

False localizing sign caused by schwannoma in cervical spinal canal at C1-2 level

A case report

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Abstract

Rationale: False localizing sign means that the lesion, which is the cause of the symptom, is remote or distant from the anatomical site predicted by neurological examination. This concept contradicts the classical clinicoanatomical correlation paradigm underlying neurological examinations.

Patient concerns: A 54-year-old man consulted for the right sciatica-like leg pain that had aggravated 1 year ago. Radiological examinations revealed degenerative spondylolisthesis with instability and right-sided recess stenosis at the L4–5 level. After initial improvement following 3 transforaminal epidural steroid injections with gabapentin and antidepressant medication, there was a recurrence of the symptoms a year later, along with wasting of the right leg for several months. Physical examination revealed difficulty in heel-walking and a weakness of extension of the right big toe; tendon reflexes were normal. Lumbar spine radiographs revealed no new findings. The initial course of treatment was repeated, but was ineffective.

Diagnoses: Further cervicothoracic spine evaluations revealed a right-sided intradural-extramedullary mass and myelopathy at the C1–2 level.

Interventions: The cervical mass was surgically resected and identified histopathologically as a schwannoma.

Outcomes: Immediately after surgery, sciatica-like pain and weakness of right leg were completely resolved.

Lessons: It is difficult to make an accurate diagnosis if there are symptoms caused by false localizing sign. Additionally, it is even more difficult to diagnose false localizing sign accurately when there is a co-existing lumbar lesion that can cause the similar symptoms.

Abbreviations: C = cervical, L = lumbar, MRI = magnetic resonance imaging, TFESI = transforaminal epidural steroid injection, VAS = Visual Analogue Scale.

Keywords: false localizing sign, myelopathy, schwannoma, sciatica, spinal cord compression

1. Introduction

In 1904, Collier proposed the concept of the false localizing sign for the first time.^[1] It can be described as a state in which the anatomical location of the lesion causing symptoms is distant or remote from the anatomical locus predicted by neurological examination.^[2] This concept contradicts the classical clinicoanatomical correlation paradigm underlying neurological examinations. The symptoms caused by false localizing signs most

likely result into missed or delayed diagnosis. Furthermore, these misleading signs lead to unnecessary or incorrect treatments and even surgical procedures at an unaffected site. In particular, co-existence of lesions in the lumbar region, which are likely to cause similar symptoms based on conventional neurological examinations, increases the difficulty in establishing accurate diagnosis and initiating proper treatment. Sciatica-like leg pain owing to cervical cord compression is a very rare symptom of false localizing sign and only a few clinical cases have been reported.^[3–7] Among them, in 1967, Langfitt and Elliott^[4] reported that cervical spinal cord compression caused by a tumor or degenerated disc material might cause lower back or leg pain that can easily be confused with a lumbar disc syndrome and accurate differential diagnosis is too easily impeded by abnormal findings in both cervical and lumbar region. Until now, however, no reports have described leg pain or sciatica as a false localizing sign caused by a schwannoma at cervical spinal canal.

The authors concluded that the cause of the patient's right sciatica-like leg pain was a schwannoma in the cervical spinal canal at C1–2 level, as clearly revealed by the results of the surgery. We report this case as a typical false localizing sign.

2. Case Report

A 54-year-old man presented with right sciatica-like leg pain that began three years ago. This throbbing pain originated from the right buttock and radiated into the posterolateral thigh and leg

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Ethical approval: This case report is not a clinical trial and just incidental interventional process, so ethical approval was not necessary.

The authors report no conflicts of interest.

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and dorsal surface of the foot. He also reported lower back pain in addition to paresthesia and numbness in his right leg that had started worsening approximately one year ago. He had received numerous examinations and various kinds of conservative treatments for the lumbar spine in different hospitals, but no improvement was noted. His Visual Analogue Scale (VAS) score was 6 or 7 out of 10. He underwent laminectomy at L3–4 level six years ago. There was nothing special about his medical history. Physical examination revealed no particularities. Lumbar spine radiographs (Fig. 1) and magnetic resonance imaging (MRI) (Fig. 2) showed degenerative spondylolisthesis with instability and right-sided recess stenosis at L4–5 level. It was considered to be the cause of symptoms. Transforaminal epidural steroid injection (TFESI) was performed at right-sided L5–S1 level every two weeks for three times. After the procedure, his lower back and right sciatica-like leg pain were reduced to VAS score 2 or 3 out of 10, and he showed great satisfaction with the results. However, paresthesia and numbness in the right leg persisted. Thus, gabapentin and antidepressant were prescribed. Three months later, his lower back and right sciatica-like leg pain were reduced to VAS 1 or 2 out of 10, and the improvement in paresthesia and numbness was also reported. He reported no difficulties in daily life. After the cessation of medication for one month, no changes in the symptoms were noted. The authors recommended him to return for consultation in the case of relapsing of symptoms or occurrence of new symptoms.

He returned to the pain clinic one year later and reported recent deterioration of initial symptoms. Nature and radiating pattern of recurred right leg pain was similar to the previous pain. Since his place of residence was so far, he also visited the pain clinic after going through several hospitals. He reported that peripheral nerve abnormalities had been identified on the electromyogram performed at another hospital several months earlier. From several months ago, he felt that his right leg was tapered slightly, although this did not affect his daily activity but there was no significant difference between the two legs in actual measurements. This time, physical examination revealed several abnormal neurological findings. He showed some difficulty in heel gait. Right great toe extension and ankle dorsiflexion were slightly weak and classified as grade IV, according to the Medical

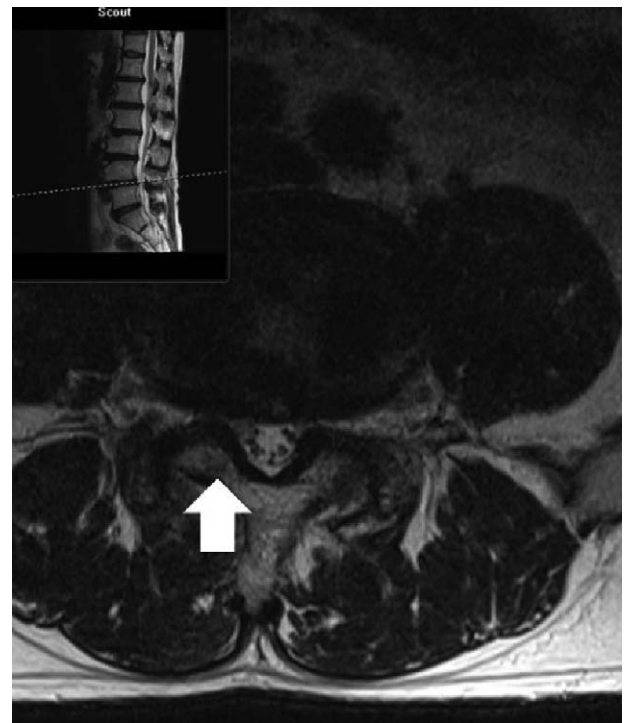


Figure 2. Lumbar magnetic resonance imaging shows right-sided recess stenosis at L4-5 level (arrow).

Research Council Muscle Strength Scale. However, no abnormal reflex sign was present. Radiological examination of the lumbar spine was immediately performed, but no significant changes were observed as compared to the observations 1 year earlier. Same as with the past treatments, the TFESI at the right-sided L5–S1 level was repeated and medications were reinitiated. This time, however, the treatment had no effect. Immediate further examinations of cervical and thoracic spine were conducted, and cervical MRI revealed a right intradural-extramedullary mass with myelopathy at the C1–2 level (Fig. 3A). He was



Figure 1. (A) Flexion view. (B) Extension view. Lumbar spine radiographs show degenerative spondylolisthesis with instability at L4-5 level (arrow).

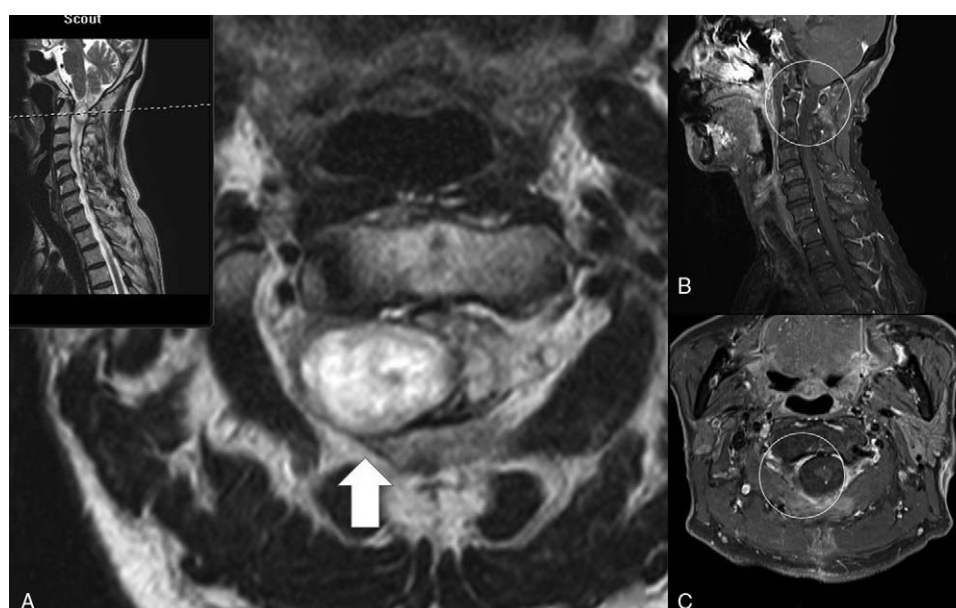


Figure 3. (A) Preoperative. (B and C) Postoperative. (A) Preoperative cervical T2-weighted magnetic resonance imaging (MRI) shows a right intradural-extramedullary mass with myelopathy at the C1–2 level (arrow). (B and C) Postoperative cervical MRI shows a complete removal of the mass after surgery (circle).

immediately referred to the department of neurosurgery, wherein right-sided C1 hemi-laminectomy was performed followed by tumor removal (Fig. 3B and C). Histopathological examination of the excised tissue identified the tumor as a schwannoma. Fortunately, abnormal neurological signs and sciatica-like pain in the right leg completely resolved immediately after the surgery. However, abnormal sensations such as paresthesia and numbness remained to a certain extent, which caused inconvenience. Gabapentin and antidepressant were prescribed again. After 3 months, he reported a great improvement in paresthesia and numbness. Thereafter, he stopped the medication and has been free of symptoms for more than a year.

Approval of this study was waived from the Ethics Committee of Kyungpook National University Chilgok Hospital, based upon their policy on case reports. The authors obtained written consent from the patient to publish this case report.

3. Discussion

False localizing sign can be described as a state in which the anatomical location of the lesion causing symptoms is distant or remote from the anatomical locus predicted by neurological examination.^[2] In 2003, Lerner classified false localizing signs into those caused by intracranial, spinal cord, and other lesions, according to the location of lesion, in addition to describing the nerves affected and the symptoms specific to each case.^[8] Until now, however, no reports have described leg pain or sciatica as a false localizing sign caused by a schwannoma at cervical spinal canal.

Sciatica-like leg pain is a very rare symptom of false localizing sign owing to cervical cord compression. Sciatica represents a symptom rather than a specific diagnosis. The most prominent symptom is the pain in the lower limb radiating to the feet and toes. Sciatica can lead to clinical signs of neurological deficit, such as muscle weakness and a change in reflexes. It can be caused by a variety of conditions and diseases, and most of these are related to lesions affecting the lumbar spine or pelvis. About 90% of these

lesions are because of lumbar herniated disc with nerve-root compression, other than lumbar spinal canal and foraminal stenosis. It can also be caused by tumors or cysts originating from the back or pelvis as well other parts of the body.^[9] Although it is very rare, sciatica-like leg pain can be caused by cervical cord compression. Only a few clinical cases have been reported.^[3–7]

Schwannoma is an intradural-extramedullary nerve sheath tumor that predominantly occurs in the third to fifth decade of life. Tumors are benign in most cases. Although most patients develop symptoms at the stage of diagnosis, a few become symptomatic months or years before diagnosis. The most common symptom of schwannoma is a local or radicular pain, and some patients also report paresthesia and numbness.^[10] In 2005, Jinnai et al^[11] performed a retrospective review of 149 patients with spinal nerve sheath tumors. They found that the initial symptoms included motor weakness in 24.2% patients, pain in 36.9% patients, and paresthesia and/or numbness in 35.6% patients. The remaining 3.3% patients were diagnosed incidentally. In particular, when the nerve sheath tumor was located at the level of the first 2 cervical nerve roots or at the cauda equina, motor weakness was hardly observed. The findings for the present case were similar. Although an extensive mass was confirmed by cervical MRI in the present case, it can explain why pain, rather than apparent neurological abnormalities, is the most predominant symptom. They also subdivided their patients into 5 groups according to the correlation between the tumor and the dura mater and/or intervertebral foramen. Tumors localized entirely within the dural sac, such as the tumor in the present case, were categorized as Group 1 tumors, and pain was the most frequent initial symptom associated with this group of tumors. The patient also presented with sciatica-like leg pain as the predominant symptom. In 2016, Murahashi et al^[12] reported the results of the retrospective review of pre-operative neurological and radiological examinations for 24 patients with cervical myelopathy caused by spinal cord compression at C1–2. Although these patients were to be operated, 10 of 24 patients showed normal deep tendon reflex. The degree of spinal cord

compression was more severe in patients with perceptible dysfunction and muscle weakness, but there was no difference in the degree or location of spinal cord compression among the patients with dysfunction. Nevertheless, no significant differences in the level and location of spinal cord compression were observed among patients with dysfunction. The results of these studies are in good agreement with the clinical symptoms of the present case.

In 1967, Langfitt and Elliott^[4] described that cervical spinal cord compression caused by a tumor or degenerated disc material might cause lower back or leg pain that can easily be confused with a lumbar disc syndrome. They described that abnormal findings in both cervical and lumbar region render accurate differential diagnosis very difficult. Furthermore, these authors already described that abnormal findings might be absent in the neurological examination even in the case of an extensive cord compression that can cause severe pain. In addition, mechanical signs, limitation in back mobility, and a positive reaction to the straight leg raising test might be absent even when there is a cord compression. This possibility may be explored in the present case.

In fact, symptoms caused by false localizing signs are rarely reported and no pathological mechanisms underlying the occurrence of false localizing signs have been described so far. However, several hypotheses have been elaborated. In 1956, Scott^[3] described the irritation of ascending spinothalamic tract as the likely cause of sciatica-like lower extremity pain caused by a tumor at high thoracic and cervical cord. Jamieson et al^[13] presented several hypotheses regarding the origin of false localizing sign in their clinical case report. Above all, these authors suggested that myelopathy originated from the destruction of the anterior horn cell by venous obstruction and subsequent static hypoxia, and this hypothesis was evaluated by Taylor and Byrnes in 1974 as the most reasonable.^[14] Another hypothesis was that disinhibition of normal ascending pain-producing pathways that regulate pain signals is interpreted as pain by the brain.^[15] In 2002, Ochiai et al^[16] reported that false localizing sign is likely to be caused by a severe compression of the midline ventral structure within the cervical spinal cord including the anterior spinal artery, leading to ischemia in the thoracic watershed zone of the artery.

After 3 TFESIs, some of the patient's symptoms improved, probably because some of the causes of the patient's right leg pain were owing to lumbar lesions. But it is believed that the schwannoma became enlarged as time progressed, and the right leg pain recurred and the neurologic abnormalities appeared. As described previously, it is very difficult to recognize false localizing sign earlier if lesions are simultaneously present in cervical and lumbar regions.

In the present case, physical examination during the first visit showed no evidence of cervical cord compression, and radiologic examinations showed degenerative spondylolisthesis with instability and right-sided recess stenosis at L4–5 level. In addition, after 3 TFESIs, the patient reported the relief of symptoms and the recovery of normal life. Thus, no other causes to explain the symptoms were investigated. Fortunately, the present case

exhibited a complete recovery immediately after surgery. However, irreversible damage would have occurred had he not undergone the appropriate examinations and received an accurate diagnosis and appropriate treatment in a timely manner. In the present case, the sciatica-like leg pain was induced by a slowly progressing schwannoma in the cervical spinal canal at C1–2 level. However; the identification of degenerative spondylolisthesis with instability and right-sided recess stenosis at the L4–5 level led to a discrepancy between the clinical level and the actual lesion, which is the most typical case of false localizing sign.

It is difficult to recognize that the patient's symptoms are caused by false localizing sign from the beginning of the diagnosis. In addition, it is more difficult to diagnose false localizing sign accurately when there is a co-existing lesion that can cause the same symptoms. In conclusion, although it is rare, the patient's symptoms may be because of a false localizing sign if the patient does not respond to appropriate treatment, so proper diagnosis should be made through appropriate examination to reduce unnecessary treatment.

Author contributions

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