

Case report

Cervical dumbbell meningioma and thoracic dumbbell schwannoma in a patient with neurofibromatosis

Jin-Cherng Chen^a, Sheng-Hong Tseng^b, Yun Chen^{b,c}, Jeh-En Tzeng^d, Swei-Ming Lin^{b,*}^a Department of Surgery, Buddhist Tzu Chi Dalin General Hospital, Dalin, Chia-Yi, Taiwan^b Department of Surgery, National Taiwan University Hospital and National Taiwan University College of Medicine, 7 Chung Shan S. Road, Taipei 100 Taiwan^c Department of Surgery, Far Eastern Memorial Hospital, Taipei, Taiwan^d Department of Pathology, Buddhist Tzu Chi Dalin General Hospital, Dalin, Chia-Yi, Taiwan

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Abstract

The occurrence of both dumbbell meningioma and dumbbell schwannoma in one patient has not been reported in the literature. We present a 16-year-old female patient, who had progressive bilateral hearing impairment for 5 years and a progressively enlarged, non-tender neck mass for 1.5 years. Mild motor weakness over her right upper limb was noted 1 week before admission. No café-au-lait spot was noted. Magnetic resonance imaging (MRI) revealed bilateral cerebellopontine angle tumors, a C1-2 dumbbell tumor, and a T5-6 dumbbell tumor. Neurofibromatosis type 2 was diagnosed. The cervical spine and thoracic spine tumors were removed via one-staged combined posterior (laminectomy) and antero-lateral (transforaminal or thoracoscopic) approaches during two operations performed 1 month apart. The pathology revealed meningioma and schwannoma, respectively. The patient had good recovery after these two operations and her motor function improved. Six months after the second surgery, radiosurgery was performed for the bilateral acoustic tumors, because of enlarged tumor size on follow-up MRI. To the best of our knowledge, this is the first case reported in the literature of a patient, having both dumbbell meningioma and dumbbell schwannoma. A literature review of the dumbbell tumors was done, and their treatment strategies were discussed. © 2004 Elsevier B.V. All rights reserved.

Keywords: Meningioma; Schwannoma; Dumbbell tumor; Intraspinal tumor; Neurofibromatosis

1. Introduction

Schwannoma and meningioma are the two most common intraspinal tumors [1–5]. Intraspinal schwannomas may be located in the spinal canal, or sometimes, may extend along the root to the extravertebral space through the intervertebral foramen and become dumbbell tumors [6]. In contrast, intraspinal meningiomas are usually located in the spinal canal, and do not extend through the intervertebral foramen [1,5,7]. Therefore, the schwannoma commonly appears as a spinal dumbbell tumor [6], however, rarely, the meningioma appears as a spinal dumbbell tumor [5,6,8–12]. In the literature,

only few reports mentioned spinal dumbbell meningioma [5,6,8–12], and there is no study mentioning the occurrence of both a dumbbell meningioma and a dumbbell schwannoma in one patient. In this report, we presented a neurofibromatosis patient who had two dumbbell tumors, with one cervical dumbbell meningioma and one thoracic dumbbell schwannoma. A literature review of the dumbbell tumors was done, and their treatment strategies were discussed.

2. Case report

A 16-year-old female patient had had progressive bilateral hearing impairment for 5 years and a progressively enlarged, non-tender neck mass, just below the right mandibular angle, for 1.5 years. Magnetic resonance imaging (MRI) done at an

* Corresponding author. Tel.: +886 2 3123456x5110;
fax: +886 2 28313787.

E-mail address: tsh@ha.mc.ntu.edu.tw (S.-M. Lin).

other hospital revealed the patient had bilateral cerebellopontine angle tumors, and a cervical and a thoracic spinal tumor. Neurofibromatosis type II (NF2) was diagnosed; however, no surgery was done at that time. However, the neck mass enlarged gradually and mild weakness of her right upper limb was noted 1 week before admission.

On admission, the patient was conscious. No café-au-lait spot was noted. One soft 1.5-cm-diametered subcutaneous tumor was noted over the nuchal area at the C2–3 levels, and a 3-cm-diametered, non-tender, non-movable, firm mass was palpable over the right side of her neck, beneath the right sternocleidomastoid muscle and 1.5 cm below the right mandibular angle. Her hearing was severely impaired bilaterally, and she used body gestures and hand language for communication. The muscle power of her right upper limb was grade 4/5, while that of other limbs was normal. The deep tendon reflex was increased on the right limbs, and the plantar response was extension on the right and flexion on the left. Her sphincter function was intact. MRI of the brain revealed bilateral cerebellopontine angle tumors, which were considered to be acoustic schwannomas (Fig. 1A). In addition, MRI of the cervico-thoracic spines revealed a cervical dumbbell tumor over the C1–2 levels and a thoracic dumbbell tumor over the T5–6 levels, multiple cervical intraspinal tumors, and a subcutaneous tumor over the nuchal area at the C2 level (Fig. 1B–E). The cervical dumbbell tumor, measuring 4.5 cm \times 2 cm \times 2.5 cm, was multilobulated and caused

widening of the right C1–2 intervertebral foramen. The cervical tumor was hypointense in both T1 and T2 weighted MRI and well enhanced by gadolinium (Fig. 1B and C). The thoracic dumbbell tumor, measuring 6.5 cm \times 3.5 cm \times 3.5 cm, was also multilobulated and caused widening of the left T5–6 intervertebral foramen. The thoracic tumor was isointense in both T1 and T2 weighted MRI and well enhanced by gadolinium (Fig. 1D and E). Both dumbbell tumors extended from the spinal canal (intraspinal component) through the intervertebral foramen (foraminal component), and finally to the paraspinal area (extravertebral component), with the extravertebral component the largest. The intraspinal component of the cervical dumbbell tumor was located at the right side of the spinal canal and displaced the spinal cord to the left, while the thoracic one was located at the left side of the spinal canal and displaced the spinal cord to the right side.

First, the patient underwent operation to excise the cervical spine tumor. A one-staged operation was performed with intraspinal component excised via laminectomy first and then the foraminal and extravertebral components were excised via antero-lateral transforaminal approach. During the operation, the patient was put in the prone position and a C1–2 laminectomy was done. When the dura was opened, a pinkish, firm, hypervascular tumor was noted at the right side of the spinal canal, with the spinal cord displaced to the left. The tumor was in between the dorsal and ventral branches of the left C2 root. Although, the left C2 root was easily

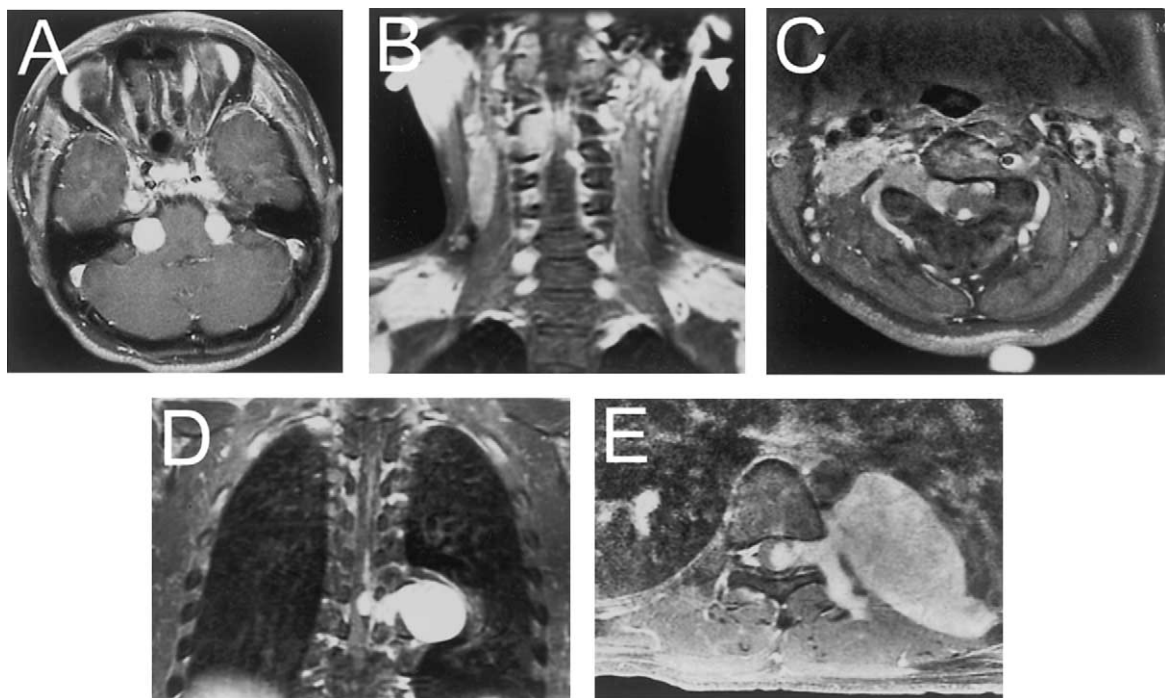


Fig. 1. Gadolinium-enhanced T1 weighted magnetic resonance imaging of the brain, cervical, and thoracic spines: (A) axial-section of the brain, showing bilateral well-enhanced cerebellopontine angle tumors. (B and C) Coronal (B) and axial (C) sections of the cervical spines, showing a well-enhanced multilobulated, dumbbell tumor over the C1–2 levels. The intraspinal component of the tumor was located on the right side of the spinal canal and pushed the spinal cord to the left. A subcutaneous tumor was also noted over the nuchal area at the C2 level. (D and E) Coronal (D) and axial (E) sections of the thoracic spines, showing a well-enhanced multilobulated, dumbbell tumor over the T5–6 levels. The intraspinal component of the tumor was located on the left side of the spinal canal and displaced the spinal cord to the right side.

separated from the tumor, however, the dorsal branch of the left C2 root was cut to facilitate tumor excision. After excision of the intraspinal component of the tumor, the tumor was found to be tightly adhered to the posterior wall of the foramen, and part of the foraminal component of the tumor was excised too. Then, the patient was put in the supine position and a right antero-lateral transforaminal approach was performed to excise the foraminal and extravertebral components of the tumor. The tumor extended from the posterior wall of the foramen to the whole foramen and to the extravertebral space along the anatomical cleaving plane of the left cervical plexus. The left C2 root was displaced superiorly by the tumor, however, there was no adhesion between the tumor and the left cervical plexus. The tumor was nearly totally excised, but a small piece of tumor over the posterior wall of the intervertebral foramen remained because of tight adhesion. The patient withstood the operation well. The pathology of the tumor revealed meningioma (Fig. 2A). The postoperative

course was uneventful, and her neurological status had no significant change after the operation. One month after the first operation, we performed another one-staged operation to remove the thoracic dumbbell tumor via laminectomy first and then the left antero-lateral thoracoscopic approach. During the operation, the patient was put in the prone position and T5–6 laminectomy was done. When the dura was opened, a yellowish, soft, elastic, hypervascular tumor was noted at the left side of the spinal canal with the spinal cord displaced to the right. The intradural component and part of the foraminal component of the tumor was excised. Then the patient was turned to the right lateral position and the left antero-lateral thoracoscopic approach was used. The extravertebral and foraminal components of the tumor were totally excised. Pathologic examination revealed a schwannoma (Fig. 2B). The patient also had good recovery after the second operation.

Because bilateral acoustic tumors were enlarged on follow-up MRI, radiosurgery was performed for these two tumors 6 months after the second operation. After 10 months of follow-up, the patient was well, and the muscle power of her right upper limb became normal. When reviewing her family history, it was noted that the patient's mother also had been found to have bilateral acoustic tumors and diagnosed as NF2.

3. Discussion

This report presents a rare case with both cervical dumbbell meningioma and thoracic dumbbell schwannoma. Spinal dumbbell schwannomas, 10–15% of all the spinal schwannomas, are quite common [5]; by contrast, spinal dumbbell meningioma is rare [5,6,8–12]. Because dumbbell tumor is considered a typical feature for schwannoma [5,6], we initially assumed both the cervical and thoracic tumors in this presented patient were schwannomas. However, the cervical spine tumor turned out to be a meningioma, although the thoracic tumor was a schwannoma. In the literature, to the best of our knowledge, a dumbbell meningioma and a dumbbell schwannoma has never been reported to occur in one patient and this is the first report to present such case.

Spinal schwannomas generally arise from the Schwann cells of the dorsal nerve roots, thus they commonly form dumbbell tumors along the nerve root [5]. In contrast, spinal meningiomas often appear as globular tumors, because the tumors originate from the arachnoid membrane, and approximately 90% are intradural in location [1,3,5,13,14]. Occasionally, meningiomas present as extradural or intradural/extradural tumors and exhibit extravertebral extension [15,16]. The extravertebral extension of meningiomas occurs by tumor growth through the intervertebral foramens, mostly to a minor extent [7,8,15,16]. However, rarely, as seen in this patient, the extravertebral component may become large and make the meningioma appear as a dumbbell [6,10,13]. The origin of the dumbbell meningioma is probably from the arachnoid villi at the nerve root exits [6,10,13]. Because, there is little intraspinal space for tumor growth, a menin-

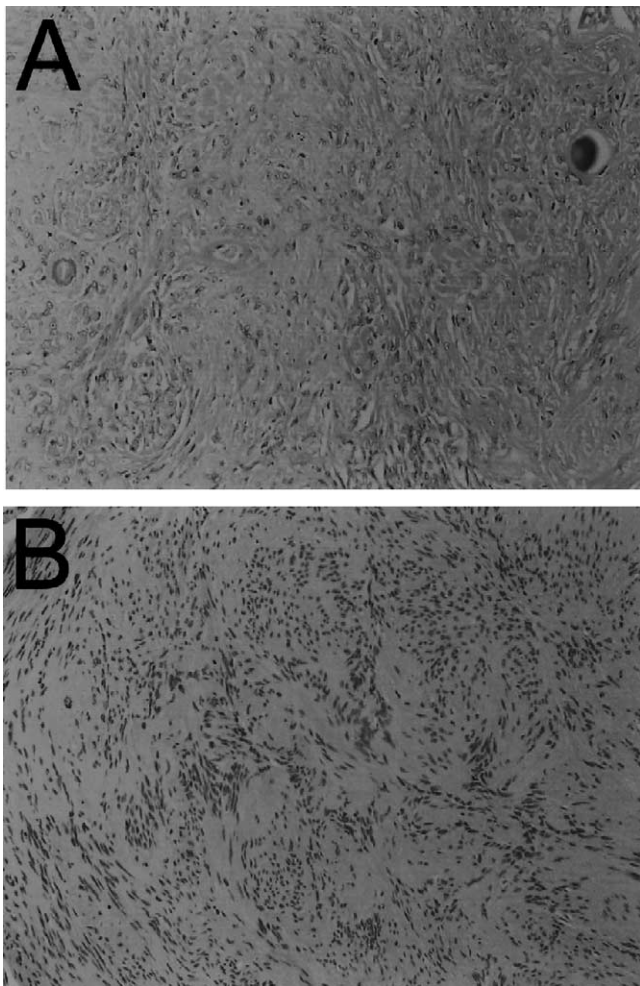


Fig. 2. Histological features of the cervical and thoracic dumbbell tumors. (A) The pathology of the cervical dumbbell tumor, showing a meningioma composed of nests or whorls of tumor cells among prominent fibrous stroma. Some psammoma bodies were noted. (H and E, 100 \times) (B) The pathology of the thoracic dumbbell tumor, revealing a schwannoma composed of spindle cells with palisading pattern (H and E, 100 \times).

Table 1
Clinical features of the patients with dumbbell meningiomas in the literature

Case [Ref] ^a	Sex	Age	NF ^b	Location	Pathology	Previous operation ^c	Surgery
1 [9]	M	60	–	C6–T1	Meningioma	–	Single approach (transuncodiscal approach)
2 [10]	M	52	–	T3–5	Angioblastic meningioma	+	Combined approaches (two stage) (laminectomy + thoracotomy)
3 [10]	F	29	–	T1–2	Meningothelial meningioma	+	Single approach (laminectomy)
4 [6]	F	26	–	T7	Malignant meningioma	+	Single approach (lateral extracavitary approach)
5 [12]	F	16	NF1	C2–4	Transitional/syncytial meningioma	–	Single approach (laminectomy)
6 [8]	F	72	–	C7–T2	Meningotheliomatous meningioma	–	Combined approaches (two stage) (laminectomy + thoracotomy)
7 [11]	M	34	–	T1–2	Atypical meningioma	–	Combined approaches (two stage) (Laminectomy + thoracotomy)
8 [5]	M	58	–	T11–12	Fibroblastic meningioma	–	Combined approaches (one stage) (hemilaminotomy + thoracoscopic approach)

^a The reference the case was cited from.

^b Neurofibromatosis.

^c History of previous operation for intraspinal meningioma.

gioma at this location is prone to grow through the dura and subsequently to the extradural/extravertebral space [10,15]. The tumor of our patient was also considered to arise from the arachnoid villi at the nerve root exits because it was tightly adhered to the posterior wall of the intervertebral foramen.

In the English literature, eight patients were found to have dumbbell meningiomas [5,6,8–12] (Table 1). Of these eight patients, 4 were younger than 35, and neurofibromatosis type 1 (NF1) was suspected in one patient who was 16 years old [12]. Five patients had tumors in the thoracic spine, two in the cervico-thoracic junction, and one in the cervical spine. Six tumors were benign meningiomas, and one tumor was atypical and one was a malignant meningioma. Three patients had previous surgery for the intraspinal meningiomas, and the tumors recurred to become dumbbell [6,10]. The clinical features of these patients suggested that young age and previous operation for intraspinal meningioma might be related to the development of dumbbell meningioma.

Meningioma seldom occurs in patients younger than 20 years old [3], and spinal meningiomas in children and adolescents raise the possibility of underlying neurofibromatosis [17,18]. Our patient was young (16-year-old) and had NF2, while the other young patient reported previously was suspected to have NF1 [12]. Therefore, combined neurofibromatosis and young age were possibly contributing factors in the development of dumbbell meningioma. As to the role of previous operation for intraspinal meningioma in the development of a dumbbell tumor, we considered that coagulation of the dura near the foramen during excision of the meningioma might cause dural necrosis or a small dural defect, which was a potential route for tumor grow-in whenever the tumor recurred, and eventually the tumor became dumbbell-shaped. Because of the small case number of dumbbell meningiomas reported in the literature, more experience on this disease is necessary before making conclusive comments about the contributing factors of the development of dumbbell meningioma.

The treatment of dumbbell tumors is primarily surgical. To decide on the surgical approach for the operation of a dumbbell tumor, we must consider thoroughly the patient's general condition and age, tumor size, axial location and level, relationship to the neural structures, and spinal stabilization requirements [5,6,8,9,19]. Surgical treatment strategies of spinal dumbbell schwannomas were commonly known [5,6,8,9,19], in contrast, surgery for a dumbbell meningioma deserved special consideration. For our patient, at first, the cervical tumor was operated on via a laminectomy to excise the intraspinal portion and decompress the spinal cord, however, the tumor turned out to be a meningioma arising from the dura mater of the posterior wall of the spinal foramen. In addition, the tumor grew along the left C2 root to the extravertebral space and the extravertebral component of the tumor extended rostrocaudally for about three spinal levels. Thus, a right antero-lateral transforaminal approach was used to excise the foraminal and extravertebral components of the tumor because this approach provided ample exposure to excise the extravertebral component of the tumor.

In summary, we presented a neurofibromatosis type 2 patient who had two dumbbell tumors, one was a cervical dumbbell meningioma and the other was a thoracic dumbbell schwannoma. Dumbbell meningioma is rare and co-occurrence of dumbbell meningioma and dumbbell schwannoma in one patient is even rarer. Each of these two tumors was excised via one-staged posterior (laminectomy) and antero-lateral (transforaminal or thoracoscopic) approaches. The patient had good neurological recovery.

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