PRIMARY MATURE RETROPERITONEAL TERATOMA INVOLVING THE ADRENAL GLAND.

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ABSTRACT
Retroperitoneal mature cystic teratoma arising from the adrenal gland is a rare retroperitoneal tumor accounting for only 4% of all primary teratomas. Though mature cystic teratomas of extra gonadal sites are unusual, (1) those arising in the adrenals are exceptionally rare. (2, 3) They are more common in childhood and rarely occur in adults. (4) Only very few cases, mostly in young patients have been reported. Most teratomas in this region are secondary to germ cell tumors of the testicles or ovaries. To be very specific, in male patients, retroperitoneal germ cell tumors are more likely to have metastasized from the testes than presenting as primary tumors. On histological examination, they are composed of variable proportions of tissue originating from the ectoderm, mesoderm, and endoderm. Although gastrointestinal epithelium is occasionally seen in these tumors, the presence of a complete intestinal wall is rare. We report a case of primary mature cystic teratoma involving the left adrenal gland with portion of the mature component being intestinal wall.

KEYWORDS:
Adrenal gland, colonic wall, mature cystic teratoma

Introduction:
Primary mature cystic teratomas are uncommon non-seminomatous germ cell tumors. They are made up of well-differentiated parenchymal tissues that are derived from more than one of the three germ cell layers (6). They usually occur in midline structures. The most common sites are gonads followed by extra gonadal sites such as intracranial, cervical, mediastinal, retroperitoneal, and sacrococcygeal regions (7). Primary retroperitoneal teratomas involving adrenal glands are exceedingly uncommon accounting for only 4% of all primary teratomas (7-9). Only a very few case reports have been documented in literature so far (10). The majority of cases are asymptomatic, present with nonspecific complaints, or identified incidentally on routine investigations (11). Confirmatory diagnosis of mature teratoma comes by histopathological examination (12). Prognosis is fortunately excellent after complete surgical excision remains the mainstay of treatment and prognosis is excellent after resection (13). Herein, we report a mature cystic retroperitoneal teratoma in the region of left adrenal gland in an otherwise healthy female patient who presented with a 1-month history of left flank pain and hypertension.

Case report:
A 45 year old female presented with flank pain for one month and incidental hypertension. On physical examination, a mobile mass in the flank was identified. All other laboratory investigations were normal. CT abdomen was done and the patient was found to have a mass in the left retroperitoneal region involving the adrenals. A diagnosis of phaeochromocytoma was made because of the coexisting hypertension and surgical resection was advised. Resection of the mass in toto was performed and sent for histopathology.

Grossly, the resected mass measured 12x10x8 cms. Surface was smooth and lobulated. Cut section shows a multiloculated cyst filled with grey white material and adipose tissue with focal areas of calcification. Multiple sections were taken and submitted for histopathology. Sections revealed fibro muscular wall lined partly by respiratory and partly by intestinal epithelium. Focal area showed a portion
histologically resembling wall of the large intestine. Lobules of mature adipose tissue, Spicules of mature bone and cartilage identified. A diagnosis of retroperitoneal teratoma involving the left adrenal gland was made. No immature elements were identified.

**Discussion:**

Generally, teratomas arise from uncontrolled proliferation of pluripotent cells: germ cells and embryonal cells. The type of pluripotent cell giving rise to the tumor greatly influences the presentation time and site of teratoma. Teratomas of germ cell sources can be congenital or acquired and are usually found in gonads (6,7). In contrast, teratomas of embryonic cell sources are always congenital and are usually found in extra gonadal locations, such as intracranial, cervical, retroperitoneal, mediastinal, and sacrococcygeal sites (11-15).

Teratomas can be diagnosed based on high index of clinical suspicion, routine laboratory, and radiographic investigations (17,18). With respect to high index of clinical suspicion, retroperitoneal teratomas involving adrenal glands may present congenitally, or later in life when they grow to massive sizes (19). Clinical presentations are variable and include nonspecific back pain, obstructive gastrointestinal and genitourinary symptoms, as well as lower limb swelling due to lymphatic obstruction. They can rarely present with complications such as secondary infections, traumatic rupture leading to acute peritonitis, or malignant transformations.

With respect to laboratory investigations, retroperitoneal teratomas can express a diversity of serum tumor markers such as elevated alpha-fetoprotein (AFP), carcinoembryonic antigen (CEA), and CA 19-9. These serum tumor markers are helpful in clinical practice and can be used to monitor successful treatment or detect relapse in patients with specific tumor marker-secreting teratomas. The diagnosis of adrenal teratoma relies predominantly on an imaging examination because the findings from laboratory examinations will often be normal. On CT scans, teratoma is frequently shown as a heterogeneous fat dense mass with calcifications. Mature teratoma in the adrenal region can mimic other types of lipomatous adrenal tumor. The differential diagnosis of retroperitoneal teratomas include ovarian tumors, renal cysts, adrenal tumors, retroperitoneal fibromas, Wilms’ tumor, sarcomas, hemangiomas, neonatal cystic neuroblastoma, xantogranuloma, congenital mesoblastic nephroma, enlarged lymph nodes and perirenal abscess.
Conventional imaging techniques cannot exactly distinguish the various types of lipomatous tumor. Histopathology gives the final confirmatory diagnosis.

**Conclusion:**

Adrenal teratomas have been reported extremely rarely in adults, it should be considered in the differential diagnosis of hormonally silent adrenal tumors. In particular, teratoma should be considered in the differential diagnosis of adrenal lipomatous tumors, not only in children and young adults, but also in elderly patients. The final diagnosis depends on the findings of the pathological examination. Once complete resection of the tumor has been made and the diagnosis is accomplished, prognosis for such patients is excellent with 5 year survival being 100%.

**CONFLICT OF INTEREST:**

Dr. Chandramouleswari, although a member of Editorial Board, did not participate in the review of this case report which was done by an independent and autonomous panel.

**REFERENCES:**


