Multiple schwannomas of cauda equina in the absence of von Recklinghausen's disease

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ABSTRACT

Multiple schwannomas in the absence of neurofibromatosis is rarely reported in the literature. We present a 56-year-old female with a history of severe leg and back pain on the left side for one year. Magnetic resonance imaging revealed 4 schwannomas located in the cauda equina in the absence of von Recklinghausen's disease.

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Multiple schwannomas have been reported in association with neurofibromatosis.[1,2] Our literature review could reveal only one previously reported case of multiple schwannoma located in the spinal intradural space without any evidence of neurofibromatosis.[3] We present a unique case of multiple schwannoma at the cauda equina region without any evidence of neurofibromatosis. The aim of this report is to describe the clinical and radiological features of this rare condition.

Case Report. A 56-year-old female was referred to our clinic with a one-year history of back pain spreading to the left leg, resulting in motor loss of the ankle and toe. Neurological examination showed a weak left ankle dorsiflexion and toe dorsiflexion with a 3/5-muscle power. Sensory examination demonstrated the presence of hypoesthesia at the dorsal aspect of the left foot. No pathological reflex was observed. Sphincter tonus was normal and no skin lesion was found. Her past medical history and family history revealed no evidence in favor of neurofibromatosis. In addition, the physical examination of her parents did not reveal any neurological of cutaneous findings of neurofibromatosis. Magnetic resonance imaging (MRI) showed 4 intradural schwannomas located in the cauda equina at spinal levels L2 and L3, resembling beads. Lesions were well bordered, mildly hyperintense in T1-weighted images, hypointense in T2-weighted images, and they had increased signal intensity on gadolinium-enhanced T1-weighted images (**Figure 1**). Laminectomy was performed at the level of L2-3. Following dural incision, 3 of the schwannomas originating from the same nerve and the other schwannoma originating from another nerve were removed. Pathological examination of the specimen showed that the excised masses were schwannomas. Muscular power and sensory loss improved post operatively.

Discussion. Nerve sheath tumors and meningiomas are the most common intradural extramedullary tumors.[2] There are 2 types of nerve sheath tumors in the spine widely known as schwannomas and neurofibromas, totally constituting 25-29% of primary intraspinal neoplasms.[3,4] Both tumors are primarily made up of Schwann cells, with the exception of optic and olfactory nerves originating from cranial and peripheral nerves. They can originate from the nerve roots within the spinal cord or can be seen in the distal part of the spine. They have also been identified in the skin, oral cavity, and lacrimal glands.[5]

Multiple schwannomas are extremely rare, and their presence has been identified mostly in association with neurofibromatosis.[3,6] Schwannomas identified in this form can suggest the presence of a tumor in the central nervous system. Doi et al[7] were the first to introduce the case of multiple intracerebral schwannomas. Without other findings of neurofibromatosis, occurrence of multiple schwannomas is rare. Attie et al[5] for the first time reported multiple schwannomas in the brachial

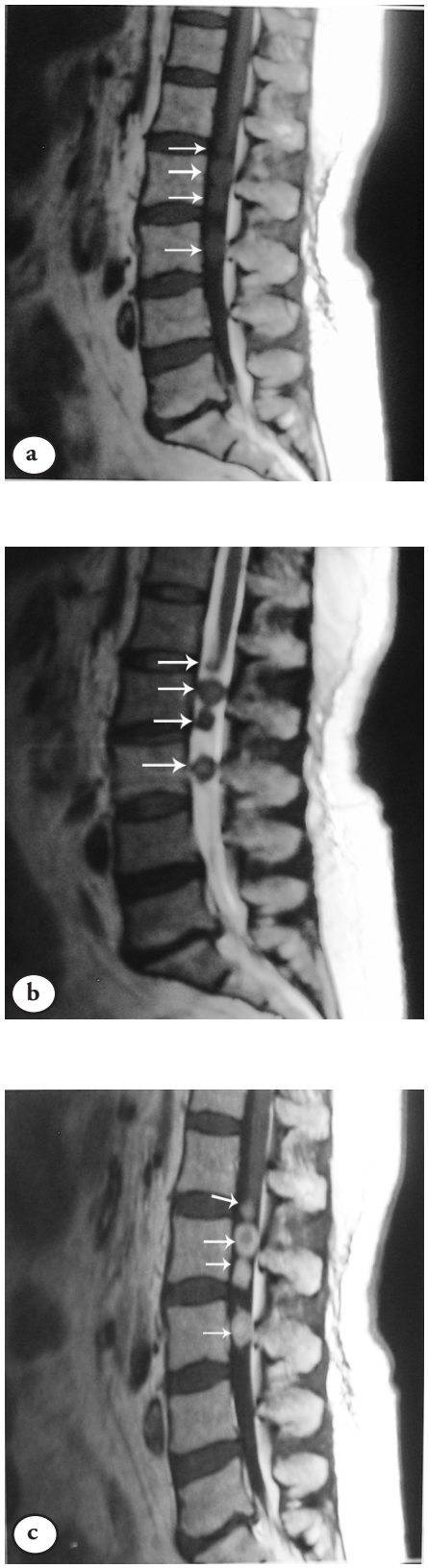

Figure 1 - Sagittal magnetic resonance images of the lumbar spine demonstrating the presence of 4 lesions at the cauda equina region, **a**) T1-weighted image, **b**) T2-weighted image, and **c**) Gadolinium-enhanced T1-weighted image.

plexus and submandibular glands. This case did not have any finding of von Recklinghausen's disease. Likewise Daras et al[3] first reported multiple spinal intradural schwannomas in the absence of von Recklinghausen's disease. A subsequent study conducted by Altinors et al[8] reported a case with 3 schwannomas at the spinal region, and one intracranial schwannoma without evidence of neurofibromatosis, and defined them as "craniospinal schwannomatosis" to emphasize that the schwannomas were not solely confined to the spine.

Our case is the first multiple spinal intradural schwannomas in the absence of von Recklinghausen's disease at the cauda equina region, which is quite different from what was reported by Daras et al.[3] Clinical signs are similar to disc herniation, radiculopathy, and low back pain being the most common symptoms. Other symptoms include paresthesia and weakness of the extremities. If the tumors compress the spinal cord, myelopathy signs inevitably develop.[1,2] Subarachnoidal hemorrhage may also be seen with tumor located at the cauda equina.[8,9] Our patient also had signs and symptoms of radiculopathy. The MRI showed 4 schwannomas at the cauda equina region in the size of beads with mild hyperintense appearance in T1 scan, and hypointense appearance in T2 scan. Cervical, thoracic, and cranial MRI results were normal. Neither our patient nor her parents had any skin or ophthalmological lesions suggestive of neurofibromatosis type 1 and neurofibromatosis type 2. All the tumors were surgically excised.

In conclusion, most nerve sheath schwannomas are solitary and benign tumors. Multiple schwannomas are frequently seen in association with neurofibromatosis and rare in its absence. The presence of a single schwannoma may entail a more detailed examination of the neural axis, and MRI investigation may be necessary to detect the possible presence of other lesions even in the absence of neurofibromatosis.

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