

Ganglioneuroma

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Definition Ganglioneuromas (GNs) are benign differentiated tumours of neural crest-derived cells in the autonomic nerves. Incidence GNs are rare, affecting one per million of the population. Most are sporadic but they can be associated with neurofibromatosis type II and MEN 2B.

Pathology GNs are benign and are most often located in the mediastinum (20%), retroperitoneum (10%) and adrenal gland (30%). However, GNs can arise anywhere sympathetic nervous tissue is found, such as tongue, bladder, uterus, bone and skin. They are composed of an admixture of ganglion cells and Schwannian stroma/cells.

Clinical presentation GNs develop in childhood but typically present later as they are non-secreting and slow growing. Two-thirds of patients are under the age of 20 years. They are usually asymptomatic

Theodore Schwann , 1810–1882, Professor of Anatomy , Leuven, Belgium. Michel Gagner , b. 1960, Professor of Surgery , Montreal, Canada. Martin K Walz , contemporary , Professor of Surgery , Essen, Germany . and are identified incidentally . When large, GNs may cause local pressure symptoms such as abdominal pain or bloating. Occasional reports of elevated metanephrines exist and the differential diagnosis is PCC.

Diagnosis Once a mass is discovered CT and MRI are usually performed to characterise the lesion. They are well defined with a capsule and calcification may be present. GN has a low T1- and a high T2-weighted signal on MRI. Radiology cannot definitively diagnose a GN; histology is required and so preoperative diagnosis can be challenging.

Treatment Excision is the treatment of choice. A laparoscopic approach is preferred. The prognosis is excellent as recurrences are rare.

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