The Master Masquerader: A Case Series on Adrenal Teratoma

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Abstract

Adrenal teratoma is a rare entity that often resembles other adrenal pathology in imaging studies and intraoperatively. A high degree of suspicion, as well as knowledge about imaging characteristics of teratoma, is essential to make a diagnosis.

Keywords: Adrenal, Adrenocortical cancer, Case report, Endocrine surgery, Heterogenous adrenal mass, Retroperitoneal tumors, Teratoma.

As surgeons, we do not like surprises when it comes to the diagnosis of our patients. We want to know and be sure. However, we have encountered intraoperative surprises during our training/practicing periods. Here, we present a case series on our institutional experience with such an intraoperative surprise. Teratomas are rare tumors having their origin in embryonal germ layers. Most of them are mature, and gonads are their most common location. However, 15% are extra gonadal. Primary adrenal teratomas are very rare. In this case series, we present two cases of adrenal teratoma.

Case Description

Case 1
A young lady aged 27 years presented with pain in the abdomen. She underwent evaluation and was found to have a large non-functional right adrenal mass. Contrast-enhanced computed tomography (CECT) abdomen was suggestive of a large (8 × 9 cm) heterogenous density adrenal mass with calcifications (Fig. 1). She underwent a right open transperitoneal adrenalectomy. The specimen showed a large adrenal mass with variegated consistency, yellow fat, hard calcified elements, and other firm areas (Fig. 2). Histopathological examination (HPE) showed it to be a mature teratoma. The patient is doing well on 3-year follow-up.

Case 2
A young male aged 23 years presented to us with right abdominal pain persistent for 1 month with no other significant history.

Fig. 1: Computed tomography image

Fig. 2: Gross specimen

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The results of the complete hematological examination and hormonal assessment were unremarkable. Contrast-enhanced computed tomography abdomen revealed a 12 × 12 × 13 cm heterogenous right adrenal mass with calcifications and cystic changes, with hypodense areas likely fat, which was reported to be Adrenocortical carcinoma (Fig. 3). He underwent a right open transperitoneal adrenalectomy. Preoperatively the tumor was densely adherent to the IVC and the right liver lobe, making mobilization difficult. A gross postoperative examination revealed a whitish-grey mass measuring 16 × 15 × 12 cm with cartilage and bone-like hard tissue (Figs 4 and 5). The postoperative period was uneventful, and on POD 4 the patient was discharged. Histopathological examination showed an encapsulated neoplasm displaying haphazardly arranged derivatives of all three germ layers consistent with mature cystic teratoma.

Discussion

Teratomas are germ cell tumors composed of mature tissues arising from more than one germinal layer, i.e., ectoderm, mesoderm, and endoderm. Childhood teratomas are usually found at locations other than the gonads whereas those occurring in adults are predominantly located in the gonads.\(^2\) The adrenal gland is a very rare location accounting for 1–11% of all primary retroperitoneal tumors.\(^3\) Earlier studies showed that adrenal teratomas are more often found in women than men.\(^4\) Zhong et al. found an equal distribution in both sexes.\(^5\) However, in our series, the teratoma was present on the right side in both patients.

Primary adrenal teratoma has no unique clinical presentation and usually has normal levels of adrenal hormones present as “incidentalomas”, and is indistinguishable from non-functioning adrenal tumors. In day-to-day clinical practice, adrenal teratoma is detected during the workup and diagnosis of other functional and non-functional adrenal tumors like phaeochromocytoma or adrenal adenoma.\(^6\) Hence arises the need to differentiate adrenal teratomas from incidentalomas, non-functioning adrenal tumors, cysts arising from adrenal, and adrenocortical carcinomas. Adrenal teratomas usually attain large size because the retroperitoneum provides sufficient space for their growth. As the tumor enlarges, patients may develop symptoms such as pain abdomen, distension of the abdomen, backache, or even obstruction of the bowel due to the mass effect on nearby organs.\(^7\)

In previous cases, adrenal teratomas have been wrongly diagnosed before surgery. Hence pathological examination of the postoperative specimen is needed to arrive at a confirmative diagnosis. Mature teratomas are generally benign tumors and can compress the nearby structures as the tumor grows. Bleeding, infection, and tumor rupture are the usual complications. After diagnosis surgery should be done at the earliest. Laparoscopic surgery is the “gold standard” for the treatment of adrenal tumors smaller than 6 cm.\(^8\) Open surgery is preferred for tumors, more than 6 cm. However, in the recent past, it has been proven that minimally invasive surgery is also safe for large benign adrenal tumors if intraoperatively or on preoperative imaging, there are no features of local invasion.\(^9\)

The etiology of teratomas is not very well known, and pathologically they are classified as mature teratomas and immature teratomas. Benign teratomas are characterized by (1) a lack of immature or malignant components in the tumor, (2) absence of other similar lesions in the body, (3) AFP and HCG are within normal range, and (4) On follow-up there is no evidence of
reoccurrence. Mature teratoma has a good prognosis. However, 1.46% of mature teratomas can turn malignant.10 Hence, it necessitates regular follow-up. However, chemotherapy and radiotherapy are deemed unnecessary. In immature teratoma, the risk of metastasis and recurrence has been significant. Therefore, chemotherapy and radiotherapy are essential after surgery. The patient has to be followed up lifelong.11

In our case series, both lesions were histopathologically mature teratomas. One of our patient is doing well on a 3-year follow-up; the other has completed an uneventful one-month post-surgery.

**Conclusion**

We have briefly described the features of pathology, imaging, and the management of the typical primary adrenal teratoma to create awareness of this disease, which needs to be considered in the differential diagnoses of adrenal lesions.

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