

## Case Report

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# Laparoscopic surgery for adrenal ganglioneuroma: Case report and literature review

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## Introduction

Ganglioneuromas (GN) are benign and well-differentiated tumors originated from the sympathetic chain of the neural crest. They can affect any part of the body along the autonomic ganglia, however the two most common sites of presentation are in the retroperitoneal space (32-52%) and posterior mediastinum (39-43%), followed by the cervical region (8-9%) [6]. They are most common in children and young adults, but can occur at any age. They have a greater predilection for females (3:1). They are usually asymptomatic and when present, clinical manifestations are symptoms of compression (epigastric pain, abdominal pain). Most adrenal ganglioneuromas do not have endocrine production, but up to 30% of tumors can secrete testosterone or catecholamines.

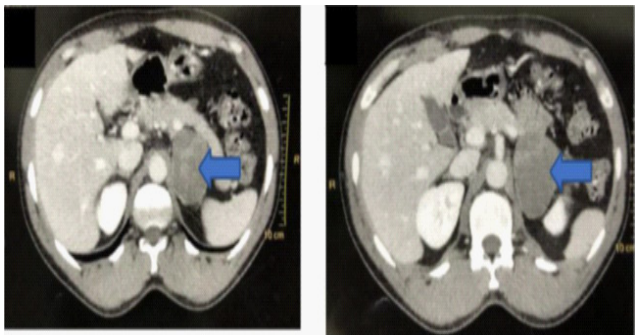
## Case report

Male patient, 39 years old, without comorbidities, referred to Hospital Santo Antônio – Obras Sociais Irmã Dulce due to a nodular image shown in routine exams. The patient was asymptomatic in a specialized medical appointment. Abdominal ultrasonography showed an expansive formation in the topography of the left adrenal, measuring 5.5 cm in the longest axis. A computed tomography scan of the abdomen showed solid expansive formation and heterogeneous post-contrast highlight in the left adrenal, with medium density of 26 UH and measuring about 6.2 X 5.8 cm in its longest axes (Figures 1 and 2). In requested metabolic assessment, all results were unchanged. The patient underwent videolaparoscopic left adrenalectomy (Figures 3 and 4). The patient evolved without complications

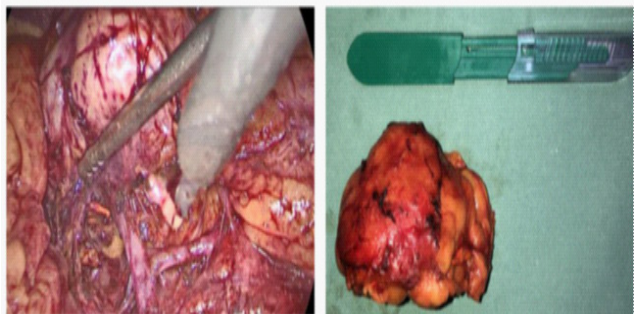
and was discharged on the first postoperative day. Pathological anatomy results revealed a mature subtype of ganglioneuroma, with ganglion cell and Schwann cell components (Figures 5 and 6). Size of neoplasm: 8.0 X 5.5 X 4.0 cm. Neoplastic components: ganglion cells and Schwann cells. Characteristics of the ganglionic component: mature with compact and eosinophilic cytoplasm with distinct cell borders, single eccentric nucleus, prominent nucleolus, varying in distribution and number, with sparse distribution. Characteristic of the Schwann cell component: they support neuritic processes without the presence of myxoid component.

### Discussion

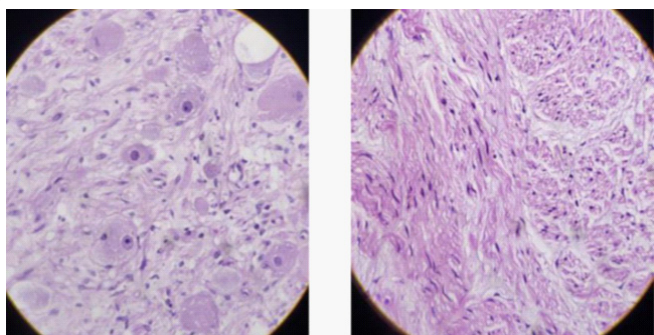
Adrenal ganglioneuroma is a rare, nonfunctioning, benign tumor of the adrenal gland that develops slowly. Although in rare cases there is metabolic production of testosterone and catecholamine, the vast majority presents as an adrenal inci-



**Figure 1 and 2:** Abdominal Tomography – blue arrows show the lesion.



**Figure 3 and 4:** Laparoscopy images and surgical specimen.



**Figure 5 and 6:** Histopathology images.

dentaloma on imaging exams [7]. Symptoms are only present if the tumor is large enough to cause a mass effect on adjacent structures or if it exhibits hormonal activity [4]. The test used to confirm the existence of adrenal incidentaloma is abdominal CT scan. Adrenal incidentalomas are rare findings on abdominal CT (0.6 to 1.3% of all abdominal CT scans performed). Adrenal ganglioneuroma represents only 0-6% of all adrenal incidentalomas [7].

Differential diagnosis includes: ganglioneuroblastoma, neuroblastoma, pheochromocytoma, adrenal adenoma, and adrenal carcinoma [6]. Diagnostic confirmation requires histopathological analysis which reveals a neoplasm formed by a mixture of ganglion cells and Schwann cells. Ganglion cells may exhibit a mature to slightly dysmorphic component. The mature component exhibits eosinophilic compact cytoplasm cells with distinct cell borders, single eccentric nucleus, and prominent nucleolus. And the dysmorphic component presents cells with single or multiple pyknotic nuclei, varying in distribution and number, which are usually quite sparse. May contain finely grained, golden to brown pigment (lipofuscin or neuromelanin). The Schwann cell component may involve neuritic processes and get organized into small, intersecting fascicles that are separated by a loose myxoid stroma. Ganglioneuroma has two histological subtypes: mature (most common, where all ganglion cells are mature) and maturing (where there is a smaller component of scattered collections of differentiating neuroblasts or maturing ganglion cells). However, unlike the mixed ganglioneuroblastoma subtype, these immature focuses do not form clear microscopic nests. A slight variation in cellularity is allowed for these diagnoses, however, no significant atypia, mitosis or necrosis should be present. Most cases are of sporadic origin, but some rare cases have been reported in association with Turner Syndrome and Multiple Endocrine Neoplasia Type II [2].

The definitive treatment for adrenal ganglioneuroma is surgical resection, open or minimally invasive (laparoscopic, robotic). Adrenal tumor larger than 6 cm is a relative contraindication for a laparoscopic approach [8]. It should only be performed in tumors larger than 6 cm by an experienced team and respecting all the oncological principles of resection. In our case, despite the tumor being larger than 6 cm, the laparoscopic approach was chosen, without violating the surgical principles of adrenal surgery, and without intraoperative complications.

### Conclusion

Adrenal ganglioneuroma is a rare tumor, usually benign, composed of mature ganglion cells and Schwann cells. It usually does not cause symptoms until it reaches a sufficient volume to cause compressive symptoms. In most cases the diagnosis is through an incidentaloma in imaging exams. We report the case of a 39-year-old young patient with an incidental finding of a left adrenal lesion measuring 6.2 cm in the longest axis, who underwent videolaparoscopic adrenalectomy without complications. The prognosis tends to be excellent in these cases, with no recurrence or need for adjuvant therapy

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