Upper Cervical Intermedullary Lipoma with Extension into the Cistern Magna in an Elderly: Report of a Case and Review of the Literature

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ABSTRACT
Intermedullary lipoma not associated with dysraphism is very rare and those locating at the cervicomedullary region are extremely rare. Majority of lipomas of this area are manifested and diagnosed in childhood or early adulthood. An intermedullary cervicomedullary lipoma becoming symptomatic and diagnosed in old age is a medical curiosity. Here in a 68 years old male with progressive quadripareseis secondary to a high cervical subpial intermedullary lipoma passing through the foramen magnum is presented

KEY WORDS: Cervical spine, Craniocervical instrumentation, Geriatric, Intermedullary lipoma, Magnetic resonance imaging, Spinal cord

INTRODUCTION
Isolated intermedullary lipomas without neural tube defect are rare. These lipomas usually extend from 4 to 5 vertebral segments (6, 10, 12). Intermedullary lipomas can occur anywhere along the length of the spinal cord, but the most common location of these tumors is in the cervicothoracic followed by lower thoracic region (1, 2,6,7,9, 12, 17). In this report we present a case of intramedullary lipoma located at upper cervical spinal cord.

CASE REPORT
A 68–year old man was admitted because of severe neck pain for six months and inability to walk for two months. In retrospect the patient noted some clumsiness in gait since 10 years ago. This symptom had a very slow course and was aggravated with time. On admission we found an old man who was hardly able to walk a few steps supported by two persons. He also complained of dysesthetic pain of the extremities and incontinence. Neurological exam revealed spastic quadripareseis with impaired position sense in the lower limbs. Cervical MRI was the first diagnostic tool used which showed a very enlarged upper cervical canal occupying with a hyperintense intermedullary mass extending from C4 up to cisterna magna both in T1- and T2-weighted images (Figure 1A,B). Cervical X-Ray revealed an exaggerated cervical lordotic curve. Scalloping of the upper cervical vertebral bodies was also noted (Figure 2). CT scan disclosed an enormously enlarged upper spinal canal and foramen magnum filled by a large, low–density mass surrounded by several patches of calcification. Canal widening with erosion and remodeling of pedicles were demonstrated (Figure 3A,B). Diagnosis of a cervicomedullary lipoma was made.

At surgery, limited suboccipital craniectomy and upper cervical laminectomy above C5 was made after dural opening a yellow subpial tumor extending from C4 up to 2 cm above the foramen magnum which had occupied a part of cistern magna was observed. Through an extensive midline myelotomy, intratumoral resection of the lipoma was performed carefully with the aid of a sonographic aspirator avoiding the rootles which were embedded inside the tumor. After subtotal resection, duraplasty with
A patch of fascia lata was used for enlarging the dural tube and in order to allow free pulsation of the cord. With regard to the tiny bony element that remained after laminectomy and the intractable neck pain, craniocervical instrumentation with application of screw rod-plate was done (Figure 4). The postoperative course was uneventful. Plain radiographs showed the proper placement of screws (Figure 5). The patient was discharged after 10 days while his neurological status remained unchanged. Surprisingly, at follow up examination 3 months after surgery, he was able to walk with only the support of a cane.

**DISCUSSION**

It is well documented that less than 1% of all intraspinal tumors are spinal cord lipomas (7, 10, 12). These lipomas are true neoplasms because there is progressive increase in tumor size over time (2). Isolated intermedullary lipomas without neural tube defect are rare. These lipomas usually extend from 4 to 5 vertebral segments (6, 10, 12). They may occupy more spinal cord levels and even appear as a holocords tumor on rare occasions (7,9,16,17). Intermedullary lipomas can occur anywhere along the length of the spinal cord, but the most common location of these tumors is in the cervicothoracic followed by lower thoracic region (1, 2,6,7,9, 12, 17). Nonetheless, its
occurrence in the cervicomedullary area is supposed to be very rare. In a review of the literature, we were able to find only 20 cases of cervicomedullary lipomas reported in the past (1-3, 6, 7-11, 13-16, 19-21). This search disclosed that the ages of the patients with cervicomedullary lipoma range from 6 months to 62 years which means that our patient diagnosed at the age of 68 is apparently the oldest one in the literature. The review also revealed that the sex distribution are almost equal in males and females (20). The survey disclosed that the cervicomedullary intermedullary lipomas can be classified in two different groups according to the age and the clinical picture of the patients. One group is confined to infancy and early childhood, usually being manifested as a floppy child secondary to general hypotonia (8, 9, 15, 16, 21). The patients of the second group are adolescents and young adults being ultimately admitted with quadriplegia (10, 13, 14, 21). The clinical course in this group is indolent and it usually takes more than 5 years to reach to the correct diagnosis (12). This rather indolent period might
occasionally be up to 10 years. The clinical picture in this group is extremely subtle and usually starts with mild quadripareisis that can remain static for years (1,12,13). Sensory disturbances in the form of dyesthesias, and affection of the posterior column manifested as a mild clumsiness are the major clinical picture in this period.

However, once the cord compromise secondary to further growth of the tumor cannot be tolerated any more, the pain is escalated and the patient will develop rapid deterioration of neurological signs obliging him/her to seek medical attention which ultimately leads to the correct diagnosis. In plain radiographs, widened canal, scalloping of the posterior border of the corresponding vertebral bodies, and increased interpedicular distance are the cardinal radiographic feature of the condition and are demonstrated specially in longstanding cases (12, 15, 20). However, MRI is the diagnostic tool of choice in intermedullary lipomas. It allows a more accurate anatomical delineation of the tumor and its exact relationship to the cord (4,5,7,16,17). As fat has a short relaxation time on T1-weighted sequences, it will be demonstrated as a hypersignal mass on T1 MR images (3,4,7,12,13,14,16,17). Relaxation of fat in T2–weighted images is variable and may appear as hyperintense, isointense or hypointense when compared to normal neural parenchyma (8,11,18,21). However, they are presented as a hypointense or isointense mass on T2 images in the majority of the reported cases (10,12,13).

CT specially reformatted appearances are quite characteristic and diagnostic as they show a low fat density (1011,14,15,16,19,21). In long-standing case, the lipoma might be circumscribed with linear calcification indicating its chronicity.

Once the diagnosis is made, surgical intervention is recommended. It should be noted that lipomas are adherent to neural parenchyma and there is no clear cleavage plane between the neural tissue and fatty tumor. Therefore, radical removal of these tumors is not recommended since attempts at complete excision carry an unacceptable risk of postoperative morbidity and even mortality (16,19). Surgical options range from partial excision to subtotal removal (13,18). Subtotal removal with duraplasty is the treatment of choice and generally provides an acceptable outcome (8,10,21). CO2 laser and sonographic aspiration are known to reduce intraoperative damage to the cord (1,15). Recognition of necessary extent of surgical excision can be aided by monitoring electrophysiological responses (7,12). Various degrees of recovery can be expected with appropriate surgery. Once surgery is done early in the course of disease, a favorable result may ensue (7,8,10,16,21). However, once the disabling symptoms are settled, significant residual disability may remain (7,11,12,20). However, catastrophic results with quadriplegia and death has been described after surgery (4,19). In decompressive surgery for such benign tumors in young children, bony fusion, reconstructive surgery or internal stabilization should be considered in order to prevent future deformity of the spinal column (7,9,14,19). Surgical stabilization is not usually necessary in adults. However, in patients with severe bony changes as well as in those with intractable neck pain supposed to be due to severe lordosis, laminectomy may result in instability or further deformity. Therefore, laminoplasty or internal fixation seems worthy in such instances.

A new case of cervicomedullary lipoma in an elderly is reported which is the first example of intermedullary lipoma in this age group. The CT features of this case were the most demonstrative up to date. Generous debulking, duraplasty and cranio-cervical instrumentation result in acceptable outcome.

REFERENCES


