Journal of Cancer Research and Practice 3 (2016) 19–22

Contents lists available at ScienceDirect

Journal of Cancer Research and Practice

journal homepage: http://www.journals.elsevier.com/journal-of-cancer-research-and-practice

Case report

Ancient schwannoma of thoracic spine in a schizophrenic patient with somatic delusion

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ARTICLE INFO

Article history:
Received 15 June 2015
Accepted 19 August 2015
Available online 17 November 2015

Keywords:
Schwannoma
Ancient schwannoma
Somatic delusion

ABSTRACT

Ancient schwannoma is a rare variant of schwannoma characterized by histopathologic degenerative changes, which are thought to be the result of long-term tumor growth and aging. However, ancient schwannoma in the spinal canal is particularly rare. We report a case of thoracic spine intradural extramedullary ancient schwannoma in a schizophrenic patient, who kept saying that "something in his back was giving him electric shock" for a long time. Unfortunately, this complaint was misinterpreted as somatic delusion symptoms. A spinal cord tumor was taken into consideration only after paraparesis developed. We have highlighted this case to remind every clinician to remain alert about the possibility of organic disease while treating patients with psychotic disorder history. Thorough neurological examination is required to avoid misdiagnosis. Spinal canal schwannoma can be totally removed successfully with good functional outcome and prognosis.

1. Introduction

Ancient schwannoma is an uncommon variant of schwannoma, which arises from the nerve sheath Schwann cells, with a typically long course of growth.[1] These tumors are termed "ancient" because of the degenerative features acquired as the tumor ages, such as cystic necrosis, hemorrhage, calcification, relative loss of Antoni type A tissue, and the presence of degenerative nuclei. These changes are believed to occur because it takes a long time for ancient schwannomas to develop.[2] Schwannomas with these degenerative changes can be misdiagnosed as malignant sarcomas or other forms of soft-tissue neoplasms owing to the nuclear pleomorphism.[3]

Ancient schwannoma is usually located deeply in the head and neck, thorax, retroperitoneum, pelvis, and extremities of elderly patients. Spinal canal ancient schwannoma is very rare; there are only few reported cases in the literature.[4–6] The most common symptom of a spinal schwannoma is pain, either localized or radicular, associated with frequent involvement of dorsal sensory nerve roots. Herein we report a case of thoracic spine intradural extramedullary ancient schwannoma in a schizophrenic patient who initially experienced severe back pain, which was misinterpreted as somatic delusion symptoms for an extended duration of time.

2. Case presentation

A 58-year-old man was seen in our emergency room (ER) presenting with bilateral leg weakness, complaining that he could not stand up or walk for two days. His unkempt appearance and incoherent hyper-talkative behavior were also noticeable. He repeatedly stated that something in his back was giving him "electric shock". According to personal and family medical records, this patient many years ago had been diagnosed with schizophrenia, with persecutory delusions.

The patient had complained of a persistent backache for more than one year, but denied any history of trauma or fall. In the last month, he also noted progressive weakness in the lower limbs. Otherwise, there were no upper limb or facial expression involvement.

Upon further review of his medical records, we determined that the patient came to our ER several times approximately two months prior to this admission, also complaining that something in his back was giving him the sensation of an electric shock. However,

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Peer review under responsibility of The Chinese Oncology Society.

http://dx.doi.org/10.1016/j.jcrpr.2015.10.002

the ER doctors could not understand what he was saying about his back pain because of his incoherent speech and psychotic symptoms. The patient's lumbar spine X-ray was checked and only mild degenerative change was found. Pain control pills were given and there was some indication that a preliminary diagnosis of schizophrenic somatic delusion was made. Furthermore, he had also visited an orthopedic surgeon and a neurologist with the same complaint and received similar feedback.

Upon neurological examinations during his current ER visit, muscle power levels in the patient's lower limbs were only grade 1/5 in right side and grade 2/5 in left side. Increased deep tendon reflex, increased spasticity and positive Babinski sign were also detected in both lower extremities. Pinprick sensation decrease was further demonstrated below the T12 level symmetrically, and a thoracic spinal cord lesion was thereafter suspected. A subsequent magnetic resonance imaging (MRI) of the spine showed an intradural extramedullary cystic tumor at T9-T10 level with marked spinal cord compression (Fig. 1).

Due to these findings, a T9, T10 laminectomy and tumor excision was then performed. We noted the presence of a well-defined yellow-grayish appearance cystic tumor attached to a nerve root (Fig. 2). The tumor was grossly total excised with the attached nerve root sacrificed. Subsequent pathology study showed spindle tumor cells with focal nuclear palisading, cystic change, hemorrhage and foamy cell infiltration. Nuclear degenerated atypia was also noted. The patient's immunohistochemistry workup also showed strong positive staining of S-100 protein (Fig. 3). After review of the results of both prior and current testing and physical examination, a diagnosis of ancient schwannoma was thus made.

Postoperatively, there was no detectable neurological deficit referable to T9 or T10 myotome and dermatome. The patient exhibited good recovery of muscle strength in both legs, though recovery was gradual. Additionally, he could walk independently with the assistance of a walker one month after surgery. Lastly, the patient had no residual complaints about the sensation of electric shocks in his back.

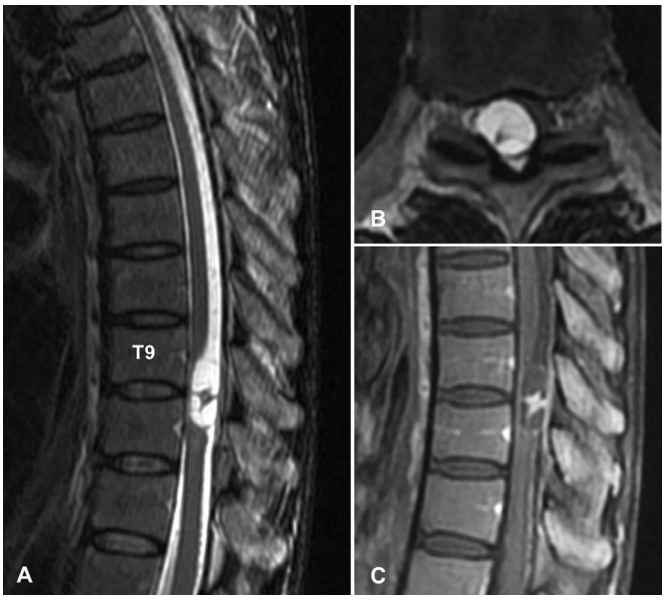

Fig. 1. (A) Sagittal T2-weighted image without contrast, showing a well-defined intradural extramedullary cystic tumor at the T9-T10 level with marked spinal cord compression. (B) Axial T2-weighted image without contrast, showing an eccentric cystic tumor push away the spinal cord to the left side. (C) Sagittal T1-weighted image with contrast, demonstrating good contrast enhancement of the central component.

3. Discussion

Schwannomas or neurilemmomas are benign and slow growing tumors that arise from the peripheral nerve sheath Schwann cells. The tumor cells are arranged in varying cell proportions and arrays named Antoni type A and Antoni type B tissues. Antoni type A area is a compact array of spindle-shaped cells with high cellularity, while Antoni type B tissue is less cellular, with a predominantly loose matrix.[7] When schwannomas demonstrate degenerative pictures including cyst formation, hemorrhage, calcification, diffused hypocellular areas (relative loss of Antoni type A tissue), and degenerative nuclei, they are termed “ancient” schwannoma.[4,5] The nuclear degenerative features, characterized by nuclear hyperchromasia and pleomorphism, are thought to be the result of long-term tumor growth and aging. Immunohistochemically, these tumors also show diffuse positivity for S100 protein in the cytoplasm.[8] This “ancient variant” is an uncommon subtype of schwannoma, which was first mentioned in the literature by Ackerman and Taylor in 1951 to describe schwannomas with calcification.[9]

The ancient schwannoma in our case showed a well-circumscribed cystic mass with heterogeneous enhancing patterns on MRI. Microscopically, the Antoni type B area occupied most of the tumors, and the Antoni type A area was smaller and located mostly around the cystic formation.

For schwannomas, the most useful image tool for pre-operative diagnosis is MRI.[3] Typical findings of ordinary schwannomas are a low signal on T1-weighted images, a high signal on T2-weighted images, and strongly enhanced by gadolinium contrast medium, which is the characteristic of the Antoni type A area.[3,10] In contrast, degenerative cyst formation, hemosiderin deposition, and calcifications are the most useful radiological clues for ancient subtype.[11] They also show different enhancing pattern, primarily enhanced in the circumference of the cyst, hematoma and tumor capsule, while illustrating the feature of Antoni type A tissue loss in the ancient variant.[3] Focal areas of higher signal on T2-weighted image could also be seen in the cystic portions.[12,13]

Most spinal schwannomas derived from Schwann cells along the dorsal sensory roots involve radicular pain as the most common initial symptom. However, symptoms may be subtle and progress slowly over time, owing to the slow-growing nature of these tumors.[6] In our case, it's more difficult to think about a spinal tumor in a schizophrenic patient with persecutory delusion history who complains that “something in his back was giving him electric shock”. Moreover, the back pain complaint was relatively nonspecific and could be multifactorial. A thorough and detailed neurological examination is required to evaluate patients presenting with back pain for any signs of myeloradiculopathy or abnormal reflexes.

We highlighted this case for another purpose, namely to remind every clinician about the possibility of organic disease in patients with psychotic delusions. A wide variety of organic diseases masquerade as psychiatric disorders.[14] Clinicians may be distracted easily by a patient's chief presentation of psychotic delusions, hallucinations, or anxiety, and ignore the possibility of organic disease. This case reminds us of the danger of misdiagnosis, and should encourage every clinician to remain alert to etiological clues when obtaining medical histories and during examination, especially when treating patients who have a history of psychotic disorder.[14]

The treatment of choice for spinal canal schwannoma is complete microsurgical resection. Solitary schwannomas are rarely found, and infrequently manifest malignant degeneration. The

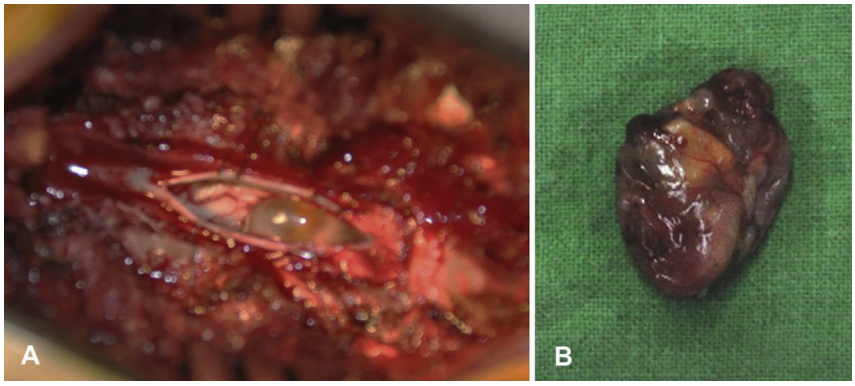

Fig. 2. (A) Intraoperative picture after dura opening, showing a well-defined extramedullary cystic tumor in the dorsal side of the spinal cord. (B) Gross appearance of the tumor: a yellow-grayish and soft tumor, measuring 1.6 × 1.5 × 0.8 cm^3 in size. The tumor is excised in total.

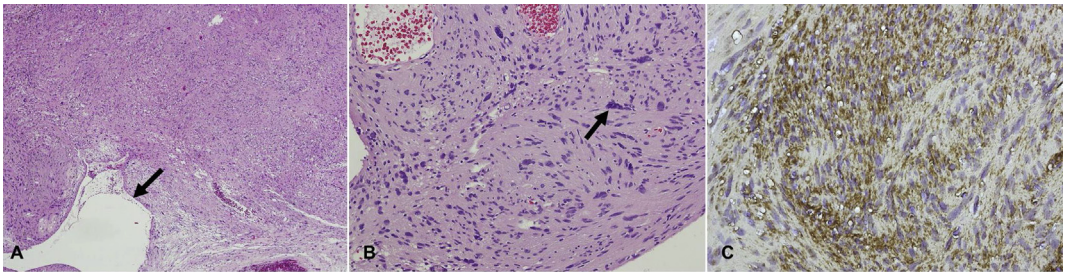

Fig. 3. Histopathological images of the tumor specimen: (A) Spindle tumor cells with focal nuclear palisading, cystic change (Arrow), hemorrhage and foamy cell infiltration (H&E stain, 100X). (B) Nuclear hyperchromasia (Arrow) and pleomorphism of tumor cell under high power view (H&E stain, 200X). (C) Immunohistochemistry showed strong positive staining for S-100 protein (200X).

prognosis of spinal schwannoma is good when complete tumor resection is achieved. They rarely recur after gross total resection, except in cases of schwannomatosis.[15]

The major goals of spinal schwannoma surgery are to perform neural decompression and total tumor resection without damaging adjacent neural tissue. Radical tumor removal sometimes requires that the nerve root origin involved in the schwannoma be sacrificed. However, several studies have reported the risk of neurological deficits following resection of the nerve roots involved in schwannomas. Kim et al[16] reported that 7 of 31 patients (22.5%) developed partial loss of motor strength or sensation postoperatively. Ohnishi et al[17] reported that 3 of 15 patients developed neurological deficits. Two of 6 patients (33.3%) with ventral nerve root origin tumors had minor postoperative motor weakness. Only 1 of 9 patients (11.1%) with dorsal nerve root origin tumors had postoperative segmental sensory numbness. In our case, there was no resulting neurologic deficit from dorsal root sacrifice. Similarly, Schirmer et al[18] even demonstrated that resection of dorsal nerve root tumors resulted in almost no detectable loss of skin sensation. Dermatomes overlapping from adjacent nerve roots could be one possible explanation.

Intraoperative electrophysiological monitoring including neurostimulation is a useful tool to establish the function of nerve root before sacrifice.[7,19] Free-running electromyography (EMG) and electrical stimulation with compound muscle action potential (CMAP) recordings, also called triggered EMG, could be used to monitor and identify root function. A monopolar stimulation electrode is used to elicit evoked EMG responses from the affected root, or any suspicious tissues that may contain neural structures before resection. Additionally, somatosensory evoked potentials (SSEP) and transcranial motor evoked potentials (MEP) could prove efficacious in monitoring the integrity of the spinal cord. SSEPs monitor the dorsal column-medial lemniscus pathway, while MEPs monitor the corticospinal tracts. The combined use of SSEPs, MEPs, and both spontaneous and triggered EMG provide optimal monitoring of spinal cord function and minimize intraoperative neurological injury.

4. Conclusions

Ancient schwannoma is a benign and slow-growing tumor which takes a longer period of time to develop, and is particularly rare in the spinal canal. Most common initial presentations of spinal canal schwannoma involve nonspecific back pain. Therefore, a thorough and detailed neurological examination for myeloradiculopathic clues is required to avoid misdiagnosis. Besides, we should remain alert to the possibility of organic disease while treating patients with psychotic disorder history. The treatment of choice for spinal schwannomas is complete surgical resection. These tumors can be successfully and totally removed, with a good prognosis, and even more so enhanced with the aid of intraoperative electrophysiological monitoring.

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