Suboccipital meningocele presenting as a huge retropharyngeal mass in a patient with neurofibromatosis Type 1

Case report

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The authors report an extremely rare case of neurofibromatosis Type 1 (NF1) with a suboccipital meningocele presenting as a huge retropharyngeal mass. A 73-year-old woman with typical cutaneous manifestations of NF1 presented with nasal obstruction and dysphagia due to a retropharyngeal mass. Magnetic resonance imaging revealed a huge mass lesion extending from the right occipital bone defect to the retropharynx through the right paravertebral region. Computerized tomography scanning after intrathecal administration of contrast material confirmed that the mass was a meningocele protruding through a right occipital bone defect. The authors attempted to ligate this meningocele, most of which was excised via a suboccipital approach, but a second transcervical operation was required. Finally, the meningocele resolved and the patient was discharged without symptoms.

KEY WORDS • neurofibromatosis Type 1 • suboccipital meningocele • retropharyngeal mass

EUROFIBROMATOSIS Type 1 (NF1) is one of the most commonly inherited disorders of the mesodermal and neuroectodermal structures, affecting approximately one in 4000 individuals. Neurofibromatosis Type 1 results in a complex cluster of developmental and tumor syndromes that include neurofibromas, hyperpigmentation, bone anomaly, and hamartomas of the iris. Some patients with NF1 may also have lesions, such as optic gliomas, dural ectasia, and aqueductal stenosis. Meningocele, a less frequently associated lesion in patients with NF1, is the result of dural ectasia. This entity most commonly occurs in the thoracic spine and rarely occurs in other areas. We report the case of a patient with NF1 who harbored a huge suboccipital meningocele that required two surgical extirpations, and we discuss this rare clinical entity.

Case Report

History. This 73-year-old right-handed woman with typical cutaneous manifestations of NF1 first noticed a nasal quality to her voice and nasal stuffiness 6 months before admission. The patient consulted her general practitioner and was treated for common cold. Because her symptoms were progressive and she noticed difficulty in swallowing for 2 weeks, she consulted our outpatient clinic.

Examination. On examination, the patient had numerous neurofibromas all over her body as well as multiple café-au-lait spots. She had a soft retropharyngeal mass and a soft posterior neck mass, but she was neurologically intact. Magnetic resonance (MR) imaging disclosed the existence of a huge mass lesion extending from the retropharyngeal region across the right paravertebral region to the suboccipital area (Fig. 1). Computerized tomography (CT) cisternography (Fig. 2) revealed a dramatic accumulation of contrast material in the mass lesion, confirming the diagnosis of meningocele. Three-dimensional CT scanning of the head (Fig. 3) demonstrated a bone defect at the right occipital bone adjacent to the foramen magnum.

First Operation. Surgical treatment was performed via a suboccipital approach. A U-shaped suboccipital skin incision was made on the right side. Edematous subcutaneous tissue and deep muscles were dissected around the meningocele, after which we recognized an occipital bone defect and the stalk of the meningocele. The posterior arch of the atlas was exposed and circumferential dissection around the stalk was attempted. The stalk of the meningocele was large and extended anterolaterally to the right occipitoatlantal joint. The dura was intact at the stalk; however, most of the meningocele wall lacked dura mater and the wall was very fragile. Because the cerebrospinal fluid (CSF) easily leaked through the dissected meningocele...
wall and the ventral portion of the meningocele neck had been left behind, we opened the meningocele wall and inspected the vertebral artery and the lower cranial nerves intradurally. Finally, the meningocele was ligated using No. 2-0 silk suture at the orifice of the occipital bone defect with the help of an aneurysm needle and multiple hemoclips. Cranioplasty was performed using methylmethacrylate.

First Postoperative Course. The patient’s immediate postoperative course was uneventful. However, the retropharyngeal mass did not collapse and, unexpectedly, it gradually enlarged, aggravating the patient’s dyspnea on the 3rd postoperative day.

Second Operation. Because CT scanning demonstrated accumulation of CSF in the retropharyngeal mass, the patient subsequently underwent a second operation on the meningocele via a right-sided anterolateral–transcervical approach. We opened the retropharyngeal sac, aspirated the CSF, and obliterated the narrow connection between the suboccipital and retropharyngeal portions of the meningocele by using a few sections of muscle. A temporary lumbar drain was placed to facilitate healing and obliteration of the meningocele.

Final Postoperative Course. After the second procedure, the meningocele disappeared, along with the patient’s initial symptoms. The patient was discharged in good condition.

Discussion

Neurofibromatosis Type 1 is known to be associated with a spinal meningocele, which occurs most commonly in the thoracic spine. The high incidence of thoracic meningocele is considered to be caused by a pressure gradient between the subarachnoid space and the intrapleural space. Widening of the internal auditory canals and optic canals due to dural ectasia has rarely been reported in patients with NF1. Meningocele very rarely occurs in the cranium. To our knowledge, there is only one reported case in the English-language literature of a meningoencephalocele in a patient with NF1. Probst reported the case of a patient with NF1 who harbored bilateral frontobasal meningoencephaloceles and a CSF fistula. He observed multiple osseous and dural defects in the patient’s anterior skull base. This meningoencephalocele was ligated and the CSF leak was repaired, but preexisting hydrocephalus later became aggravated. The present case seems to be the first reported case of NF1 involving a suboccipital meningocele.

The pathogenesis of suboccipital meningocele is unclear. It is known that NF1 is sometimes associated with calvarial defects, most of which occur adjacent to the lambdoid suture. Our case had an occipital bone defect.
Suboccipital meningocele in NF1

There are two possible explanations for the development of this defect. One is that this condition results from defective embryological development in patients with NF1 or a congenital abnormality. Hunt and Pugh\(^4\) reported finding one large midline calvarial defect in a patient with NF1; they assumed that the defect was a bifid cranium. The other explanation is that this defect is acquired. It is known that a calvarial defect results from erosion by neurofibromatous tissue.\(^9\) In addition, Kaufman, et al.,\(^11\) stated that the normal but pulsatile pressure of CSF in the brain can lead to thinning of bone, resulting in the development of a skull defect. Therefore, one or more small bone defects could allow the gradual development of a meningoencephalocele. Recently, Macfarlane, et al.,\(^13\) presented the case of a patient with NF1 in whom typical sphenoid dysplasia with absence of the greater wing appeared to be acquired. These authors assumed that either the growth of the neurofibromatous tissue or a local vascular abnormality resulted in the local expansion of the superior orbital fissure, which became further enlarged by pressure from a concomitant increase in cerebral volume. In the present case, we assume that, based on the fragility of the dura and the possible occipital bifid cranium, a pulsatile CSF pressure and repeated neck motion might dilate and extend the dural sac, resulting in a large meningocele surrounding the upper cervical spine.

The goal of surgical treatment of a basal meningocele is neck ligation and resection of the sac. However, treatment is not easy because dural defects are multiple and the meningocele wall is very fragile, as previously described.\(^4,14\) In our case, the ventral portion of the meningocele neck was obscure from the suboccipital route. We thus opened the meningocele to identify the anatomical relationships between the meningocele sac and surrounding structures such as the vertebral artery, lower cranial nerves, and occipitoatlantal joint. However, it was difficult to obtain a watertight closure despite our meticulous attention to repairing the meningocele, and the retropharyngeal meningocele unexpectedly enlarged, possibly as a ball valve–type phenomenon. A second transcervical exploration was needed to obliterate the meningocele completely. In most circumstances, a simple dorsal approach should suffice if the meningocele neck is dorsal in location. However, a combined suboccipital–transcervical approach is useful for better visualization of the ventral portion of the meningocele neck and complete obliteration of this type of huge suboccipital meningocele extending to the retropharynx.

References

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