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 100,000(1,2). 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(2,3,4,5,6,7,8). For spinal cord tumors, some cases of dual pathology in a single patient have been reported(2,3,4,5).

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 appeared 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 physiological examination performed 3 months after surgery showed no neurological deficits.

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Discussion

The subtypes of spinal cord tumors are anatomically classified by their relationship to the dura mater and spinal cord parenchyma. Intramedullary 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 the these lesions. The remaining 5% of tumors include: paraganglioma, lipoma, drop metastases and granulomas, all of which are rare.

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.

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.

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. and Abord, et al. 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 pluripotent 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.
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 discussed. 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 Markesbery 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 pluripotent embryonic cells that normally disappear during the completion of neural tube formation. These pluripotent cells may become neoplastic in some patients. We postulate that the proposed mechanism for the pluripotent remnant cells may be important to the formation of the dural 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.

References


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.


