ELSEVIER

The Spine Journal 13 (2013) e49–e54

THE SPINE JOURNAL

Case Report

Tanycytic ependymoma of the filum terminale associated with multiple endocrine neoplasia type 1: first reported case

Toru Funayama, MD[a,*], Masataka Sakane, MD, PhD[a], Tomohiro Yoshizawa, MD[a], Yousuke Takeuchi, MD[b], Naoyuki Ochiai, MD, PhD[a]

[a]*Department of Orthopaedic Surgery, Graduate School of Comprehensive Human Sciences, University of Tsukuba, 1-1-1 Tennoudai, Tsukuba, Ibaraki 305-8575, Japan*

[b]*Department of Spine and Orthopaedic Surgery, Japanese Red Cross Medical Center, 4-1-22 Hiroo, Shibuya-ku, Tokyo 150-8935, Japan*

Received 21 March 2012; revised 25 February 2013; accepted 25 February 2013

Abstract

BACKGROUND CONTEXT: Ependymoma associated with multiple endocrine neoplasia type 1 (MEN-1) is an extremely rare clinical entity. To the best of our knowledge, only five cases of ependymoma associated with MEN-1 have been previously described. Furthermore, there has been no case of tanycytic ependymoma of the filum terminale associated with MEN-1.

PURPOSE: The present case report illustrates a 53-year-old man with tanycytic ependymoma of the filum terminale associated with MEN-1. We review the literature on ependymoma with MEN-1 and tanycytic ependymoma of the cauda equina region and also discuss the risk of recurrence.

STUDY DESIGN: A case report.

METHODS: The patient presented with complaints of nocturnal pain in the lower back, accompanied by numbness around the anus and intermittent claudication for approximately 1 year. Magnetic resonance imaging (MRI) identified an intradural-enhancing, large mass lesion at the level from Th12 to L2 vertebrae, with a cranial cystic lesion.

RESULTS: Open-door laminoplasty of the Th12, L1, and L2 and en bloc tumor resection with thickened filum terminale were performed. Histopathologic examination of the tumor specimens showed tanycytic ependymoma (World Health Organization Classification Grade II). At the time of the 2-year and 8-month follow-up examination, MRI did not show tumor recurrence.

CONCLUSIONS: This is the first reported case of this clinical entity. A careful follow-up of patients with this unusual tumor is strongly recommended.

Keywords: Tanycytic ependymoma; Filum terminale; Conus medullaris; Multiple endocrine neoplasia type 1

Introduction

Multiple endocrine neoplasia type 1 (MEN-1) is a rare autosomal syndrome, with an estimated prevalence of 1 to 17.5 per 100,000 inhabitants [1] or 1 per 30,000 individuals [2]. Multiple endocrine neoplasia Type 1 is characterized by neoplasia of the parathyroids, anterior pituitary, and endocrine pancreas and is correlated with mutations in the *MEN1* gene on chromosome 11q13 [3,4]. Other endocrine and nonendocrine tumors with lower frequency have been reported. These include adrenal cortical tumors; foregut neuroendocrine tumors of the gastrointestinal tract, thymus, and bronchi; facial angiofibromas; collagenomas; lipomas; and leiomyomas [1,2]. Although many kinds of tumor associated with MEN-1 can occur, central nervous system tumors, except intracranial meningiomas, have rarely been reported [5]. The occurrence of ependymoma associated with MEN-1 is very rare, and only a few such cases have been reported [6–10].

Tanycytic ependymoma is a rare subtype of ependymoma, which is formally recognized as a pathologic entity according to the latest World Health Organization Classification of Tumors of the Central Nervous System [11]. It preferentially arises in the spinal cord [12] as an

FDA device/drug status: Not applicable.

Author disclosures: ***TF:*** Nothing to disclose. ***MS:*** Nothing to disclose. ***TY:*** Nothing to disclose. ***YT:*** Nothing to disclose. ***NO:*** Nothing to disclose.

* Corresponding author. Department of Orthopaedic Surgery, Graduate School of Comprehensive Human Sciences, University of Tsukuba, 1-1-1 Tennoudai, Tsukuba, Ibaraki 305-8575, Japan. Tel.: (81) 29-853-3219; fax: (81) 29-853-3214.

E-mail address: funatoru3@gmail.com (T. Funayama)

1529-9430/$ - see front matter
http://dx.doi.org/10.1016/j.spinee.2013.02.066

intramedullary tumor and, less frequently, in other regions of the central nervous system.

Here, we present an extremely rare case of tanycytic ependymoma of the filum terminale associated with MEN-1.

Case report

A 53-year-old man visited a local hospital with complaints of nocturnal pain in the lower back, accompanied by numbness around the anus and intermittent claudication for approximately 1 year. Three years before presentation, he had experienced numbness in both the legs. This symptom had spontaneously disappeared within a year. Magnetic resonance imaging (MRI) of the lumbar spine showed an intradural, large mass lesion at the level from the Th12 to L2 vertebrae. The condition was diagnosed as a spinal cord tumor of the cauda equina region. The patient was subsequently referred to our institute for further evaluation and treatment. His family history was negative. The patient was diagnosed with MEN-1, with multiple pancreatic gastrinomas, at the age of 51 years. His medical history was as follows: resection of pituitary adenocarcinoma at the age of 20 years, recurrent duodenal ulcer since he was 25 years old, total parathyroidectomy and immediate forearm autotransplantation at the age of 43 years, and resection of the transplanted glands at the age of 51 years.

Physical examination

At the time of admission to our institute, the patient's complaints had spontaneously disappeared. However, physical examination showed that the left patellar tendon reflex, both the right and left Achilles' tendon, and bulbocavernosus reflex were reduced. The results of a detailed motor examination via a manual muscle test in both the legs were normal. Sensory loss around the patient's anus was noted. He experienced some dysfunction of the bladder and bowel, including nocturia and mild constipation. Moreover, a urinary test indicated a high risk of retention because of incomplete contraction and relaxation of the bladder.

Laboratory data

The laboratory data of the patient's endocrine function were normal with respect to intact parathyroid hormone, hemoglobin A1c, insulin, adrenocorticotropic hormone, and cortisol levels. However, his gastrin and glucagon levels were slightly high, that is, 289 pg/mL (normal, $<$200 pg/mL) and 210 pg/ml (normal, $<$180 pg/mL), respectively.

Radiological findings

Plain radiographs of the lumbar spine showed no abnormalities. Magnetic resonance images obtained at our institute showed a lesion occupying the intradural space at the level from the Th12/L1 to the L2 vertebrae, with a cranial cystic lesion. The tumor appeared isointense in relation to the spinal cord on T1- and T2-weighted images and was heterogeneously enhanced when gadolinium was used as a contrast agent (Fig. 1A–C). The regions of epiconus and conus medullaris were compressed cranially by the tumor and highly curved. Spinal cord edema and enlargement of the central canal were observed. A string that suggested the thickened filum terminale was connected to the caudal part of the tumor (Fig. 1D). The spinal nerves of the cauda equina were equally separated by the tumor into the right and left sides in the dural space (Fig. 1E). The preoperative diagnosis was myxopapillary ependymoma.

Operation

Open-door laminoplasty of Th12, L1, and L2 was performed using a diamond T-saw (Threadwire Saw; MANI, Inc., Tochigi, Japan) for cutting the middle of the spinous processes and a high-speed burr for creating gutters on both sides of the laminae. The dura mater surface was normal. Dissection of the dura mater and arachnoid mater under a microscope showed an approximately 28$\times$60 mm reddish soft tumor with a rough surface and rich vascularity (Fig. 2). The ends of the tumor were directly connected to the conus medullaris and the thickened filum terminale. Some of the spinal nerves adhered to the tumor. The tumor was carefully separated from these nerves and the conus medullaris and resected *en bloc* with the connected filum terminale. After the dura mater was closed using a continuous suture, the opened laminae were closed by fitting the split spinous processes with nonabsorbable suture threads.

Histologic examination

Histologic examination of the tumor with hematoxylin and eosin (H&E) staining showed that it contained slender and elongated, bipolar, spindle cells, the so-called ''tanycytes,'' with many capillaries (Fig. 3, Top Left). True ependymal rosettes and perivascular pseudorosettes were absent, but a perivascular, anuclear zones were evident (Fig. 3, Top Right). Immunohistochemical staining showed strong, positive staining of the tumor cells for glial, fibrillary, acidic protein (Fig. 3, Bottom); moreover, the tumor showed dot-like, positive staining for epithelial membrane antigen and occasionally positive staining for S-100 protein. These findings indicated a diagnosis of tanycytic ependymoma (World Health Organization Classification Grade II). The mindbomb homolog 1 (MIB-1, also known as Ki-67 antigen) labeling index of the degree of cell proliferation was 4.6%. Pathologic findings suggestive of myxopapillary ependymoma, such as a papillary arrangement of tumor cells, or the deposition of myxoid material in the perivascular region were not obtained.