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A Purely Paraspinally Located Schwannoma from a Dorsal Root of a Spinal Nerve

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Authors' contributions

Both authors contributed equally to this work. Author VO wrote the manuscript. Authors VO and CS interpreted the images and accompanied the patient. Author CS performed the surgery. Both authors read and approved the final manuscript.

Review Article

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ABSTRACT

Background: Spinal Schwannomas are benign nerve sheet tumors and the most common intradural extramedullary tumors of the spine, mainly occurring in the cervical and lumbar region.
Case Description: Here we illustrate the case of a 34-year-old woman with a paraspinal lesion presenting with severe lumbar pain. There were no focal sensory or motor deficits. Magnetic resonance imaging (MRI) disclosed a nodular lesion embedded in the paraspinal lumbar musculature, suspicious of tumorous growth. Complete surgical excision of the lesion was performed with clear margins and neuropathological analysis revealed a Schwannoma. Postoperative imaging confirmed a gross total resection with no recurrence after 6 months.
Conclusion: Although Schwannomas are frequently encountered in the lumbar spine, a location distinct from the spinal canal is very rare. To be adequately treated, it needs to be considered in the differential diagnosis of any spinal or paraspinal mass. As with other accessible and symptomatic lesions, the treatment of choice is complete excision with clean margins to avoid local recurrence.

Keywords: Schwannoma; neurinoma; paraspinal lesion; lumbar tumor.

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1. INTRODUCTION

Schwannomas are benign nerve sheet tumors which rarely undergo malignant transformation, representing approximately 30% of all spinal neoplasms and show a male-to-female predilection of 1.23 to 1.5:1, slightly favoring males [1-4]. Occurrence is sporadic and the incidence in Caucasians is estimated to be 0.3 – 0.4 cases per 100.000/persons per year. Although Schwannomas have been linked to some inherited syndromes such as neurofibromatosis type 2 [5], schwannomatosis and Carney complex, there is no known genetic trait for de-novo spinal Schwannomas.

Spinal Schwannomas mostly occur intradurally (>80%), but sometimes extend extradurally through the intervertebral foramina [6] assuming a typical dumb-bell shape [7]. They usually appear as painless, well encapsulated firm tumors arising from a single nerve fascicle and mainly occur in the cervical and lumbar regions [8].

Depending on location and size, patients present with- pain upon pressure, radicular symptoms, sensory impairment or if intracanalicular- paraparesis [9,10].

Contrast enhanced MR-Imaging is the neuroradiological gold standard for assessment. Surgical removal of the lesion with curative intent is the treatment of choice for symptomatic lesions or lesions progressing in size on sequential scans.

To the best of our knowledge this purely extraspinal occurrence is the first case to be reported for a Schwannoma in this location.

2. PRESENTATION OF CASE

A 34 year old female without pertinent familial past medical history presented with lumbar back pain characterized as deep, dull and diffuse. There were no sensory or motor deficits. Sagittal T2 weighted and contrast enhanced T1 weighted MRI showed a clearly demarcated tumorous growth, lateral to the spinous process of L4 located within the right multifidus muscle measuring 1x1.3 cm in size (Fig.1A-1C). There was no indication of bony destruction of the L4 posterior elements. Based on radiographic criteria differential diagnosis included neurofibroma, paraganglioma, pallisaded encapsulated neuroma, myxoma, fibroma, liposarcoma and metastatic disease amongst others. Due to the debilitating symptom of this tumor as well as the concern for a potentially more aggressive neoplasm, the patient opted for an open surgical approach with the intent of a complete surgical removal or at least excisional biopsy.

The patient was placed under general endotracheal anesthesia and positioned prone on a Wilson frame. A standard posterior midline approach was performed, the subcutaneous layers split and the thoracolumbar fascia divided. The tumor was clearly visualized and was distinct from its surrounding. An intracapsular resection of the encapsulated tumor was performed en bloc. No distinct nerve or fascicle from which the tumor arose could be identified.

Intraoperative histopathological frozen-section analysis indicated a low-grade spindle-cell neoplasm that favored the diagnosis of Schwannoma.

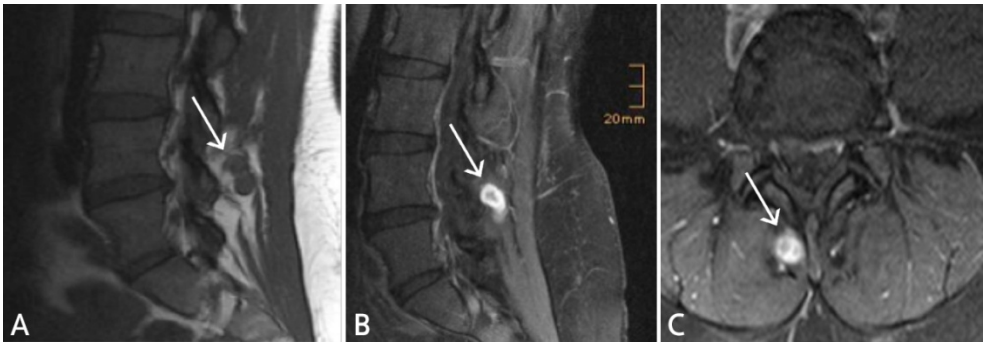

Fig. 1. Lumbar MRI: sagittal T1 precontrast (A), sagittal T1 post contrast (B) and axial T1 post contrast images (C) round and well demarcated mass is found at the level of L4 just lateral to the spinous process.

On gross examination the specimen showed a yellow, well circumscribed ovoid soft mass, approximately 1x1x0.9 cm in size. The cross cut surfaces displayed regions with nodular appearance. Margins were clear. Neuropathological stainings were positive for S100 and Vimentin. Pathological examination revealed the characteristic features of alternating Antoni A and Antoni B areas. While Antoni A areas are composed of spindle shaped Schwann cells arranged in interlacing fascicles showing nuclear palisading, Antoni B area consist of a loose meshwork of gelatinous and microcystic tissue. Based on the histopathological features and classic immunostainings, the diagnosis of Schwannoma was made. After an unremarkable postoperative course, the patient was discharged on the second postoperative day.

Further workup such as genetic analysis of the identified INI1 gene as well as MRI scan of the entire neuroaxis was not needed as schwannomatosis without NF syndrome is an inherited autosomal dominant disease and diagnosed by 2 or more histologically confirmed peripheral Schwannomas.

As the risk of recurrence is estimated to be less than 10% in these cases with gross total resections [11], no further therapeutic treatment was indicated. Short-term follow up has shown a gratifying outcome with significant improvement of pain and the patient has meanwhile returned to normal daily life activities.

Due to the unexpected diagnosis of this lesion as an extraspinal lumbar Schwannoma originating either from the medial or lateral part of the posterior branch of the corresponding spinal nerve (L3 or L4), a more frequent follow-up surveillance schedule was established, which thus far has not shown clinical nor radiological signs of recurrence at 6 months post surgery.

3. DISCUSSION

Most spinal Schwannomas present as firm encapsulated tumors of a nerve fascicle, initially causing radicular pain followed by weakness eventually leading to myelopathy [12].

These tumors may occur at any level of the spinal axis, but are mostly located intradurally [13]. They may extend along the spinal nerve and impose as a dumbbell mass, consisting of an intradural and extradural component [12]. However, to the best of our knowledge, no cases of purely extraspinal Schwannoma of the paraspinal region have been reported

previously.

These sporadic occurring tumors usually affect adults at about 40-50 years of age [13,14], with a male predominance. The tumor is usually well- circumscribed and the average size at the time of diagnosis varies greatly [15].

Characteristic MR-imaging features include rather homogenous isointense signal intensity on T1 weighted MR sequences with heterogeneous contrast enhancement and high signal intensity on T2 weighted MR sequences. The majority of lesions have well-defined margins.

On histological examination, two distinct patterns of cellular architecture known as Antoni A and Antoni B regions can be recognized. Although not highly specific, S-100 protein is a reliable marker being especially prevalent in the Antoni A areas [16,17].

Due to her blank family history and clinical presentation with complete lack of signs indicating neurofibromatosis such as café au lait spots or cutaneous fibromas, neuroradiological diagnostic studies (MRI) were limited to the lumbar spine. An ophthalmological examination to exclude Lisch bodies is scheduled postoperatively.

Based on imaging criteria alone, the differential diagnosis includes: Neurofibroma, Schwannoma, Paraganglioma, palisaded encapsulated Neuroma, Myxoma, Fibroma, Liposarcoma and metastatic disease besides other rare entities. As all Neurofibromas and Schwannomas show moderate to strong expression of S-100 protein, its absence should lead to further investigations to ensure the diagnosis.

4. CONCLUSION

Although intradural-extramedullary Schwannomas are frequently encountered in their practice, neurosurgeons should also be aware of a purely paraspinal location. As even "extraspinal" Schwannomas may cause debilitating effects, total microsurgical resection may become a necessity. As seen in our case complete surgical excision can be considered curative, reliefs the patient from their symptoms and should hence be seen as the treatment of choice.

CONSENT

All authors declare that written informed consent was obtained from the patient publication of this case report and accompanying images.

ETHICAL APPROVAL

Not applicable.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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