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Neuroendocrine Tumor (Paraganglioma) of the Cauda Equina: MR and Pathologic Findings

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Summary: The MR and pathologic features of a case with neuroendocrine tumor (paraganglioma) of the cauda equina are presented. MR showed the tumor to be hyperintense on postcontrast examination and also showed serpiginous flow voids suggesting vessels capping the tumor. A neuroendocrine tumor should be considered in the differential diagnosis of tumors in this location.

Index terms: Cauda equina; Spinal cord, neoplasms; Spinal cord, magnetic resonance

Neuroendocrine tumors (NETs) are so called because they are composed of cells that have neural features of peptide-containing secretory granules and potential endocrine function. This term has largely replaced the term apudoma which is derived from the concept, proposed by Pearse (1), of a widely dispersed system of cells displaying amine precursor uptake and decarboxylation. These cells are located in a wide variety of sites, including the adrenohypophysis, thyroid gland, pancreatic islets, gastrointestinal tract, tracheobronchial tree, adrenal medulla, skin, and chemoreceptor system, among others. Many but not all of the NETs arising from these cells produce symptoms caused by hypersecretion of peptide hormones.

We report here the magnetic resonance (MR) findings in a patient with a pathologically proved NET (paraganglioma) of the cauda equina.

Case Report

A 44-year-old woman presented with a 1½-year history of low back pain radiating into her right leg and foot. A computed tomography (CT) scan done at another hospital 1 year previously was reported as showing mild "degenerative changes" in the lumbar spine but was otherwise normal. Treatment by a chiropractor and at the pain clinic at another hospital afforded little relief. The patient's pain continued to progress and a repeat CT scan at another hospital a few weeks before admission was reportedly unremarkable. An attempt at a myelogram resulted in a bloody tap.

At presentation to our hospital, she stated that there had been increasing pain with inability to dorsiflex her right foot and toes for the past few weeks. She denied any bladder or bowel symptoms. On examination, there was slight right leg weakness. The right knee jerk was absent, and the plantar reflexes were flexor bilaterally. There was decreased pin prick sensation in the right L4-L5 distribution and decreased rectal tone.

An unenhanced axial CT scan of the lumbar spine obtained at our institution was interpreted as normal. Myelography was attempted but yielded only a bloody tap. An MR examination of the lumbosacral spine with and without Gd-DTPA (gadolinium diethylene triamine pentacetae) was then obtained. This revealed an intradural mass extending from below the level of the conus medullaris down to the S1 level (Fig. 1).

Surgical exploration revealed a sausage-shaped, soft, whitish, encapsulated tumor with a cyst in its caudal portion originating from the filum terminale. It had a rich vascularity that was most marked superiorly. A gross total removal of the tumor was achieved. A follow-up MR examination has shown recurrence of a small tumor nodule 2 years and 3 months later (Fig. 2) but, so far, without the return of neurologic symptoms.

Pathology

The excised tumor was composed of epithelial cells with a predominantly papillary pattern (Fig. 3A). The papillae had fibrovascular cores covered by crowded tumor cells having small round nuclei with moderately dispersed chromatin and occasional nucleoli and either vesicular or pale slightly granular cytoplasm. No mitotic figures were present.

Immunocytochemistry showed strong positive staining for neuron-specific enolase and chromogranin, neuroen-
Fig. 1. A, Nonenhanced sagittal T1-weighted spin-echo image (600/20/4, TR/TE/excitations) reveals a large intradural tumor (arrows) that is almost isoïntense to the spinal cord except for a caudal hypointense area.

B, Proton density- (2400/35/2) and C, T2-weighted spin-echo images (2400/70/2) reveal bulk of the tumor (arrows) to be isoïntense to the spinal cord, but the superior and inferior poles are markedly hyperintense. The fluid collection in the soft tissues posteriorly is related to a recent attempt at a myelogram.

D, Sagittal T1-weighted spin-echo image (600/20/4) 8 minutes after intravenous injection of Gd-DTPA shows heterogeneous but intense enhancement of the tumor (arrows). The superior pole enhances most intensely, whereas the caudal cyst does not enhance. Note the flow void within the prominent vessels related to the superior pole of the tumor (best seen in A, B, and C).
Russell and Rubinstein (3) have continued to use the term paraganglioma for all NETs of the cauda equina regardless of their histologic variations or immunocytochemical profile. In fact, the tumor in our patient had certain histologic and immunocytochemical features (absence of S-100 protein and the presence of α-subunit as well as the presence of keratin and the histologic papillary pattern) which may be thought to favor a diagnosis of carcinoid tumor rather than paraganglioma. Nonetheless, most of the pathologic features of the tumor in our patient have been documented in tumors reported as paragangliomas. In addition, the clinical presentation and operative findings were typical of the NET reported to date as paraganglioma. The patient was not known to have a primary NET at any other

Discussion

NETs from a wide variety of sites have been given distinctive names including Merkle cell tumor (skin), oat cell carcinoma (lung), islet cell tumor (pancreas), medullary carcinoma (thyroid), paraganglioma (chemoreceptor system), and carcinoid (gastrointestinal and respiratory tracts). Occasionally, NETs are found at sites where the cell of origin is obscure. The cauda equina is one such site and the NETs reported at this site have been identified as paragangliomas except for a single report of a metastatic carcinoid tumor (2).
The hypointense caudal cyst which was marked by T1-weighted spin-echo images, except for the partial or complete block—a nonspecific finding of a case with NET (paraganglioma) of the cauda equina. MR showed postcontrast enhancement to be diagnostic—particularly useful than unenhanced T2-weighted images. The enhanced scan was particularly useful in accurately delineating the superior and inferior extent of the tumor. In summary, we have presented the MR findings of a case with NET (paraganglioma) of the cauda equina. MR showed postcontrast enhancement of the tumor and also showed its serpiginous flow voids. The latter were felt to be caused by either hypervascularity of the tumor or compression of veins by the large mass resulting in their dilatation. A (paraganglioma or other) NET, albeit rare, should be considered in the differential diagnosis of an intradural tumor in this location.

References
1. Pearse AGE. The cytochemistry and ultrastructure of polypeptide hormone-producing cells of the APUD series and the embryologic.
physiologic and pathologic implications of the concept. J Histochem Cytochem 1969;17:303–313