

Co-existence of Lipoma and Myxopapillary Ependymoma in a Filum Terminale Tumor

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A 65-year-old woman presented with a history of severe lower back pain on forward-flexion for 2 months duration. Magnetic resonance imaging revealed a high signal mass with a tail on T1-weighted images at the L3 level. A total surgical resection was performed via a posterior approach with the aid of a microscope. Histopathological examination of the tumor revealed two pathological components: lipoma and myxopapillary ependymoma. The presence of dual histological components in one spinal cord tumor is rare. There are no prior reports of both types of cells (adipose and ependymal) grown simultaneously in a single tumor of the filum terminale in the medical literature. We report a unique case of the co-existence of lipoma and myxopapillary ependymoma within the same tumor located at the filum terminale and review related literature.

KEY WORDS : Co-existence · Lipoma · Myxopapillary ependymoma · Filum terminale.

Introduction

The incidence of primary central nervous system (CNS) neoplasm in adults has been reported to be from 11 to 12 per 100,000²¹⁾. Spinal cord tumors consist of a reported 15% of CNS neoplasm. In reported cases of intracranial tumors, the co-existence of dual pathology in one patient and the pathological co-existence in one tumor are rare events^{4,5,8,16,18,20,22)}. For spinal cord tumors, some cases of dual pathology in a single patient have been reported^{2,7,11,13,17)}.

However, there is no prior report of the pathological co-existence in a single tumor of the filum terminale. We report a unique case of the co-existence of lipoma and myxopapillary ependymoma within the same tumor of the filum terminale and review of the related literature.

Case Report

A 65-year-old woman presented with one-year history of low back pain after a slip and fall accident, which was aggravated by forward bending for 2-month prior to admission. On admission, her neurological examination was normal. Plain lumbosacral x-rays revealed an old compression fracture at the T12 vertebral body. The stress views revealed no instability.

Lumbar magnetic resonance imaging revealed an intradural extramedullary tumor with a tail at the level of L3 vertebral body. The tumor observed to be attached to the low-lying conus medullaris. The lesion showed high signal intensity on T1-weighted images, iso-intensity on T2-weighted images and no enhancement on gadolinium enhanced images (Fig. 1). The patient underwent a midline L3-4 laminectomy. After dural incision, a yellowish fatty tumor with a tail was exposed to be immediately dorsal to the filum terminale, it measured 35 × 6mm in size. The tumor and tail were completely removed as one fragment (Fig. 2). On microscopic examination (Fig. 3), the tumor showed well demarcated margin and mostly consisted of mature adipose tissue, which was consistent with lipoma. In addition, admixed tumor with adipose tissue showed a localized proliferation of cuboidal to elongated ependymal cells radially arranged in a papillary manner around vascularized stromal cores as well as in nodular manner with myxoid appearance. The tumor cells showed focal positive reaction to glial fibrillary acidic protein (GFAP) and S-100 protein antibodies. Mitotic activity was absent. The findings were consistent with myxopapillary ependymoma. The patient made an excellent recovery. The postoperative course was uneventful. The physical examination performed 3 months after surgery showed no neurological deficits.

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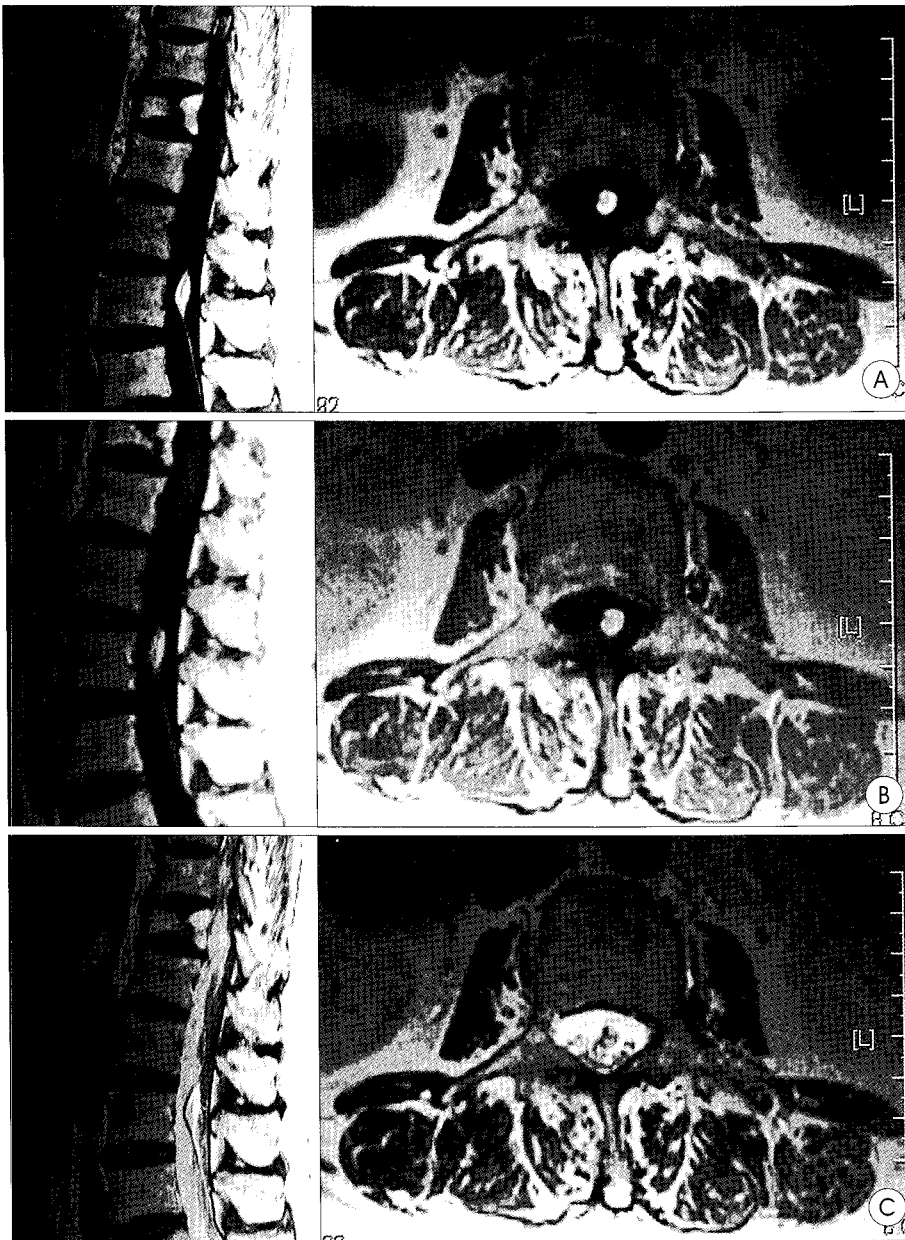


Fig. 1. Preoperative magnetic resonance images show a tumor at L3 level with a tail. The tumor is attaching to low-lying conus medullaris. T1-weighted image reveals high signal mass and tail (A). T1-weighted image after administration of gadolinium intensity visualizes no enhancement (B). T2-weighted image shows iso signal intensity (C).

Discussion

The subtypes of spinal cord tumors are anatomically classified by their relationship to the dura mater and spinal cord parenchyma. Intradural tumors can be intramedullary or extramedullary and account for roughly three fourths of all spinal tumors. About two thirds of tumors are extramedullary, well circumscribed and histologically benign. Meningiomas and nerve sheath neoplasms account for 80% of extramedullary spinal cord tumors and filum terminale ependymomas

make up 15% of these lesions. The remaining 5% of tumors include: paraganglioma, lipoma, drop metastases and granulomas, all of which are rare⁹⁾.

Lumbosacral lipoma is a congenital abnormality in which subcutaneous fat tissue extends into the spinal canal and attaches to the distal part of the spinal cord. It is the most common occult dysraphic lesion that leads to the tethered cord syndrome. Its incidence is reported to be 8~25% of myelomeningoceles or about 1 in 4,000 live births^{3,12,19)}.

Ependymomas are slow-growing tumors that derive from the cells that line the ventricular spaces of the central nervous system, including the central canal of the spinal cord. They are the most common low-grade neuroepithelial tumors of the spinal cord, accounting for 50 to 60% of spinal cord gliomas. Myxopapillary ependymomas represent the most frequent type of ependymomas; they are found at the conus medullaris-cauda equina-filum terminale level^{6,10)}.

In our case, the tumor was composed of both adipose and glial tissue. The adipose tissue represented the largest component. The findings were for a tumor and therefore we could exclude a hamartomatous origin. The microscopic examination and immunostaining suggest that both types of cells (adipose and ependymal) grew simultaneously in the same mass.

Lemire, et al.¹⁵⁾ and Alvord, et al.¹⁾ have proposed a mechanism of caudal neural tube formation that explains the persistence of mesenchymal remnants such as lipomas. Following neural tube formation or neurulation, the second phase of caudal neural tube formation occurs. A coalescence of pluripotential cells into a caudal cell mass stretches from the posterior neuropore into the sacrococcygeal level. The third phase of caudal neural tube formation had been termed "retrogressive differentiation". In this phase, selective death of some of the poorly differentiated cells of the caudal cell mass

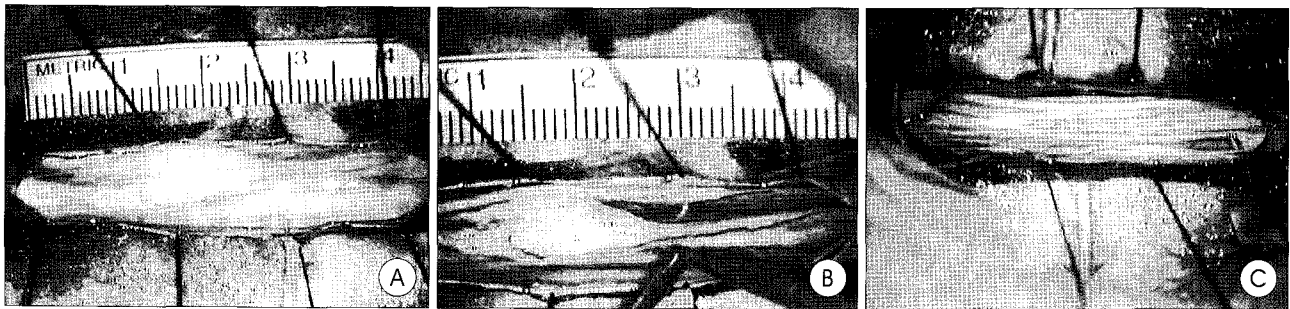


Fig. 2. Operative findings demonstrating a yellowish fatty tumor. Tail is revealed to be immediately dorsal to a filum terminale, which is measured 35×6mm in size (A&B) Complete removal of tumor and tail could be done by microsurgical technique without damaging nerve roots (C).

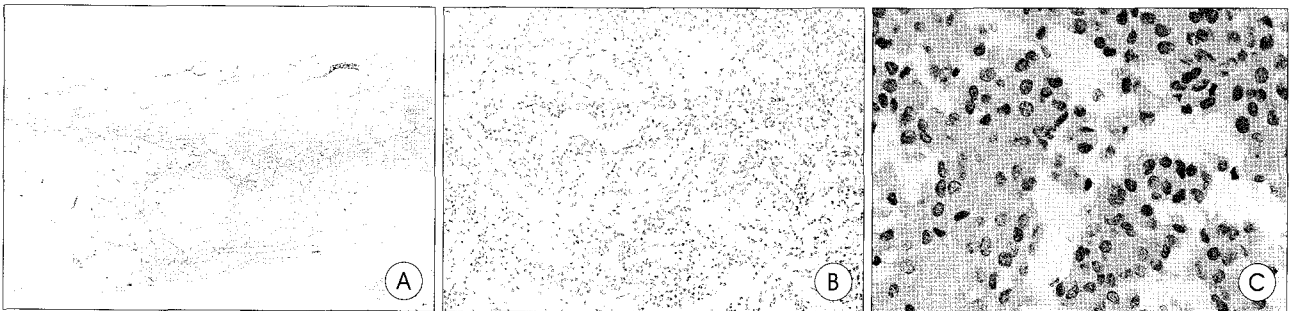


Fig. 3. Microscopic findings. A tumor shows well demarcated margin and mostly consisted of mature adipose tissue, which is consistent with lipoma (A). In addition, admixed tumor with adipose tissue shows a localized proliferation of cuboidal to elongated ependymal cells radially arranged in a papillary manner around vascularized stromal cores as well as in nodular manner with myxoid appearance. The findings are consistent with myxopapillary ependymoma (B&C). (H&E, original magnification, A×10, B×100, C×400).

occurs and the rest of the mass differentiates into the conus medullaris (with the ventriculus terminalis) and the filum terminale. The potential for these structures to play a role in the pathogenesis of tumors and malformations has been previously discussed¹⁴. It is possible that the lipoma results from a “clone of lipomatous cells” that escapes the programmed cell death during retrogressive differentiation. In addition, Walsh and Markesbery²³ have suggested that other tissue types found at this location such as neuroglia, ependyma, smooth and striated muscle, may be accounted for by the persistence of pluripotential embryonic cells that normally disappear during the completion of neural tube formation. These pluripotential cells may become neoplastic (eg. myxopapillary ependymoma) in some patients. We postulate that the proposed mechanism for the pluripotential remnant cells may be important to the formation of the dual pathologic components in the same tumor located at the filum terminale.

This is the first report of the co-existence of dual pathology in a single spinal cord tumor found at the filum terminale^{2,7,11,13,17}.

Conclusion

We report a unique case of the co-existence of lipoma and myxopapillary ependymoma within the same tumor of the filum terminale. The manifestation of tumor growth should be taken into consideration in the future.

References

1. Alvord EC Jr, Shaw CM, Sumi SM : Central nervous system developmental abnormalities in Thompson RA, Green JR(eds) : *Pediatric Neurology and Neurosurgery*. Orrville, OH : Spectrum Publications, 1978, pp97-166
2. Angiari P, Corradini L, Falasca A, Crisi G, Botticelli AR, Merli GA : Associated spinal meningioma and neurinoma. A case report. *Acta Neurol* 9 : 218-223, 1987
3. Bruce DA, Schut L : Spinal lipomas in infancy and childhood. *Childs Brain* 5 : 192-203, 1979
4. Bunick EM, Mills LC, Rose LI : Association of acromegaly and meningiomas. *JAMA* 15 : 1267-1268, 1978
5. Cannavo S, Curto L, Fazio R, Paterniti S, Blandino A, Marafioti T, et al : Coexistence of growth hormone-secreting pituitary adenoma and intracranial meningioma : a case report and review of the literature. *J Endocrinol Invest* 16 : 703-708, 1993
6. Cooper PR : Outcome after operative treatment of intramedullary spinal cord tumors in adults : intermediate and long-term results in 51 patients. *Neurosurgery* 25 : 855-859, 1989
7. Dorizzi A, Crivelli G, Marra A, Scamoni C, Dario A, Bonfanti N, et al : Associated cervical schwannoma and dorsal meningioma. Case report and review of the literature. *J Neurosurg Sci* 36 : 173-176, 1992
8. Elizabeth J, Menon G, Nair S, Radhakrishnan VV : Mixed tumour of schwannoma and meningioma in a patient with neurofibromatosis-2 : a case report. *Neurol India* 49 : 398-400, 2001
9. Guidetti B, Mercuri S, Vagnozzi R : Long-term results of the surgical treatment of 129 intramedullary spinal gliomas. *J Neurosurg* 54 : 323-330, 1981
10. Hanbali F, Fournay DR, Marmor E, Suki D, Rhines LD, Weinburg JS, et al : Spinal cord ependymoma : radical surgical resection and outcome. *Neurosurgery* 51 : 1162-1174, 2002
11. Honda E, Hayashi T, Goto S, Oshima Y, Kikuchi N, Utsunomiya H, et al : Two different spinal tumors(meningioma and schwannoma) with von Recklinghausen's disease in a case. *No Shinkei Geka* 18 : 463-468, 1990
12. Kanev PM, Lemire RJ, Loeser JD, Berger MS : Management and long-term follow-up review of children with lipomyelomeningocele, 1952-1987. *J Neurosurg* 73 : 48-52, 1990
13. Lee TH, Cho YJ, Cho YE, Jung HJ : Co-Existence of dual spinal cord tumor : Schwannoma and Meningioma. *J Korean Neurosurg Soc* 33 : 425-427, 2003

14. Lemire RJ, Beckwith JB : Pathogenesis of congenital tumors and malformations of the sacrococcygeal region. *Teratology* 25 : 201-213, 1982
15. Lemire RJ, Loeser JD, Leech RW, Alvord EC : **Normal and Abnormal Development of the Human Nervous System**. Hagerstown, MD : Harper and Row, 1975, pp421
16. Mathuriya SN, Vasishta RK, Dash RJ, Kak VK : Pituitary adenoma and parasagittal meningioma : an unusual association. *Neurol India* 48 : 72-74, 2000
17. Nishiura I, Koyama T, Tanaka K, Aii H, Amano S : The occurrence of different types of spinal tumours in one patient. A case report and review of the literature. *Neurochirurgia* 32 : 52-55, 1989
18. Santoro A, Minniti G, Paolini S, Passacantilli E, Missori P, Frati A, et al : Atypical tentorial meningioma 30 years after radiotherapy for a pituitary adenoma. *Neurol Sci* 22 : 463-467, 2002
19. Schut L, Bruce DA, Sutton LN : The management of the child with a lipomyelomeningocele. *Clin Neurosurg* 30 : 446-476, 1983
20. Sharma MC, Vaish S, Arora R, Gaikwad S, Sarkar C : Composite pituitary adenoma and intrasellar tuberculoma : report of a rare case. *Pathol Oncol Res* 7 : 74-76, 2001
21. Surawicz TS, Davis F, Freels S, Laws ER Jr, Menck HR : Brain tumor survival : results from the National Cancer Data Base *J Neurooncol* 40 : 151-160, 1998
22. Tripathi RP, Gupta A, Gupta S, Kumaran SS, Khushu S, Dev A, et al : Co-existence of dual intracranial pathology clinical relevance of proton MRS. *Neurol India* 48 : 365-369, 2000
23. Walsh JW, Markesbery WR : Histological features of congenital lipomas of the lower spinal canal. *J Neurosurg* 52 : 564-569, 1980