Genuine neoplasms arising from the filum terminale are uncommon with ependymal tumors, particularly myxopapillary ependymoma as the most common, followed by paraganglioma. Other genuine neoplasms (such as teratomas) and tumor-like conditions (such as lipomas and malformations) can also affect the sacrum, with involvement of the filum terminale. Microscopic clusters of ganglionic cells have been described in normal human filum terminale. Heterotopic dorsal root ganglion cells and ganglionic tissue have been described in patients with spinal malformations including spina bifida and diastematomyelia. We report a centimeter-sized, heterotopic ganglion in the filum terminale that was not associated with malformation of the spinal cord or vertebral column. This lesion mimics a neoplasm on imaging, and on clinical presentation it was initially thought to account for the patient’s subjective symptoms. To our knowledge, no instance of ectopic or heterotopic ganglia within the cauda equina, in the absence of preexisting neural tube pathology, has been described in the English-language scientific literature.

Ectopic ganglion in cauda equina: case report

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Macroscopic ectopic or heterotopic ganglionic tissue within the cauda equina is a very rare pathological finding and is usually associated with spinal dysraphism. However, it may mimic genuine neoplasms of the cauda equina. The authors describe a 29-year-old woman with a history of back pain, right leg pain, and urinary incontinence in whom imaging demonstrated an enhancing mass located in the cauda equina at the L1–2 interspace. The patient subsequently underwent biopsy and was found to have a focus of ectopic ganglionic tissue that was 1.3 cm in greatest dimension.

To the authors’ knowledge, ectopic or heterotopic ganglionic tissue within the cauda equina in a patient without evidence of spinal dysraphism has never been reported. This patient presented with imaging and clinical findings suggestive of a neoplasm, and an open biopsy proved the lesion to be ectopic ganglionic tissue. The authors suggest that ectopic ganglionic tissue be added to the list of differential diagnoses of a space-occupying lesion arising from the cauda equina.

http://thejns.org/doi/abs/10.3171/2015.11.SPINE141304

KEY WORDS ectopic ganglion; cauda equina tumors; spinal oncology; ectopic dorsal root ganglion; paraganglioma; lumbar

Case Report

History and Examination

The patient was a 29-year-old woman who presented with a long history of back pain, several months of intermittent incontinence, and new-onset pain and numbness in her right leg. On physical examination the patient appeared healthy and in no acute distress. No constitutional conditions were noted. Her neurological examination revealed normal strength and sensation, and examination of her spine demonstrated no stigmata of congenital anomalies. Urological evaluation, including formal urodynamic studies, revealed neurogenic bladder.

Imaging Findings

An MRI study of the lumbar spine demonstrated a small nodule, 1.3 × 0.5 × 0.7 cm, located at the cauda equina at the L1–2 level. The lesion was well circumscribed, isointense to the spinal cord on T1-weighted images, and showed focal enhancement (Fig. 1A and B). The T2-weighted images demonstrated a hypointense lesion that...
T2-weighted imaging.
weighted imaging with Gd enhancement.
arrow
well-demarcated, enhancing nodule (Fig. 1.

- pain had resolved, whereas her numbness remained un-
1-month follow-up, the right lower-extremity neuropathic
home in good condition after a 2-day hospital stay. At
experienced transient neuropathic pain that was successfully
treated with steroids and gabapentin. She was discharged
patienteoed neuropathic pain to be absent. At last follow-up,
the patient continued to demonstrate a neurogenic bladder.
Her neurological examination was otherwise unchanged.

Pathological Findings
Intraoperative cytological preparations demonstrated a
high number of huge, round cells with centrally located
large round nuclei and large nucleoli that were diagnostic
of normal-appearing ganglion cells (Fig. 2A). These
cells were admixed with small spheroids of tightly packed
cells that had small nuclei without nuclear atypia. No men-
ingeal or glial components were noted. Histological sections
showed classic features of a ganglion that included
ganglionic neurons rimmed by sustentacular cells and
embedded in a background of Schwann cells with susten-
tacular cells (Fig. 2B and C). On immunohistochemistry
(Fig. 2D–F), the Schwann cells but not the ganglionic cells
were positive for S100 protein (Polyclonal, Ventana). The
ganglionic cells were positive for synaptophysin (Clone
SP11, Ventana) and neurofilament proteins (Clone FNP7,
Invitrogen). Note that peripheral nerve bundles were
highlighted by immunohistochemistry for neurofilament proteins (arrow in Fig. 2F). Immunohistochemistry was
performed by a Benchmark Autostainer (Ventana) with
protocols provided by the vendor.

Discussion
The histology and immunohistochemical profile in this
case was that of a peripheral nerve ganglion. The electrophysiological observation that this nodule was closely
associated with functional nerve fibers, the surgical ob-
servation that nerve fibers coursed around the nodule, and
the fact this nodule could not be cleanly dissected from
other fibers confirmed that this was an ectopic ganglion
and probably electrically active. Our report is the first de-
scription in the English-language literature of an ectopic
ganglion that presented as a tumor of the filum terminale.

Microscopic clusters of ganglionic cells have been de-
scribed in normal human filum terminale. Heterotopic
dorsal root ganglion cells are found in 66.3% of cases of
spina bifida aperta, according to 1 study. Small ectopic
ganglionic tissue intercalated in the ventral root of 1 hemi-
cord has been described in an asymptomatic 63-year-old
patient with sacral diastematomyelia, and paramedian
dorsal root ganglia have been described in an 18-month-
old infant with a split cord malformation. The etiology of
these rare findings is thought to be associated with abnor-
mal migration of neural crest cells during primary neu-
rogenesis—more specifically, secondary to a premature
division of the neural plate prior to closure of the neural
tube.
The nodule in this case was 1.3 cm in greatest dimension and was far larger than any of the aforementioned ectopic ganglionic tissue nodules associated with spinal and/or vertebral malformations. This unique case illustrates that ectopic or heterotopic ganglionic tissue can occur in the absence of underlying spinal dysraphism. Radiographically, the current lesion raises a concern of paraganglioma. Clinical findings (midline location, urodynamic studies, and radiculopathy) seem to be of no value in differentiating this congenital lesion from an acquired lesion in the filum terminale.

Space-occupying lesions arising from the filum terminale are essentially intradural extramedullary lesions. Spinal intradural extramedullary tumors are uncommon, and the more commonly encountered entities include schwannoma, meningioma, neurofibroma, myxopapillary ependymoma, paraganglioma, and leptomeningeal metastasis. Among these entities, myxopapillary ependymoma and paraganglioma typically arise from the cauda equina and/or filum terminale. Recognition of myxopapillary ependymoma radiographically, grossly at surgery, or through pathological examination is usually not a major challenge. Paraganglioma is unique in that it often presents as a single round nodule hanging by the filum terminale. In most situations, recognition of this entity is not difficult. Paraganglioma, however, can have gangliocytic differentiation. Nearly half of cauda equina paragangliomas contain mature ganglion cells and cells that are transitional between chief and ganglion cells. These tumors are often referred to as gangliocytic paraganglioma. Interestingly, gangliocytic paraganglioma are frequently seen in the duodenum, and a concomitant ganglioneuroma component has also been described. Whereas classic paraganglioma can be easily distinguished from ectopic ganglion tissue, gangliocytic paraganglioma and paraganglioma with a ganglioneuromatous component have certain histological overlap with ectopic ganglionic tissue. Demonstration of areas with the classic paraganglioma component is the best way to differentiate these tumors from ectopic ganglionic tissue.

Conclusions

Although this patient’s lesion did not represent a neoplasm, its appearance on imaging and the clinical presentation warranted biopsy. The patient’s preoperative neuropathic pain resolved after biopsy of the lesion. Although the lesion resembled neural tissue on intraoperative inspection, pathological identification of spinal mass lesions remains dependent on obtaining tissue. The decision whether to perform a biopsy is dependent on the localizing quality and severity of the patient’s symptoms as well as radiographic and intraoperative findings. At this point, the patient’s pain is being successfully managed with neuropathic pain medications. Given her limited but present urinary and bowel function, complete resection of this lesion has not been pursued.

References


Disclosures
The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author Contributions
Conception and design: Martin, Conner, Fung. Acquisition of data: Conner, Peterson. Analysis and interpretation of data: Conner, Fung, Peterson. Drafting the article: Martin, Conner, Peterson, Glenn. Critically revising the article: Martin, Conner, Glenn. Reviewed submitted version of manuscript: Conner, Fung, Glenn. Statistical analysis: Fung. Administrative/technical/material support: Conner, Fung, Peterson. Study supervision: Martin, Fung.

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