Oncology

A big encapsulated adrenal mass with well-circumscribed margin

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ABSTRACT

We present a case on adrenal schwannomas. The CT shows enhancement of distal adrenal gland lesion. We named it rabbit tail sign. Adrenal schwannomas are rare and presents a very small proportion of retroperitoneal schwannomas. It tends to be misdiagnosed because of lacking characteristics clinical presentations and CT features. And there is debate about its origination. The rabbit tail sign of distal adrenal gland is one typical radiographic feature of adrenal schwannomas. It also suggests the tumor may originate from dominate nerve of adrenal medulla.

Introduction

In 1920, schwannomas are discovered in the peripheral nerve sheath tumors by Nils Antoni. Schwannoma is a benign tumor originating from the myelin sheath of autonomic, peripheral, or cranial nerves. It is predisposed to head and neck regions. Furthermore, retroperitoneal schwannomas are rare (accounting for 0.7–5.0%). And primary juxta-adrenal or adrenal schwannomas, represents a very small proportion of them. It is mostly benign, no hormonal production, lack of characteristics clinical presentations. Clinically, it tends to be misdiagnosed as other adrenal tumors. There is no debate about its origination. Histologic examination is the gold standard. Positive labeling for S100 and Sox10 plays important roles in immunohistochemical examination. So, if we can dig out some preoperative radiological features about it, the unnecessary extensive surgery could be avoided. However, there are few studies examining the CT imaging features of this tumor. A report, published in 2016, demonstrated 4 patients’ CT scans displayed enhancement of distal adrenal gland lesions (rabbit tail sign). We report an asymptomatic case in which rabbit tail sign also be found.

Case presentation

A 56-year-old woman was admitted to our department for 2-month history of occasional right lumbar pain. Her medical history included the thyroid neoplasms surgery therapy. Abdominal multiphase contrast-enhanced CT scan was taken immediately and showed a big (60 × 40mm) encapsulated oval mass with well-circumscribed margin in right adrenal region. The mass displays almost homogeneous hypodense and patchy calcifications in CT plain scan. Patchy moderate enhancement, enhanced capsule and rabbit tail sign could be seen on corticomedullary phase a nephrographic phase (Fig. 1). The CT values on three phases are 45, 60, 66Hu, respectively. The brain, lung CT and whole body bone scan are all normal. Laboratory investigations indicated that no hormonal production had been detected (Table 1).

Considering the diameter of tumor was over 60mm and the possibility of malignancy, the retroperitoneal laparoscopic adrenal gland tumor excision was taken smoothly.

After operation, final pathologic report proved the mass to be adrenal schwannoma. The photomicrograph of lesion included spindle-shaped cells (Fig. 2). Immunohistochemically, the tumor cells were consistently and diffusely positive for S100 and Sox 10. Repeat CT scan done 2 weeks after the operation showed no signs of recurrence. Patient’s blood biochemistry data were all within the normal range. Her right lumbar pain was disappeared.

Discussion

For adrenal incidentaloma, the American Association of Endocrine Surgeons recommend that if the tumor is < 4 cm with benign features and no hormonally reactive evidence, then it can be followed up; tumors >4 cm should be removed. National Institutes of Health consensus statement mentioned that lesions <4cm with benign characteristics are generally not removed; lesions between 4 and 6 cm can be either observed or resected, and those >6 cm should be excised. But, the recommendations above are applicable to adenoma. Considering the diameter of our patient’s tumor was over 6 cm and occasional lumbar

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pain, the retroperitoneal laparoscopic adrenal gland tumor excision is appropriate. However, the unnecessary extensive surgery is ubiquitous, especially the lesions’ diameter between 4cm and 6cm. Generally, this cohort of patients would be misdiagnosed of nonfunctioning adrenal adenoma (NAA). Most NAA patients are asymptomatic. The masses are usually found incidentally during CT or MRI conducted. Cysts and lipomas make up most of the remainder. Histopathologic examination is the only way to make a finally correct diagnosis. It’s the gold standard. Adrenal schwannoma has various histologic features, including Antonia A and B areas, foamy cells, verocay bodies, capsules and spindle-shaped cells. Positive labeling for S100 or Sox10 plays important roles in adrenal schwannoma’s immunohistochemical examination. Based on the situations above, the more imaging features be found out, the more unnecessary surgeries can be avoided. The enhancement of distal adrenal gland lesion can be considered one of them.

In 2016, a report by the Shanghai east hospital and Tongji university school of medicine, Cao Kaiming and Wang Wei analyzed 8 cases and indicated that half of them displayed rabbit tail signs. There are two opinions about the origination of adrenal schwannoma. One is adrenal tissue, the other is sympathetic nerve of adrenal medulla. According to the rabbit tail signs, we speculate that it stems from sympathetic nerve of adrenal medulla. The CT scans may help the accuracy of diagnosis. But histologic examination is still the gold standard. However, clinicians must be aware of the potential diagnosis of schwannoma when the pre-operative imaging shows the enhancement of distal adrenal gland lesion, especially asymptomatic patients. Further studies are needed.

In conclusion, the rabbit tail sign of distal adrenal gland implicit the adrenal schwannoma and speculate that it stems from sympathetic nerve of adrenal medulla. Adrenal schwannoma is an extremely rare tumor and mostly appears as an incidentaloma. This cohort of patients would be misdiagnosed of NAA due to unfamiliarity with the clinicopathologic features of adrenal schwannoma.

Data and materials availability statements

All data generated or analyzed during this study are included in this published article (and its supplementary information files).

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Authors’ contributions

Yi Yi drafted the report and approved the final version of the manuscript. Chaolu Lin cared for the patient and approved the final version of the manuscript. Feng Yu, JL Zhang, Qinqi Chen and LU Song reviewed the report and approved the final version of the manuscript.

Declaration of competing interest

The authors declare that they have no competing interests.
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