Primary Mature Cystic Teratoma of the Retroperitoneum Presenting as a Suprarenal Mass in an Adult Patient: A Case Report

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Received 2022 April 24; Accepted 2022 August 25.

Abstract

Introduction: Primary retroperitoneal mature cystic teratomas (PRPMCT) are uncommon in adults. They typically occur in infants under six months old and young females. So far, some limited case reports have been documented in the literature. This case report presents a rare case of an adult patient with PRPMCT aged over 30 years.

Case Presentation: The patient was a 34-year-old female with PRPMCT in the site of the left adrenal gland and abdominal discomfort. Computed tomography (CT) scan revealed a 66 mm hypodense lesion with fat attenuation and calcification in the left retroperitoneum. Laparotomy with resection of the retroperitoneal (RP) lesion was done. Gross examination showed a solid multicystic mass measuring 8x6.5x6 cm. Histopathologic examination revealed a neoplastic tissue composed of mature osseous and cartilaginous tissue associated with stratified squamous epithelium and lumens lined by ciliated columnar epithelium, and the diagnosis was mature cystic teratoma. The patient had an uncomplicated postoperative course without recurrence after seven months of follow-up.

Conclusions: PRPMCT in adults is extremely rare. RP metastasis of primary gonadal teratoma and non-functioning adrenal tumors should be excluded first. Due to the diagnostic difficulty of posterior peritoneal teratoma with radiological imaging and its risk of malignancy, surgical resection would be the most effective diagnostic and therapeutic option. Complete resection of the mass is important to judge the existence of immature and solid components that require a long-time follow-up because of the increased risk of malignancy.

Keywords: Suprarenal Mass, Retroperitoneum, Primary Mature Cystic Teratoma

1. Introduction

Primary retroperitoneal mature cystic teratomas (PRPMCT), known as dermoid cysts, are rare non-seminomatous germ cell tumors, consisting of well-differentiated parenchymal tissues from a minimum of two layers of germ cell (1, 2). They usually arise in paraxial structures, mostly inside the gonads (1, 3). The extra-gonadal sites make up 15% of all teratomas, and retroperitoneum is below 4% of the cases (1, 3-5). They are more common in infants or young girls, usually within the sacrococcygeal region but very rarely in adults (1-6). Since gonadal malignancies usually metastasize to the retroperitoneum, it is important to distinguish primary retroperitoneal neoplasms (6). Most of the cases are asymptomatic or present with vague symptoms or are identified incidentally. Surgical removal of mature teratoma persists as the basis of treatment and is necessary for a conclusive diagnosis (6, 7).

This report presents an infrequent case of a PRPMCT in an adult patient aged over 30 years. The current case confirms the restriction of image-based diagnosis of adrenal masses.

2. Case Presentation

A 34-year-old woman was introduced with an obscure history of left upper quadrant fullness and abdominal pain for three months. There were no other relevant symptoms, such as fever, appetite or weight loss, nausea, or vomiting. The patient had no medical and surgical history, and her family history was unremarkable. The social history was negative for cigarette smoking and alcoholism. Physical examination was normal, without any palpable mass or tenderness. Laboratory tests to exclude a functional adrenal tumor and tumor markers (CA19.9, CEA, AFP, hCG, and LDH) were within normal limits.

On evaluation with ultrasound, there was a multilocular lesion without vascularity in the left side of the
retroperitoneum at the suprarenal region with an 8 cm diame-
ter. Ovarian and other sites were normal. An abdomi-
nal contrast-enhanced computed tomography (CT) scan
revealed a 66 mm circumscribed, fluid component hypo-
dense lesion with fat attenuation and calcification in the
left retroperitoneum above the kidney. No abnormal en-
hancement and RP lymphadenopathy were identified.

A core needle aspiration and biopsy of the cystic mass
were performed. However, the collected sample was not
satisfactory.

Laparotomy was performed by resection of the RP le-
sion of the left adrenal gland. The bulky cystic tumor was
encapsulated and no invasion of renal tissue was observed.
Left adrenal and bilateral ovaries were normal.

Gross examination showed a solid multicystic mass
measuring 8x6.5x6 cm filled with pasty material. The max-
imal wall thickness was 1.2 cm. Histopathologic examina-
tion revealed a neoplastic tissue composed of mature os-
seous and cartilaginous tissue associated with stratified
squamous epithelium and lumens lined by ciliated colum-
nar epithelium. Mucinous salivary gland, adipose tissue,
and smooth muscles were also present, and the diagnosis
was mature cystic teratoma (Figure 1).

There were no postoperative complications, and she
was discharged on day 4. Currently, she is well seven
months after resection.

3. Discussion

A PRPMCT in the left adrenal gland space in a 34-year-
old woman is an uncommon phenomenon, and only infre-
cent cases have been presented (6). Teratomas are non-
seminomatous germ cell tumors developed from pluripo-
tent embryonic cells (1, 8). They are categorized based
on the degree of tissues maturity (mature or immature)
and the tumor components (cystic, solid components, or
mixed) (9). Although teratoma is most common in the
ovaries and testes, it can arise in nearly any area of the
body. Most of the RP neoplasms include metastasis of go-
nadal malignancies, and primary neoplasms contain only
0.1 - 0.3% of cases; and 1% (6) – 11% (10) of these cases are ter-
atomas (1, 10-12). They are usually seen in children within
the primary six months and at younger ages (6, 13, 14). Merely 10 - 20% of these lesions happen in adults over 30
years old (2, 6, 14). The most admitted theory regarding
the origin of the primary RP teratoma is that they are a
residue of the Wolffian and Müllerian ducts or are devel-
oped from pronephric or mesonephric tubules that corre-
spond to their midline and paramedian position (6). As in
the reported case, RP teratomas in adults are usually de-
tected on the left side of the body, in the pararenal area (4,
6, 13).

These lesions are generally asymptomatic or exist with
nonspecific complaints, which makes them hard to detect.
The position of the teratoma leads to the impaction symp-
toms of the neighboring organization such as vomiting,
 constipation, back pain, distention, and edema. Systemic
symptoms such as fever, chills, night sweats, and weight
loss could be found too (6). PRPMCT may present with
chemical peritonitis following the cyst rupture (15), and
may also get infected (16). Uncommon expressions con-
sist of ocular myasthenia and hypertension that were elimi-
nated following resection of the lesion (17, 18).

Serum markers, like CA19.9, CEA, and AFP, can be ele-
vated in PRPMCT. Serum levels of AFP and HCG are good
representatives of the presence of malignant germ cell
elements (19). Increased AFP measures were present in
50% of immature teratomas and 6% of mature teratomas
(20). These markers are evaluated preoperatively to ex-
clude other germ cell tumors and follow up postopera-
tively to use for tumor surveillance (6). If the PRPMCT in-
vades the adrenal gland, plasma catecholamines, renin, al-
dosterone, ACTH, and cortisol levels should be measured
too (21). These markers were evaluated in the reported case
preoperatively, and the results were in normal ranges. The
report confirms that these tumors must be evaluated for
the differential diagnosis of adrenal masses because a ra-
diological assessment did not affirm the source of the RP
mass (22).

Radiographic evaluations have an important role in
the assessment and planning of the teratoma’s surgical
treatment. Ultrasound can distinguish between cystic and
solid components, but it has limited sensitivity, which
makes CT scan a more suitable imaging (22, 23). CT scan
can provide better soft-tissue resolution and detect malig-
nant characteristics; it is the imaging modality of choice
and helps present the extent of disease in gonadal malig-
nancies to the retroperitoneum, and involvement of blood
vessels (22, 23). CT scan results suggested that primary cys-
tic teratoma contained a well-restricted fluid element, hy-
poattenuating fat, and calcifications in the reported case
(2, 13). Even though magnetic resonance imaging (MRI)
was not a part of the assessment, in this case, they can pro-
vide better soft tissues resolution, possible diagnosis of be-
nign and malignant features, and are superior for tumor
staging (22).

Nevertheless, a certain diagnosis is formed on histo-
pathological examination of the resected tumor. Complete
excision is the choice of treatment and is recom-
mended in all cases (6, 7, 24). Prognosis is favorable after
complete surgical resection with an overall 5-year survival
rate close to 100% (25). Malignant features are unusual in
mature cystic teratoma, and these neoplasms are greatly
resistant to radio- and chemo-therapy (6). Therefore,
invasive surgical excision is suggested for the treatment of these patients with clear oncological margins.

The rate of malignant RP teratomas in adults is notably higher than in children (36.3% vs. 6.8%) and it is elevated with age, male gender, and the existence of immature tissues and solid elements (4, 6, 9, 10). The absence of malignant or immature components within the tumor, absence of other similar lesions in other parts of the body or recurrence, and normal measures of AFP and hCG are the pathological criteria for nonmalignant lesions (7). Regardless of the benign feature of mature cystic teratoma, close follow-up is advocated due to ~3-6% incidence of malignant transformation (7). Our case had no symptoms, recurrence, or other complications for seven months postoperatively.

3.1. Conclusions

PRPMCT rarely occurs in adult patients and must be placed in the differential diagnosis of nonfunctional adrenal masses. RP metastasis of primary gonadal teratoma should be excluded. They are usually asymptomatic, but even if they are detected by CT and MRI before surgery, a definitive diagnosis is established upon histologic assessment.

Complete resection of the tumor is needed to recognize immature and solid components that require long-time follow-up because of the greater danger of malignancy. Surgical excision is the base of management for PRPMCT, which results in 5-year survival of approximately 100% if the margins are oncologically clear.

Footnotes

Authors’ Contribution: Farhood Khaleghimehr: Study concept and design; critical revision of the manuscript for important intellectual content; administrative, technical, and material support; and study supervision. Maryam
Abolhasani: Drafting of the manuscript; study concept and design; critical revision of the manuscript for important intellectual content; administrative, technical, and material support; and study supervision. Yasin Zakeri: Drafting of the manuscript and study concept and design.

Conflict of Interests: Funding or Research support: No
Conflict of Interests: Intellectual content; administrative, technical, and material support; and study supervision. None of the authors is an editorial board member or a reviewer of this journal.

Ethical Approval: Written informed consent was obtained from the patient.

Ethical Approval: This case report was approved by the ethical committee of Hasheminejad Kidney Center (HKC).

Funding/Support: This study was not funded by any organization.

Informed Consent: Written informed consent was obtained from the patient.

References


