A 45-year-old man was diagnosed with retroperitoneal ganglioneuroma. The tumor was a large encapsulated mass of firm consistency with a homogeneous, solid, grayish white-cut surface, having a focally edematous appearance (Figures, Top). Microscopically, the overall appearance of ganglioneuroma resembled that of a neurofibroma (it is designated as a schwannian stroma-dominant tumor) except for the presence of numerous collections of abnormal but fully mature ganglion cells, often with more than one nucleus (Figures, Bottom).

Neuroblastic tumors constitute 4 basic morphologic categories: neuroblastoma; ganglioneuroblastoma, intermixed; ganglioneuroma; and ganglioneuroblastoma, nodular. Ganglioneuromas are fully differentiated and invariably benign, mostly seen in the elderly. They can be multiple and/or associated with other independent types of neural/neuroendocrine neoplasms. Their most common location is the posterior mediastinum and the retroperitoneum. Although catecholamine synthesis is an almost constant feature of all the neurogenic tumors, ganglioneuromas rarely lead to symptoms. Thorough microscopic examination is crucial; areas of different colors or consistency are particularly suspicious for harboring less differentiated foci, namely ganglioneuroblastoma. If there is a minor component of scattered collections of differentiating neuroblasts and/or maturing ganglion cells, the tumor is named maturing subtype, which blends with the intermixed subtype of ganglioneuroblastoma, from which it differs in that the immature foci do not form distinct microscopic nests. Exceptionally, the Schwann cell component of the ganglioneuroma may show features of a malignant nerve sheath tumor.

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