A Case of Schwannomatosis, Distinct from NF I & II

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ABSTRACT

Multiple schwannomas without vestibular schwannomas (schwannomatosis) are new classification of neurofibromatoses. Understanding of this disease entity is emerging, recently. We experienced a case of schwannomatosis, and reviewed some literatures related to schwannomas. A 45-year-old woman visited our hospital with low back pain and radiating leg pain. The patient’s spine MR images revealed that she had multiple intradural extramedullary masses in lumbar spinal canal, extending to paravertebral space through neural foramen. On brain MR images, there were no vestibular schwannomas. We took an operation to resolve her symptoms. After we removed the intradural extramedullary masses in lumbar spinal canal, her symptoms were improved.

KEY WORDS: Schwannomatosis · Neurofibromatosis · Multiple schwannomas.

Introduction

Schwannomas are benign tumor of nerve sheath origin, and they can develop anywhere, where there exists nerve sheath. Several subtypes of schwannaoma are described in the literature, but only neurofibromatosis Type I (NF 1) and Type II (NF 2) are well-known. Multiple schwannomas without vestibular schwannomas (Schwannomatosis) may be a new classification of neurofibromatoses. Understanding of this disease entity is emerging, recently.

Schwannomatosis is as common as NF 2, and recognized third major type of neurofibromatoses.2 There are several clinical points that distinguish schwannomatosis from NF 1 and NF 2. In general, café au lait spots, skin fold freckling, Lisch nodules occur in NF 1, and bilateral vestibular schwannomas are the hallmark of NF 2. But these features do not occur in schwannomatosis. In genetic point of view, schwannomatosis shares chromosome 22 with NF 2, but schwannomatosis has different locus from NF 2.8,9 We experienced a case of multiple spinal schwannomas without vestibular schwannomas.

Case Report

A 45-year-old woman, visited our hospital with back pain and radiating leg pain. Several years ago the pain started, and progressed day by day. We could not find any abnormal neurologic finding on her leg except radiating pain. And she has no cranial nerve deficits. Neuroimaging study revealed that multiple intradural extramedullary masses in lumbar spinal canal, extending to paravertebral space through neural foramen (Fig. 1A, B). Under suspicion of NF 2, brain MR images (Fig. 1C) were performed. However, we could not find evidence of vestibular schwannomas in brain MR images. There was also no abnormality on ophthalmologic examination. In addition, she had no familial history.

We thought the tumors were the major cause of her pain. L2−5 laminectomy and intradural extramedullary tumor re-
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moval was done (Fig. 2, 3). On surgical field, the tumors were originated from dorsal rootlet and the origin rootlet was intact, relatively. Tumors were compressing the other rootlets posterolaterally. In actually, after the surgery, she didn’t complain any pain on her leg, but mild back pain remained. The tumor was ancient schwannoma on the pathologic report (Fig. 4).

Discussion

Schwannomas are slow-growing benign tumors of nerve sheath origin. Several subtypes of schwannoma are described in the literature, but only neurofibromatosis Type I (NF 1) and Type II (NF 2) are well-known. NF 1 is the most common among the neurofibromatoses family (> 90% of neurofibromatoses), and it occurs in approximately 1/2,500 at birth. NF 2 is occurring in about 1/25,000 at birth, and more aggressive than NF 1, because of the higher incidence of intracranial tumors. NF 1 and NF 2 are known as autosomal dominant tumor syndrome. But over 50% of patients have no familial history, and occur sporadic mutation of genes. NF 1 is associated with a