (Table 1). Barazi et al.[6] reported two adult women with meningocele that acted as a CSF "sump," leading to changes in intracranial pressure depending upon the patient's position and resulting in symptoms, such as headache. Similar to the present case, both patients suffered headaches when supine or mechanical pressure was applied to the meningocele mass. One of the patients had been followed up conservatively for 2 years, but the meningocele ruptured when she fell downstairs. Thereafter, she developed low CSF pressure syndrome, and her symptoms changed to severe orthostatic headache while standing due to decreased intracranial pressure. Both patients underwent surgical treatment for the meningocele, and their symptoms were completely resolved.

Versteegh et al.[7] also reported a 24-year-old woman with

**Table 1 Previous cases of adult-onset meningocele with sacral bone defects**

| Case | Sex | Age (years) | Symptoms | Location | Surgery |
|---|---|---|---|---|---|
| 1[6] | F | 44 | Headache upon meningocele compression, nausea, and visual disturbance | Posterior sacral | yes |
| 2[6] | F | 28 | Headache upon meningocele compression and visual disturbance | Posterior sacral | yes |
| 3[7] | F | 24 | Headache when riding a bicycle | Anterior sacral | yes |

All patients became symptom-free after surgical resection.

Clarino syndrome and a meningocele in the anterior sacral region. She also suffered from a headache when riding a bicycle, and this was resolved by surgical treatment. Compression of the mass when sitting or riding a bicycle led to increased intracranial pressure and headache. Interestingly, all of the reported patients with meningocele acquired or manifested in adulthood were women. Differences in muscle volume, connective or adipose tissues, or pelvic structure from men could be possible reasons, but further case accumulation and investigation will be necessary.

In addition, there have been several reports of pseudomeningocele after spinal surgery. De Luca et al.[8] reported pseudomeningocele formation after spinal surgery with L5/S1 unilateral laminectomy, causing intracranial hypotension and headache fluctuating with position and was improved after surgical resection and repair. This also supports the notion that meningocele in adulthood may appear in association with spinal bone defect and pressure load, causing a headache due to fluctuations in intracranial pressure. The pressure load may cause further bone erosion or scalloping in the sacral region, possibly leading to meningocele recurrence, thereby necessitating long-term observation. The patient has been followed up in the outpatient department for over 6 years, and further follow-up will continue.

Surgical intervention allowing resection is essential to treat acquired or surgically induced meningocele, and reinforced dural repair is important because of the dynamic influence of orthostatic increased CSF pressure.

## Conclusion

We have reported a case of meningocele manifested in adulthood as a characteristic headache due to fluctuations in intracranial CSF pressure. Surgical resection with secure dural repair and reinforcement is essential to improve symptoms and prevent the recurrence of such cases.

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## Conflicts of Interest Disclosure

All authors have no conflicts of interest to declare.

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