CASE NOTE

Nodular ganglioneuroblastoma in adults

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Ganglioneuroblastoma is a primary malignant tumour of the sympathetic nervous system. It is very uncommon in the adult population, with fewer than 40 cases reported in the literature. We present here the case of a woman with a retroperitoneal ganglioneuroblastoma that was discovered in spite of a false-negative preoperative biopsy result.

CASE REPORT

A 34 year-old otherwise healthy woman was admitted to hospital for atypical right-sided back pain that was radiating to her thigh. A computed tomography (CT) scan showed a 4-cm homogeneous retroperitoneal tumour without contrast enhancement (Fig. 1). We performed a CT-guided biopsy. The pathological examination result was interpreted as a probable solitary fibrous tumour (immunohistochemical tests did not contribute to the diagnosis).

The growing tumour (8 cm 3 months later) and the patient's pain compelled us to remove the tumour surgically. We performed the resection via a transabdominal approach (Fig. 2). The tumour was encapsulated and easy to remove except near the L2 paraspinal region, where dissection was difficult. There were no perioperative complications, including blood loss. We discharged the patient on the sixth postoperative day.

Pathological examination revealed a lobulated circumscribed solid mass. Histological and immunohistochemical examination results (Table 1) confirmed the diagnosis of a nodular ganglioneuroblastoma. The resection

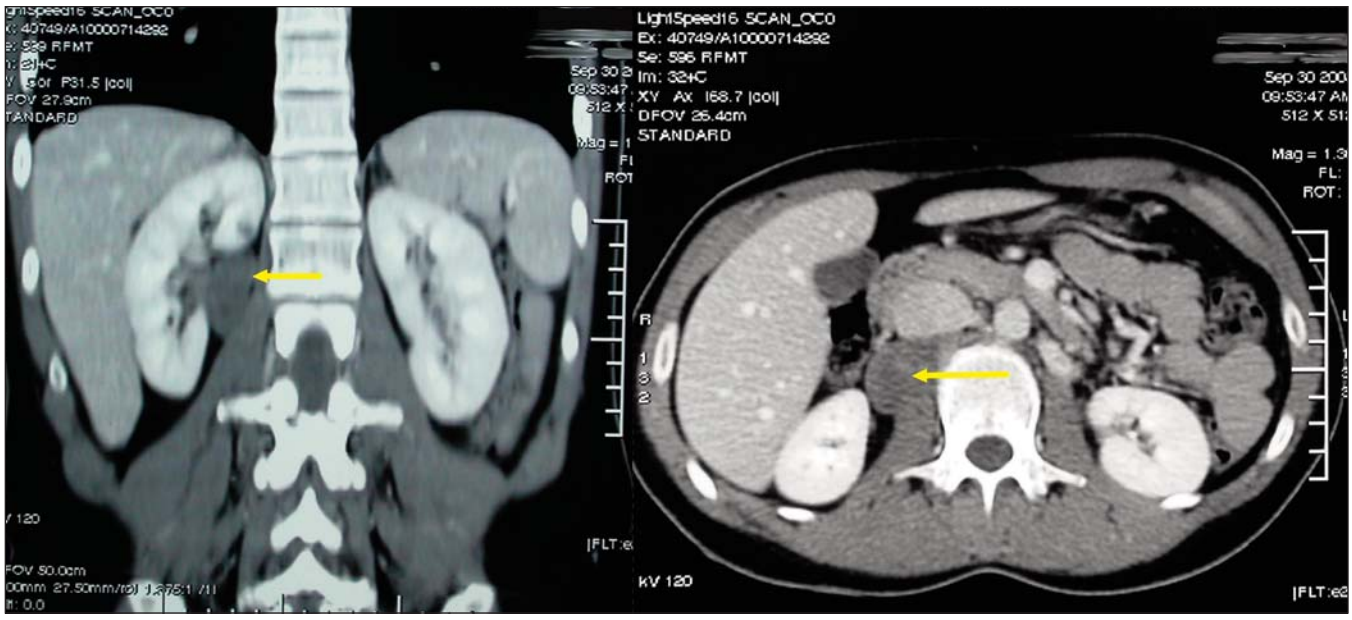

Fig. 1. Computed tomography scan of a 34-year-old woman showing a retroperitoneal ganglioneuroblastoma (arrows).

margins revealed microscopic residual tissue (R1) in the L2 paraspinal region.

All the regional and general paraclinic examination results were normal, including blood and urinary catecholamine concentrations, spinal magnetic resonance images (MRI), thoracic computed tomography scans and ^{111}In-octreotide scintigraphy scans. To locally control the R1 resection, we proceeded with adjuvant radiotherapy at a total dose of 45 Gy over the L2 paraspinal region. Thirty-nine months after surgery, the patient was healthy, and CT scans showed no recurrence.

Discussion

Ganglioneuroblastoma is a rare childhood tumour and rarely appears in adults. More than 90% of cases are reported in children younger than 5 years of age, with a spike at 18 months. In 1976, Kilton and colleagues[1] published a review of 33 cases of ganglioneuroblastomas in adults worldwide. It was updated in 2003 by Koike and colleagues,[2] who reported 37 cases. Ganglioneuroblastomas occur most frequently in cervical, mediastinal, adrenal and retroperitoneal locations. We report what we believe to be the eighth published occurrence of a retroperitoneal ganglioneuroblastoma in an adult.

In adults, this tumour is generally discovered by accident or by compression. Our patient's atypical back pain, for example, can be explained by compression of the psoas muscle and the nervous roots. Preoperative diagnosis is difficult, but CT scans generally reveal a mass with smooth margins, and it is sometimes possible to discern a heterogeneous pattern, tumour calcifications and contrast enhancement.[3] Characteristics of the ganglioneuroblastoma on MRI have been detailed elsewhere[2-4] and can help to discriminate between an adenoma or a cyst. However, MRIs cannot discriminate between adrenal malignancy, neuroblastoma, phaeochromocytoma and ganglioneuroblastoma.

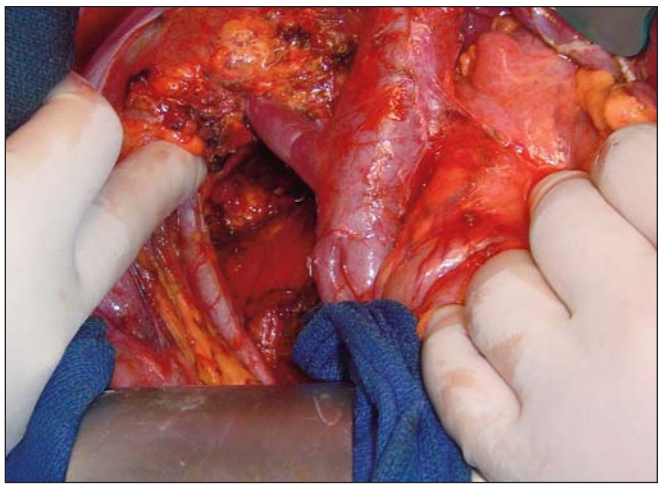

Fig. 2. Operative view of the resection region between the right renal pedicle, vena cava and right ureter.

It is also important to note that preoperative biopsies can produce false-negative results. Ganglioneuroblastomas can look like a neuroblastoma in a partial ganglioneuroma stroma. There are 2 subtypes described in the literature:[5] the nodular subtype contains gross nodules of neuroblastoma (immature small cells) in large expanses of ganglioneuroma (big mature cells in a fusiform stroma), whereas the intermixed subtype consists of microscopic nests of neuroblastoma situated in a ganglioneuromatous stroma. In the nodular subtype, the neuroblastoma nodules and ganglionic cells can represent less than 5% of the tissue: a large biopsy is required for the diagnosis. In our patient's case, the fine-needle biopsy removed only fibrous stroma tissue, so we diagnosed a solitary fibrous tumour (viewing only fusiform cells).

Sometimes, laboratory test results can be helpful in making a preoperative diagnosis. Fifty-seven percent of patients with a ganglioneuroblastoma show increased concentrations of serum serotonin and urinary catecholamine and their metabolites (vanil mandelic acid and homovanillic acid).[2] When they are initially positive, hormonal examinations are reliable for the follow-up of recurrence in children.

In adults, prognosis depends on surgical margin resection. According to Koike and colleagues,[2] every adult patient with a ganglioneuroblastoma that was partially resected (R1 or R2) or not resected died within 24 months.[2] We proposed adjuvant radiotherapy in our patient for regional control of the partial R1 resection because there was no distant metastasis. We required 6-month follow-up with CT imaging of the region and would have proposed an aggressive large resection had there been any regional recurrence.

Ganglioneuroblastoma in adults is very uncommon. Characteristics of this tumour remain poorly understood, and preoperative diagnosis is difficult. Our patient's case shows that a negative fine-needle biopsy cannot reliably rule out a diagnosis of nodular ganglioneuroblastoma. Curative treatment should be a complete resection. However, in case of only partial resection, adjuvant radiotherapy may be proposed with a close follow-up and regular re-evaluation.

Competing interests: None declared.

Table 1. Ganglioneuroblastoma immunohistochemical patterns

Cell	Immunohistochemistry
Fusiform Schwann	PS100+
Ganglioma and neuroblastoma	NSE+ Chromogranin+ Synaptophysin+ CD56 (NCAM)+

NCAM = neural cell adhesion molecule;
NSE = neuron-specific enolase.

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