Gangliocytic paraganglioma in cauda equina region, with biochemical and neuropathological studies

Case report

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Biochemical analysis of a nonfunctional paraganglioma in the cauda equina region demonstrates that its catecholamine content is predominantly dopamine with small amounts of noradrenaline and adrenaline. Scattered mature large neurons characterize the tumor as gangliocytic paraganglioma. Ultrastructural study shows intracytoplasmic neurosecretory granules in the neoplastic cells.

KEY WORDS • nonfunctional paraganglioma • cauda equina • dopamine • gangliocyte • neurosecretory granules • spinal tumor

PARAGANGLIOMA in the region of the cauda equina has been reported several times since the initial report by Lerman, et al., in 1972. None of the cases described was associated with a clinical hormonal syndrome. Biochemical analysis of the tumor in this location has not been reported previously. We are reporting the catecholamine content of a nonfunctional paraganglioma removed from the cauda equina region. Light and electron microscopic study of the tumor showed the presence of neurons, a characteristic feature of the gangliocytic variant of paraganglioma.

Case Report

This 42-year-old man was admitted to Montefiore Hospital and Medical Center with low-back pain of 4 years’ duration precipitated by a work-related injury. He had experienced intermittent tingling sensations in both lower extremities for 2 years, and a feeling of weakness in the right leg for 3 weeks.

Examination. General physical examination and blood pressure were normal. There was tenderness over the lumbar spine, paravertebral muscular spasm, and bilateral Lasègue signs. There was no muscular atrophy or weakness, although the right patellar reflex was depressed. There was hypalgesia of the entire right lower extremity and the left saddle region.

Lumbar spine x-ray films were normal. Myelography with metrizamide revealed a complete block at L-1, consistent with an intradural extramedullary tumor. Cerebrospinal fluid protein was 4576 mg/dl.

Operation. A 3.5 × 2 × 1.5-cm intradural encapsulated tumor was removed intact through a thoraco-lumbar laminectomy, using microsurgical technique. The tumor was adherent to the roots of the cauda equina. It was not attached to the conus medullaris or filum terminale. The postoperative course was uneventful, and the patient remained well at follow-up examination 1 year later.

Pathological Examination. A 0.597-gm portion of the tumor was excised from the intact specimen, rinsed in normal saline, and immediately placed and stored in deep freeze. The specimen was transported in dry ice to the Upjohn Esoteric Center in Kalamazoo, Michigan, where a radioenzymatic assay of the catecholamine content of the tumor was performed following the basic principles of Passon and Peuler, with modifications. The analysis revealed the following values: dopamine 306 ng/gm; epinephrine 6.9 ng/gm; and norepinephrine 37.2 ng/gm.

Histologically, the tumor had irregular islands and lobules of uniform polyhedral cells in a vascular stroma. In one area, large mature neurons were dispersed among the tumor cells (Fig. 1). At the ultra-
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structural level, most of the tumor cells, as well as the neurons, contained numerous dense-core vesicles consistent with neurosecretory granules (Fig. 2). The granules were intimately associated with fibrils in some tumor cells (Fig. 3). The endothelial cells of some blood vessels were fenestrated.

Discussion

In common with other neuroendocrine tumors, paraganglioma is characterized ultrastructurally by numerous clusters of intracytoplasmic dense-core vesicles consistent with neurosecretory granules. The usual biochemical correlate of this morphological feature is the capability of these cells for amine precursor uptake and decarboxylation (APUD),\textsuperscript{13} This phenomenon is reflected in the catecholamine content of the neoplasm. Functional tumors usually contain noradrenaline predominantly, while nonfunctional tumors may contain variable amounts of noradrenaline, dopamine, and adrenaline.\textsuperscript{9} The tumor in our case contained dopamine principally, and had small amounts of noradrenaline and adrenaline. The human carotid body has been found to have similar proportions of catecholamines.\textsuperscript{17} The lack of clinical hormonal syndrome in tumors with documented catecholamine content may be due to an inherent failure of the tumor cells to secrete the stored substances or

![FIG. 1. Photomicrograph of the tumor specimen showing large neurons. H & E, \( \times 100 \).](image1)

![FIG. 2. Electron micrographs of the tumor cell cytoplasm. Left: Dense-core vesicles surround a blood vessel with fenestrated endothelium. \( \times 7500 \). Right: Neurons are seen with abundant dense-core vesicles. \( \times 4200 \).](image2)
an inability of the secreted products to evoke a clinically evident response. 4

The pattern of interspersed neurons within clusters of paraganglioma cells in our case is similar to that of the gangliocytic paraganglioma in the duodenum, initially described in 1971 by Kepes and Zacharias. 7

The presence of ganglionic elements in paragangliomas of the carotid body and glomus jugulare has also been suggested previously. TM This feature is most likely just a reflection of normal structure, since rare ganglion cells have been observed in normal paraganglia. 1

Paraganglioma has also been described to form a variable admixture of patterns with other neuroendocrine tumors such as ganglioneuroma in different locations 7,15 including the cauda equina region. 10 The histopathological spectrum of paraganglioma in the latter location will be better defined as more cases are documented.

References


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