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Case Report

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Summary: We report a case of a ganglioneuroma that was found incidentally on a CT of the abdomen in a 27-year-old woman with gastrointestinal symptoms. The tumor, though located in the posterior mediastinum, demonstrated an unusual feature of abundant adipose tissue on MR imaging. The mass was later resected, and adipose tissue intermixed within a ganglioneuroma was identified microscopically. Adipose tissue is a rare finding in a ganglioneuroma, but this radiographic feature may be useful in the evaluation of posterior mediastinal masses.

Ganglioneuroma is one of several tumors reported to occur in the posterior mediastinum (1–5). The list includes Schwannoma, neurofibroma, ganglioneuroblastoma, neuroblastoma (1–3), malignant peripheral nerve sheath tumor (1, 2), lymphoma (1, 3), primitive neuroectodermal tumor (2), pheochromocytoma (1–3), aortic aneurysm (2), prostate (2), skin (2), retroperitoneum (2), and peripheral autonomic system (2). These tumors are grossly well-circumscribed, solid, encapsulated masses (4, 5, 11, 12) and may demonstrate calcifications (4, 10) or a whorled pattern on cut section (5). The hallmark microscopic feature of ganglioneuromas is the presence of mature ganglion cells and the absence of immature neuroblasts. These tumors occasionally are associated with active hormone secretion, including vasoactive intestinal polypeptide, catecholamines, and testosterone (4, 5).

Ganglioneuromas are one of a few tumors that arise from neural crest cells. This category of neoplasms also includes peripheral nerve sheath tumors, melanomas, and neuroendocrine cell tumors (2). More specifically, ganglioneuromas arise from the autonomic ganglion cells of the peripheral nervous system, usually the sympathetic ganglion. They represent the benign end of the spectrum for the ganglion cell lineage, whereas ganglioneuroblastosomas and neuroblastomas comprise the malignant end of this spectrum. It is not surprising that ganglioneuromas may arise anywhere along the peripheral autonomic ganglion sites. They most commonly occur within the posterior mediastinum and retroperitoneum and less commonly in the adrenal medulla, parapharyngeal region, visceral ganglia, or cranial nerve ganglia (4, 5). They have been reported rarely in the tongue, mandible, bladder, uterus, ovary, spermatic cord, testes, prostate, and skin and bone (5). These tumors are circumscribed, solid, encapsulated masses (4, 5, 11, 12) and may demonstrate calcifications (4, 10) or a whorled pattern on cut section (5). The hallmark microscopic feature of ganglioneuromas is the presence of mature ganglion cells and the absence of immature neuroblasts. The ganglion cells can be identified by their abundant

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eosinophilic cytoplasm, large nuclei, and prominent nucleoli. Other elements are usually seen and include Schwann cells, perineural satellite cells, axons, and collagen fibers (4, 5). Varying degrees of myxoid stroma and an interlacing pattern of Schwann cells and collagen fibers have also been reported (5, 11).

These latter 2 histopathologic features seem to correlate well with imaging findings (11). Fat, which was found in our case, is an uncommon finding but has been reported elsewhere (10, 11).

Ganglioneuromas appear radiologically as well-defined, oblong masses that are located anterior and

Fig 1. A–C, Sagittal T1-weighted images (TR 366/TE 14), from right to left, showing a right paraspinal mass with predominant high T1 signal intensity and interspersed areas of intermediate signal intensity. The mass extends through the intervertebral foramina, causing widening of the foramina.

D, Axial T1-weighted image (TR 366/TE 14) showing a right paraspinal mass that extends through the intervertebral foramina causing widening of the foramina and scalloping of the posterior margin of the vertebral body.

E, Axial T2-weighted fast spin-echo image (TR 3400/TE 110) showing mild effacement of the right lateral aspect of the thecal sac without any significant compression.
lateral to the spine. They typically span 3–5 vertebral levels (1). On CT imaging, these tumors may be homogeneous or heterogeneous masses with low to intermediate attenuation (13). Calcification has been reported in approximately 20% of cases (5, 13) and is usually punctate as opposed to the coarse pattern seen with ganglioneuroblastomas and neuroblastomas (1, 13). Following contrast administration, the tumors demonstrate mild to moderate enhancement (13).

On MR imaging, ganglioneuromas appear as homogeneous masses with low and, less commonly, intermediate signal intensity on T1-weighted images (11, 13). In contrast, on T2-weighted images, the signal intensity is usually heterogeneous and either intermediate to high or markedly high (11–13). In a series of 10 patients published by Zhang et al (11), the microscopic presence of abundant myxoid stroma was found to correlate with very high signal intensity on T2-weighted images. Also reported in this study and other sources is the finding of a whorled appearance on T1-weighted and, more commonly, T2-weighted images within the tumor that corresponds to the microscopic interlacing patterns of Schwann cells and collagen fibers (5, 11). There may be no, mild, or heterogeneous enhancement (11, 13). Ring enhancement may correspond to the capsule of the tumor (11).

Our case was unusual because the tumor demonstrated areas of high signal intensity on both T1- and T2-weighted images. Furthermore, there was low signal intensity of the mass on the fat-suppressed postcontrast images. These findings suggested the presence of a fat-containing tumor before the resection of the mass. Microscopically, the presence of adipose tissue was confirmed along with the typical features of a ganglioneuroma.

The list of fat-containing tumors found in the posterior mediastinum is shorter than the list of posterior mediastinal tumors given in the introduction. In our review of the English literature, neoplasms that have been reported within the posterior mediastinum and characterized by adipose tissue are teratomas (8, 9), parathyroid lipoadenoma (14), and other lipomatous mesenchymal tumors, including lipomas, liposarcomas (3), myelolipoma (15), and angiolipoma (16). Schwannomas, which have a similar appearance to ganglioneuromas on MR imaging, including low signal intensity on T1-weighted images and high signal intensity on T2-weighted images (1), have also been
reported to contain fat (17). Other non-neoplastic masses in the posterior mediastinum with adipose tissue elements include fatty transformation of extramedullary hematopoiesis and panniculitis (10).

Ganglioneuromas (1, 5) and Schwannomas (1) are known to widen the neural foramina and extend into the spinal canal. Lipoblastomas (18) and angiolipomas (16) have also been reported to enter the spinal canal. Therefore, we conclude that the presence of fat in a posterior mediastinal mass that widens the neural foramina and enters the spinal canal narrows the differential diagnosis to a gangli-

Fig 2. A, Microscopic picture demonstrating adipose tissue (short arrow), Schwann cells, lymphocytes (arrowhead), and ganglion cells (long arrow); original magnification 10×.

B, On the left, S100 immunostain demonstrates that the tissue is composed predominantly of Schwann cells. On the right, Synaptophysin immunostain confirms the presence of ganglion cells (arrows); original magnification 20×.

C, High-power picture of multinucleated ganglion cells (arrows); original magnification 40×.
oneuroma, Schwannoma, or lipomatous mesenchymal tumor.

References