Neuroblastoma, ganglioneuroblastoma and ganglioneuroma are tumors of varying maturity derived from the primordial neural crest cells that form the sympathetic nervous system. A primary extra-adrenal retroperitoneal ganglioneuroma was found incidentally in a 12-year-old boy during a series of examinations for abdominal distension, left upper quadrant pain and diarrhoea.

Key words: Ganglioneuroma; retroperitoneum; sympathetic nervous system.

CASE REPORT

A 12-year-old boy presented with a 18-month history of abdominal distension, left upper quadrant pain and diarrhoea. The patient was referred to our clinic for further investigation of left retroperitoneal mass incidentally discovered by ultrasonography. Abdominal computed tomography (CT) revealed a 70x54x53 mm solid tumor between the upper pole of the left kidney and aorta (Fig. 1). Hormonal studies were insignificant. Intraoperatively, no abnormality was observed in the left kidney. The excised tumor was well-demarcated. The tumor specimen was noncystic, weighing 82 g and measuring 7.4x5.8x5 cm (Fig. 2).

Histologically, hematoxylin-eosin-stained slides revealed a tumor composed of two major components. Spindle cells, which hosted groups of mature ganglion cells with plenty cytoplasm, big nuclei and prominent nucleoli, did not
show any atypia (Fig. 3). The spindled tumor cells showed strong positive reaction for S-100 protein and were negative for synaptophysin, desmin and α smooth muscle actin immunohistochemically (Fig. 4).

**DISCUSSION**

Neuroblastoma, ganglioneuroblastoma and ganglioneuroma are three tumors that differ in their degree of cellular and extracellular maturation; immature tumors tend to be aggressive and occur in younger patients (median age, just under 2 years), whereas mature tumors occur in older children (median age, approximately 7 years). \[1\]

Ganglioneuroma is relatively rare and difficult to distinguish from other tumors due to lack of image findings specific for ganglioneuromas. \[4\] Ultrasonography reveals a homogeneous, hypoechoic mass with well-defined borders. Computed tomography is the most commonly used imaging modality for assessment of neuroblastic tumors, because it reveals extent of tumor, organ of origin, regional invasion, vascular encasement, adenopathy and calcification. \[1\]

In this case report we point out that the correct evaluation of this lesion is of crucial importance, as there are some malignant neoplasms that have similar clinical presentations. \[8\]
However, ganglioneuromas can cause symptoms due to local expansion and pressure on adjacent structures.[6] In our case, informed consent must include the possibility of nephrectomy to avoid any medicolegal complications because a tight adhesion of the tumor to the left kidney can necessitate an en bloc resection. Treatment consists of complete surgical resection of tumor when possible.

Increasing numbers of these tumors are being found incidentally by ultrasonography or CT. Ganglioneuroma is a benign tumor with symptoms depending on location and can be surgically dissected with favorable prognosis. Local recurrence has been reported, so periodic radiologic surveillance is performed after resection.

REFERENCES