Cervical rudimentary meningocele in adulthood

Case report

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Almost all cases of cervical spinal dysraphism published to date have involved cystic lesions and were treated in very early childhood. The authors describe a unique case of a 21-year-old woman who harbored a solid cervical rudimentary meningocele. On preoperative CT and MR images, a cutaneous solid mass was shown to be connected to intraspinal contents by a stalk traversing the C-3 lamina defect. The authors resected the cutaneous mass and released the tethering neural band from the vertical axis of the spinal cord without causing injury. Pathological examination demonstrated a dense collagenous tissue containing clusters of meningocytes and psammoma bodies in the cutaneous mass. This rare entity, with a spinal dysraphism with a benign natural history, may contribute to the current classification of cervical spinal dysraphism.

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Key Words • cervical region • rudimentary meningocele • adulthood

Cervical dysraphism inside the spina bifida aperta is a rare disease, accounting for 1%–5% of all neural tube defects.1 In the literature, these lesions have been classified into various types based on the content of the cutaneous protuberance.11,12 Almost all cases of this congenital condition published to date are cystic in nature and were treated in very early childhood. Rudimentary meningocele, in which meningotheelial elements occupy most of a solid skin-covered mass in the midline posteriorly, is extremely rare in adults. We described a unique case of a 21-year-old woman who harbored a cervical rudimentary meningocele and in whom a surgical cure was achieved. The clinical presentation, operative findings, and distinct importance of this rare entity are discussed and the relevant literature is reviewed.

Case Report

The ethics committee of Hangzhou First People’s Hospital approved this study.

Abbreviation used in this paper: CDCT = cystic spinal dysraphism of the cervical and upper thoracic region.

History and Examination. This 21-year-old woman presented to the hospital with a posterior cervical region mass. The lesion had been detected immediately after the patient was born, and it gradually increased in size. The patient had no complaints of cervical pain or any fluid exudation from the mass. She requested an excision of the “tumor” for a cosmetic reason. Her family history was noncontributory. Physical examination revealed a posterior cervical midline protrusion measuring 4 cm in width, 4.5 cm in length, and 2.5 cm in height. The lesion manifested as an oval-shaped, hard, mobile mass without fluctuation. The top of the lesion was covered with purplish and poor-quality skin, whereas the remaining portion was covered with normal full-thickness skin. No signs of CSF leakage were noted. The lesion had a wide base and typical “hair collar” sign, which was characterized by a ring of coarse hair surrounding the mass consistent with a dysraphism (Fig. 1A). No neurological deficits were found.

Cervical 3D CT revealed a fusion defect at the level

This article contains some figures that are displayed in color online but in black-and-white in the print edition.
of the C-3 lamina (Fig. 1B). Preoperative MR imaging showed a cutaneous solid mass connected to intraspinal contents by a stalk traversing the lamina defect. The stalk and the deep portion of the mass were hypointense to neural tissue on both T1- and T2-weighted images, while the superficial aspect of the mass demonstrated a heterogeneous intensity on T2-weighted MR images. The dorsal aspect of the cervical spinal cord was tented posteriorly at the level flush with the inner end of the stalk. The enlargement of the central canal of the corresponding spinal cord indicated a local backward traction force. Magnetic resonance imaging also detected a small cystic lesion, which was hypointense on T1 sequences and hyperintense on T2 sequences, located beneath the tented neural band (Fig. 1C and D). No contrast enhancement was detected and all other radiological imaging, including cranial and thoracolumbosacral MR scans, was normal.

**Operation and Postoperative Course.** The cervical dysraphic lesion was excised under general anesthesia with the patient in the prone position. An elliptical skin incision was made around the base of the cutaneous mass, and a solid dural stalk extending from the core of the mass was isolated subcutaneously on opening the ligamentum nuchae. The stalk passed through the C-3 laminar defect with its dura-like integument, which was continuous with the normal intraspinal dura mater. A 2-level laminectomy centered on the C-3 defect was performed to expose the underlying dorsal dural diverticulum (Fig. 2 left). An intradural exploration was performed after the dural stalk was resected. The dorsal surface of the spinal cord was tented posteriorly via a slender neural band to connect the center of the dural stalk. In addition, a small CSF-filled cyst surrounded by a thickening arachnoid membrane was visualized just underneath the neural band. The cyst was resected by fenestration of its arachnoid wall without difficulty, and the tethering neural band was released from the vertical axis of the spinal cord without damage to normal tissues. The redundant dural sac, with the end of the dural stalk, was also removed, and the dural defect was repaired with flaps of paraspinal fascia. The patient made an uneventful recovery and was discharged to home on the 5th postoperative day. Cervical MR imaging 6 months after surgery confirmed that the dysraphic lesion including the arachnoid cyst was totally removed and the cervical cord had no further tethering. The patient exhibited no signs of neurological deficits at the 12-month follow-up.

**Pathological Findings.** All resected tissue was examined in conventional and immunoperoxidase sections. In the cutaneous mass, there was a loose network of pseudovascular spaces with distorted hair follicles in the superficial portion compared with the deeper portion, which presented with dense collagenous tissue containing clusters of meningocytes. The meningocytes were identified by their round to oval nucleus, inconspicuous nucleolus, eosinophilic cytoplasm, and an indistinct cell border. These meningothelial cells had a marked tendency to form a whorled pattern, and some whorls had already degenerated into psammoma bodies (Fig. 2 right). The dural band, covered by a layer of normal dura mater, had fibroconnective tissue and scant blood vessels in its center. There were no neural elements in the extraspinal part of the dysraphic lesion. On immunohistochemical evaluation, the cutaneous mass strongly expressed vimentin and epithelial membrane antigen but showed no immunoreactivity to CD31, CD34, S100, and glial fibrillary

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**Fig. 1.** Appearance of the large posterior cutaneous mass (A), which caused the cosmetic problem. Note the purplish, poor-quality skin on its top and typical “hair collar” on the bottom. Cervical 3D CT scan (B) demonstrates a bony defect at the level of the C-3 lamina. Sagittal T1-weighted (C) and T2-weighted (D) MR images show a cutaneous solid mass connected to the intraspinal contents by a stalk traversing the lamina defect, in addition to a small CSF-filled cyst located beneath the tented neural band.

**Fig. 2.** Intraoperative photograph (left) showing the stalk passed through the lamina defect with its dura-like integument, which was continuous with the normal intraspinal dura mater. Photomicrograph (right) demonstrating dense collagenous tissue containing clusters of meningocytes and a psammoma body (*black arrow*) in the cutaneous mass. H & E, original magnification ×200.
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acidic protein. Histopathological examination confirmed the arachnoid features of the wall of the intradural cystic lesion. In view of the fact that the tethering band from the dorsal aspect of the spinal cord was mainly composed of dysplastic glia and neurons, the band was classified as neuroglial.

Discussion

Several systems are used in naming and classifying cervical spinal dysraphism. According to a multicenter research, CDCT can be divided, based on the content of cutaneous protuberance, into 3 subgroups: neuroglial or fibrovascular stalk, myelocystocele, and true meningocele. In the classification by Rossi et al.,

11 cervical spinal dysraphism, as a kind of nonterminal myelocystocele, has 2 subtypes: abortive and complete myelocystocele. From their description, it is clear that the counterpart of the abortive myelocystocele is the "stalk" subgroup in the CDCT classification, while the complete myelocystocele corresponds to "myelocystocele." Because many cervical dysraphic lesions have nerve tissue in their walls, they were also called cervical myelomeningocele by some authors.

Almost all of these lesions reported in the literature had a CSF-filled cyst in the midline cutaneous mass. However, there is another kind of cervical dysraphism, just as that in the present case, that can be classified as a rudimentary meningocele, a rare congenital spinal malformation with a solid cutaneous mass mostly occupied by meningothelial elements. Most reported rudimentary lesions were on the midline scalp, preferentially involving the occiput and vertex. In general, there is only a meningothelial scalp mass, which is not accompanied by an underlying bony defect or any connection to the meninges. For this reason, a rudimentary meningocele has been referred to as a cutaneous meningiomata, because an opinion exists that this entity develops by proliferation of meningeal cells along the routes of cutaneous nerves, but not as a form of meningocele in which the underlying connection to the meninges is obliterated.

However, new evidence emerged to prove the viewpoint that rudimentary meningocele is a remnant of a neural tube defect. In a study from University Graz, common locations of dysraphism were seen to coincide with the anatomical distributions observed in cases of rudimentary meningocele. In addition, familial occurrence, as one feature of neural tube defect, was also noted in 2 siblings with rudimentary meningocele and in an autosomal dominant inherited pattern. The discovery of the connection to meninges through a bony defect may provide more strong evidence for the genuine pathogenesis of this kind of lesion. There are only 3 cases of rudimentary meningocele with such a connection reported in the literature—1 in the cervical and 2 in the lumbar region—but all of them were in infants and represented only inconspicuous fibrous tracts plus a small osseous defect or none at all. As such, our case may shed light on the pathogenesis of this lesion, with the fibrous stalk and the lamina defect becoming apparent with the advance of age.

A recent study indicated that the average rate of abnormal neurological status in patients with a CDCT was 15%. The most common symptoms and signs were cervical pain, motor weakness of extremities, hyporeflexes, CSF leakage, and urinary incontinence. Associated anomalies in CDCT are common. The average rates of Chiari malformation Type II and hydrocephaly are 40.1% and 40.8%, respectively. The main purpose of surgery in cervical dysraphism is to prevent neurological deterioration and infection due to CSF leakage. Some series have emphasized that the resection of a cutaneous lesion with intradural un tethering is essential in symptomatic patients, because clinical outcome is almost always unsatisfactory otherwise during follow-up. Stretch-associated injury in the tethered spinal cord is considered to be the main cause of subsequent myelopathy. However, in their largest adult series of untreated CDCTs (n = 5 patients), Duz et al. demonstrated that 2 patients had normal neurological status, 1 experienced intermittent CSF leakage, and 2 had only mild neurological deficits.

Surgery is beneficial in children and adults with symptomatic cervical dysraphism. When such lesions are present in asymptomatic adults, however, the indications for surgery are controversial, particularly if the lesion is a rudimentary meningocele. Tethering impairment is particularly pronounced when a lumbar or sacral lesion causes a craniocaudal traction, whereas lesions in the cervical region are less apt to cause this type of traction. There was indeed some tethering force from the stalk, but that force may be actually closer to that offered by normal cervical nerve roots. Moreover, the solid cutaneous lesion prevented any CSF leakage. The present case raises an issue about the natural history of cervical rudimentary meningocele, which may be benign and needs to be considered before any discussion of treatment options. After providing complete informed consent, it is reasonable for asymptomatic adults to undergo follow-up and observation. Surgery is reserved for adults with symptoms or a strong wish to have the cutaneous mass removed. Once surgery is chosen, all possible tethering elements including fibrovascular stalk, thickened arachnoid membrane, and other aspects should be carefully assessed and addressed. It is important to keep in mind that detethering procedures are not risk free, and the risks must be weighed against the risk of deterioration if the lesion is not immediately operated on.

Conclusions

The present case demonstrates that rudimentary meningocele is remnant of a neural tube defect, and this subgroup should be included in the existing classification of cervical spinal dysraphism. Our experience, limited as it is, indicates that this entity may have a relatively benign natural history. The decision to perform resection should be well considered, and it is reasonable for asymptomatic adults to be followed up and observed after providing complete informed consent.

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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