Dermoid tumors occurring at the site of previous myelomeningocele repair

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In 1984 and 1985, four children were treated at our institution for dermoid tumors in the area of a previous myelomeningocele repair. Because of the paucity of reports on this condition, the clinical presentations, radiographic studies, and operative findings in these patients are reviewed.

Case Reports

Case 1

This 9-year-old girl had been followed continuously in the myelodysplasia clinic since birth, when repair of a myelomeningocele was carried out. At 3 weeks of age she underwent ventriculoperitoneal (VP) shunting for hydrocephalus and she has had four subsequent shunt revisions. She had an L2–3 sensory level bilaterally and was able to ambulate with bracing and crutches; urinary continence was maintained on an intermittent catheterization regimen. She had an L2–3 sensory level bilaterally and was able to ambulate with bracing and crutches; urinary continence was maintained on an intermittent catheterization regimen. At approximately 8 years of age, she exhibited a gradual deterioration in both leg strength and urological status. By the age of 9½ years, she was totally incontinent despite a variety of pharmacological regimens, and she was unable to climb stairs or rise from a seated position. The myelomeningocele incision became sensitive to touch, and she complained of pain radiating down the anteromedial aspect of the left leg.

Examination. The computerized tomography (CT) scans and VP shunt studies revealed that the ventricles were unchanged from previous scans and that the shunt was functioning normally. Myelography, performed in order to rule out the presence of cord tethering, demonstrated a filling defect at L4–5 in the area of the previous myelomeningocele repair (Fig. 1 left). A CT scan following the myelogram demonstrated an atrophic dorsally displaced cord with a large, bilobed, low-density, pedunculated mass lying anterior to it (Fig. 1 right).

Operation. At surgery, the nerve roots both above and below the area of the old myelomeningocele placode were taut, and the cord was tethered dorsally by scar tissue both at and above the area of previous repair (Fig. 2 upper left). When these adhesions were released, a large, smooth, glistening mass could be seen anterior to the placode (Fig. 2 upper right), which on incision was found to represent a typical dermoid tumor containing sebaceous material, cholesterol debris, and hair (Fig. 2 lower left). The tumor could not be completely removed because of dense adherence of the capsule to the undersurface of the placode and functioning nerve roots (Fig. 2 lower right).

Postoperative Course. At follow-up evaluation 6 months postoperatively, the patient’s leg strength had returned to previous capacity, but she was still incontinent of urine. Spinal fusion was carried out for progressive scoliosis. At 1½ years after surgery, she was once again continent on an intermittent catheterization regimen supplemented by Ditropan (oxybutynin chloride). She has no back tenderness or leg pain.
Case 2

This 13-year-old girl had been continuously followed in the myelodysplasia clinic following repair at birth of a myelomeningocele. She underwent VP shunting at 2 months of age and had one subsequent revision. Yearly neurological examination revealed a stable L4–5 sensory level. At the age of 12 years, during a period of rapid growth, she developed progressive back and leg pain and weakness. Her posture became progressively more flexed, and she began to use a wheelchair in preference to walking. Concomitantly, urinary continence, which had been maintained on intermittent catheterization, was lost. She also noted progressive pain in the area of the myelomeningocele repair, which became exquisitely sensitive to touch.

Examination. A CT scan revealed no ventricular dilatation when compared with previous studies, and no further testing of shunt function was carried out. Unenhanced CT scanning of the lumbar region revealed a low-density, irregularly septated lesion filling the enlarged canal. Myelography disclosed a lobulated filling defect in the low lumbar area, and a CT scan following the instillation of metrizamide outlined a septated mass in the mid-lumbar area.

Operation. At surgery, the spinal cord above the placode was found to be atrophic and tightly adherent to the dorsal and lateral dura. Caudal to the open placode, which was similarly adherent to either side of the dura, was a soft irregular mass containing squamous debris and hair. The lesion was gutted and its superior capsule excised but, because of dense adherence to normal nerve roots, it could not be completely removed.

Postoperative Course. There was significant improvement from the patient's preoperative condition, and 6 weeks after surgery her back and leg pain had disappeared. She was continent on intermittent catheterization and was able to walk with braces. Four months later, an episode of shunt malfunction, heralded by progressive tenderness in the back surgery area, necessitated rehospitalization and shunt revision. Seven months postoperatively, she again complained of tenderness at the incision site and fullness in the lumbar area. Neurological examination revealed no deterioration, but she was readmitted and CT scanning of the head and lumbar and thoracic cord was carried out. The studies revealed no evidence of hydrocephalus, recurrent tumor, or hydromyelia. The scoliosis had measurably increased during this period, however, and spinal fusion was carried out. Urinary continence has been maintained with intermittent catheterization and a Ditropan regimen.

Case 3

This 8-year-old girl, who had previously undergone neurosurgical care at other institutions, was initially evaluated at the Boston Floating Hospital because of progressive scoliosis and lower-extremity spasticity. At birth, a malformation, termed a "low myelomeningocele," was repaired. A VP shunt was placed for hydrocephalus, but at 18 months of age an episode of shunt obstruction resulted in quadripareisis. A cervical laminectomy was carried out but only upper-extremity function was recovered. At 3 years of age, she experienced progressive arm weakness and underwent a repeat cervicomedullary exploration with drainage of a small hydromyelia. She has also had several subsequent operations for slit-ventricle syndrome. In addition to progressive scoliosis and spasticity, bladder function had also deteriorated, with diminished bladder capacity and worsening of continence between catheterizations. Myelography performed several years previously had demonstrated a "tethered cord," although the exact configuration of the caudal spinal cord was not described.

Examination. Prior to the present admission, an unenhanced CT scan revealed a pocket of diminished density within an area of increased density in the lumbar sacral region, consistent with hydromyelia or a low-density mass in the conus. Because of the progressive neurological symptoms and scoliosis, a combined lumbo-sacral exploration and fusion was planned.

Operation. At exploration, the spinal cord appeared atrophic at its caudal end and a large distal cystic mass containing sebaceous and squamous debris and hair was found. The lesion was situated quite dorsally with no obvious anterior components. Because of the patient's preexisting paraplegia and spasticity, the lesion was excised en bloc along with adjacent neural tissue. Spinal fusion was then carried out by the orthopedic surgeons.

Postoperative Course. The patient's spasticity improved considerably at once after surgery, but 6 months later an impressive amount of proximal leg spasticity had returned. Ten months postoperatively, she was admitted for reevaluation. Studies demonstrated excellent shunt function and no ventricular dilatation. Total myelography and delayed spinal CT scanning failed to demonstrate a recurrent mass or hydromyelia. One year postoperatively, the patient underwent mid- and upper-lumbar rhizotomy to relieve the proximal leg spasticity, with good results maintained at the 6-month follow-up review. Urinary continence has likewise fluctuated, with improvement immediately after excision of the dermoid cyst lasting for 6 weeks, then rapid deterioration to her preoperative state, followed by resumption of continence on intermittent catheterization after rhizotomy.

Case 4

This 15-year-old boy underwent repair of a myelomeningocele as a newborn, followed by ventricular shunting. The shunt was subsequently revised twice, and he did well with an L4–5 neurological sensory level until the age of 14 years, when he began to develop difficulty maintaining urinary continence. He had pre-
Dermoid tumors at myelomeningocele repair site

**FIG. 1.** Case 1. *Left:* Metrizamide myelogram, anteroposterior view, showing a filling defect at the L4–5 level (arrowhead). *Right:* Metrizamide-enhanced spinal computerized tomography scan demonstrating a low-density mass (vertical arrows) projecting anterior and lateral to the atrophic cord and placode (horizontal arrow).

**FIG. 2.** Case 1. *Upper Left:* Initial surgical exposure (patient’s head to the right) demonstrating adherence of the cord and placode to the lateral dura (arrowheads). The area of the placode appears full and rounded. *Upper Right:* After release of the adhesions, the placode is retracted to the right, revealing a smooth white mass. *Lower Left:* The tumor capsule is opened, revealing squamous debris and hair. *Lower Right:* The appearance of the nerve roots, placode, and spinal cord following radical but subtotal removal of the mass.
Previously been continent on an intermittent catheterization regimen, but now urine began to dribble between catheterizations and his parents reported that his gait was deteriorating. Urodynamic studies confirmed the deterioration of bladder function with progressive bladder spasticity.

**Examination.** The patient denied any pain in the area of his myelomeningocele repair. The L4–5 sensory level appeared unchanged, but the repair itself was full and bulging, and a subcutaneous mass could be felt extending into the left paraspinous tissues. Studies revealed that the VP shunt system was functioning well. Myelography was difficult to interpret because of the patient’s scoliosis and because of multiple loculations of dye noted in the lumbar area, but postmyelography CT scanning demonstrated a large subcutaneous area of soft-tissue density adjacent to what was interpreted as either a thick filum terminale or a tethered atrophic cord. Magnetic resonance imaging confirmed the presence of a soft-tissue structure extending to the surface of the skin, but its exact relationship to neural structures was difficult to ascertain because of the patient’s scoliosis.

**Operation.** At surgery, a large dermoid cyst extending into the left flank and containing hair and sebaceous debris was totally excised from the subcutaneous tissue and removed from the distal tethered placode to which it was densely adherent. Reddish tissue lying ventrally at the base of the cyst was found on pathological examination to represent respiratory epithelium in an otherwise typical dermoid cyst.

**Postoperative Course.** Since his surgery, the patient’s gait has returned to the baseline condition. A suprapubic tube was inserted to treat a persisting severe urinary tract infection. At 1-year follow-up review no improvement of the preoperative urinary dysfunction was noted.

**Discussion**

Dermoid tumors are congenital lesions which most commonly arise from embryonic rests.1 They are most frequently located in the lumbosacral spinal canal, and in children may be associated with dermal sinus tracts and recurring meningitis.2,3 Although dermoid tumors, along with other congenital inclusions such as epidermoid tumors, lipomas, and ectopic renal and intestinal tissue, have also been associated with myelomeningocele,1,4 these lesions are usually reported as curiosities noted at initial repair.1 In our patients with previous myelomeningocele repair, the dermoid tumors became symptomatic toward the end of the first decade or in the early part of the second decade of life and caused deteriorating lower-extremity and bladder function.

These symptoms are indistinguishable from those related to spinal cord tethering from any cause, and of themselves are not typical of dermoid tumors alone. For example, in Reigel’s series5 of patients with tethered spinal cords, most of which were noted subsequent to repair of a myelomeningocele or myelomeningocele, the most common presenting complaints were also changes in gait, progressive leg weakness, or pain. Reigel also noted in several patients the development of progressive postural changes with flexion of the knees and increasing lumbar lordosis, a finding that we observed in one of our patients as well. Dermoid tumors following myelomeningocele repair do appear to be relatively rare, however, and there are very few reports in the literature of these tumors in the myelodysplastic population. In Reigel’s series of 102 patients, 13 had epidermoid tumors and two had dermoid tumors in the area of a myelomeningocele repair. In an article on tethered spinal cord following myelomeningocele repair, Venes and Stevens4 mentioned that dermoid tumors are “not rare” in this situation, but none of their eight patients had dermoid or epidermoid tumors. Heinz, et al.,5 in their series of 16 patients with tethered cord after myelomeningocele repair, reported that five patients had “neoplasms.” Four of these lesions were lipomas and one was a neurenteric cyst; none was a dermoid tumor.

The etiology of these benign tumors in children with myelomeningoceles is probably mixed. Certain of these lesions, particularly those that are found dorsally and relatively superficially at exploration, must be related to incomplete incision of dermal elements at the time of the original repair. Two of the children in our series were initially operated on by the same neurosurgeon, but review of the operative notes failed to disclose any variation from ordinary findings or operative technique that might predispose to this late result. Lesions found ventral to the placode must be related to congenital dermal inclusions. Venes and Stevens4 refer to the work of Bouton, et al., and Lichtenstein, who postulated that teratomas and dermoid cysts occur following invagination of mesodermal and cutaneous elements as the neural folds fuse.

The diagnosis in these children was readily made by myelography and/or metrizamide-enhanced CT scanning of the spine. Myelography revealed an irregular, lobulated, filling defect adjacent to the area of previous repair, and CT scanning demonstrated low-density lesions; septations were noted in one patient. In those patients in whom the spinal cord was adequately visualized, it was atrophic and displaced dorsally and laterally in the canal. Magnetic resonance imaging was helpful but not diagnostic in one of our patients; as this technique becomes refined, it will probably simplify the diagnosis and treatment of these children in the future. We wish to emphasize that, as part of the radiographic work-up of these patients, it is imperative that the adequacy of ventricular shunting be confirmed by cranial CT scanning and dynamic shunt function studies if indicated, since ventricular shunt malfunction can at times mimic the neurological deterioration seen with cord tethering.

The long-term prognosis remains guarded in this group of patients. Only one of our four children continues with an excellent result over a 1-year follow-up
Dermoid tumors at myelomeningocele repair site

period. Two others have required additional procedures and repeated hospitalizations in order to maintain neurological stability or improvement. Because these tumors contain sebaceous and squamous debris, surgery may be followed by additional scarring and re-tethering; it was necessary to monitor all of these children very closely while their neurological course fluctuated in the months after surgery.

References

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