Congenital Intramedullary Lipoma of the Dorsocervical Spinal Cord with Intracranial Extension: Case Report

Naim-Ur-Rahman, F.R.C.S., F.R.C.S.Ed., F.A.C.S.,
Mustafa A.M. Salih, M.P.C.H., M.D.,
Abdul Hakim Jamjoom, F.R.C.S. Ed.,
Zain Alabeledin Jamjoom, M.D.

Departments of Neurosurgery (N-U-R, AHJ, ZAJ) and Pediatrics (MAMS), College of Medicine, King Saud University, Riyadh, Saudi Arabia

THE UNUSUAL COMPUTED tomographic findings of an extensive intramedullary lipoma of the dorsocervical spinal cord, which extended into the brain stem, in a 9-month-old infant are described and correlated with the surgical findings. The operative therapeutic options are discussed. (Neurosurgery 34: 1081–1084, 1994)

Key words: Computed tomography, Congenital intramedullary lipomas, Floppy infant syndrome, Hamartoma, Intracranial extension, Lipoma, Spinal cord tumors

Extensive intramedullary lipomas of the cervicothoracic spinal cord, which extended into the brain stem, were not associated with spinal dysraphism, and presented in the infancy/neonatal period, are extremely rare and poorly documented. Most of the cases reported in the literature (1, 2, 5, 6, 13), were confined to four or five segments of the cord (3), presented in adult life (1, 3, 5), and extended rarely into the posterior fossa and brain stem (16). Most of the reported intradural lipomas of the cord were subpial (3, 7, 9) and not truly intramedullary. Thus, in McLone’s (9) series of 130 spinal lipomas, 5 were totally intradural. Of these five, only one was completely surrounded by spinal cord tissue. A few reports of cervical cord lipomas with an intracranial extension have appeared (11, 12, 16), but none presented in the neonatal period or was as extensive as the present case.

CASE REPORT

This 9-month-old infant girl, who had a normal full-term delivery and an unremarkable prenatal history, was admitted to the neurosurgical ward with dense tetraparesis and respiratory difficulty. Before admission, the infant was treated elsewhere for hypotonia and lack of limb movement observed soon after birth. A diagnosis of floppy infant syndrome was made, but the cause remained obscure for many months. A physical examination revealed a tetraplegic child with flailing areflexic upper limbs and spastic lower limbs, marked ankle clonus, and bilateral upgoing plantar response was noticed. Her bladder and bowel control was impaired. Her sensation was impaired up to the mandible/upper cervical region. Her respiration was shallow, quick, and respiratory movement was confined mostly to lower chest. Plain x-rays of the spine revealed a marked increase of the interpedicular distance and of the anteroposterior diameter of the cervicothoracic spinal canal (Fig. 1). Cranial and spinal computed tomographic (CT) scans (Fig. 2) disclosed an enor-
and posterior fossa was made, and surgery was performed to confirm the histological diagnosis of the mass and to decompress the neural structures in the posterior fossa and spinal canal. The patient was placed in a prone position. A vertical midline incision from above the inion to below the T5 spinous process was made. A posterior fossa craniectomy with a cervical and upper dorsal laminectomy was performed. The cervicothoracic dural tube was extremely tense and nonpulsatile. Incision of the posterior fossa and spinal dura down to the upper thoracic spine revealed a large, tense, and expanded cervicothoracic spinal cord and a bulging floor of the 4th ventricle with upward displacement of the cerebellar tonsils (Fig. 4). An extensive posterior midline myelotomy exposed the posterior surface of the entire intramedullary lipoma with dense fibrous septa and poor cleavage between the neural and fibrofatty tissue. A Cavitron ultrasonic aspirator was used to debulk the centrum of the tumor. Adequate decompression was achieved, leaving a relaxed pulsating cord and posterior fossa (Fig. 5). The histological examination revealed a lipoma. The patient's postoperative course was uneventful. Although the mobility of the patient's upper extremities has improved, the improvement of motor activity in her legs is less pronounced.

**DISCUSSION**

To determine the nature of pathological changes responsible for tetraparesis and hypotonia in infancy is often a difficult matter (15). This case shows that congenital cervical cord lipomas can masquerade during the first few months after birth as the "floppy infant syndrome." This syndrome includes all infants who show severe, generalized weakness and hypotonia soon after birth (15). In the literature (14, 15), the causes of the floppy infant seldom include congenital spinal cord lesions like the one described here. An investigation of unexplained tetraparesis and hypotonia in infancy should include myelography, spinocranial CT scanning, or better still, magnetic resonance imaging.

CT scans can detect spinal intramedullary/intracranial lipomas as low-density mass lesions. Because of the low attenuation values of these lipomas (-115 Hounsfield units in our case), CT scan appearances are quite characteristic and diagnostic (Figs. 2 and 3). A noncontrast CT scan allows a definitive diagnosis of these lesions because of low fat density. Being quick and cost effective, CT scans can be used as a screening test whenever a cord lipoma is suspected. Magnetic resonance imaging, because of the high-signal intensity of fat on T1-weighted images, allows a more accurate anatomical delineation of the tumor and its exact relationship to the cord (4). Because such extensive intramedullary lipomas of the dorsocervical cord with brain stem extensions are extremely rare, no clear guidelines are available in the literature regarding their management. Surgical treatment in our case consisted of combined suboccipital craniectomy, spinal laminectomy, and subtotal tumor decompression. The Cavitron ultrasonic aspirator (Cooper Medical Corporation, Mountain View, CA) was found to be very helpful in debulking the centrum of this intramedullary tumor. A lack of cleavage plane and the intermingling of neural and fibrofatty tissue at the periphery of the tumor made total tumor removal impossible. The reported operative options include a simple posterior fossa decompression combined with cervical laminectomy or a laminotomy with flap elevation (12) along with a biopsy (7), and partial (1, 4, 11, 16) or subtotal removal (3, 12). In very extensive lesions involving most of the spinal cord and brain stem, a two-stage procedure may be a reasonable option. Total excision of these lesions is seldom possible because of the lack of a clear cleavage plane between the neural and fatty tissue (3, 7, 11). Severe cord damage resulting in death (7) or tetraplegia (1) usually follows attempts at the complete removal of these lesions (1, 7). Thus, the main purpose of surgery for lipoma is not total removal but decompression of the adjacent neural structures (11). The usefulness of a Cavitron in debulking these lesions is well established (11), and the laser, with its selective fat melting/ evaporating properties, has proved to be another effective tool for this purpose (10).

The follow-up period in our case has been very modest, but judging from early postoperative improvement in the neurological status and reports of good
outcome in patients whose preoperative neurological deficit was slight (8), a good long-term prognosis can be anticipated.

Several hypotheses regarding the genesis of spinal cord lipomas have been proposed (1,8). Many characteristics of the present case and its presence at birth suggest a hamartomatous origin. Maldevelopment with the resultant inclusions of embryonic rests of fat cells during the formation of the neural tube is regarded as the most probable explanation of the origin of these lipomas (1,8).

To conclude, spinal cord lipoma should be considered in the differential diagnosis whenever a neonate presents with hypotonia, tetraparesis, or floppy infant syndrome and can be ruled out by craniocervical CT/magnetic resonance imaging scans.

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Reprint requests: Dr Naim-Ur-Rahman, Professor of Neurosurgery, Department of Surgery (57), College of Medicine, King Saud University, P.O. Box 2925, Riyadh 11461, Saudi Arabia.

REFERENCES

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COMMENTS

Spinal lipomas make up 1% or less of primary spinal cord tumors, with the majority being subpial at the conus and extending into the cauda equina, actually even extending extradurally into the soft tissues of the back. Only a small minority of lipomas are found that are truly intramedullary, and this very extensive, totally intramedullary tumor in an infant is a unique case. It would be helpful to have a long-term clinical follow-up as well as follow-up magnetic resonance imaging available on this case.

Edward S. Connolly
New Orleans, Louisiana

This article reports the extensive enlargement of the spinal canal in a 9-month-old, with long-standing massive enlargement of the underlying spinal cord. Both computed tomography and magnetic resonance imaging can certainly help us early to better understand the origin of the intraspinal lesion that would cause such dilatation of the spinal canal. In this case, the described findings certainly are consistent with lipoma, and although surgery cannot accomplish a cure in such a problem, I do agree with the authors that, even though the yield of finding a spinal cord tumor such as this is extremely rare, one has to consider magnetic resonance imaging in the evaluation of the hypotonic infant.

I think it is also very important that the spinal stability of these patients be carefully monitored in the postoperative period. Infants and children who have undergone extensive laminotomies, especially for relatively benign disease, need to be followed very closely for signs or radiographic evidence of progressive spinal deformity. Sometimes it, in fact, is appropriate to consider spine stabilization and fusion even at the time of the initial surgery. In other cases, a laminotomy with replacement of the removed lamina after tumor removal should be considered. Even if some deformity develops or progresses after such surgery, the now replaced lamina can provide us with a number of additional corrective surgical options that would not exist if the lamina had not been replaced. The need to carefully follow such patients after extensive laminectomies and, for that matter, any infant or child with a spinal-induced neurological deficit cannot be overemphasized.

Michael J. Ebersold
Rochester, Minnesota