Revista Portuguesa
de Endocrinologia, Diabetes e Metabolismo

www.spedmjournal.com

Caso Clínico

Retroperitoneal Teratoma: A Differential Diagnosis of Adrenal Adenoma

Monica Laureano * [a]

[a] Serviço de Cirurgia 2, Centro Hospitalar de Leiria, Leiria, Portugal

INFORMAÇÃO SOBRE O ARTIGO

Historial do artigo:
Received/ Recebido: 2017-12-29
Accepted/Aceite: 2018-09-01
Online: 2019-09-30

Keywords:
Adrenalectomy
Retroperitoneal Neoplasms/diagnostic imaging
Retroperitoneal Neoplasms/surgery
Teratoma

A B S T R A C T

Case report of a 28-year-old female patient, presenting with a left dorso-lombar pain, with a few months evolution. During the investigation a left adrenal lesion has been found, confirmed by computed tomography.
Laparoscopic left adrenalectomy was performed. Intraoperatively, a posterior abdominal wall nodule was found, independent of the left adrenal gland and kidney. Histopathologic examination revealed a mature cystic teratoma.
Teratomas are congenital tumors, derivate from the three germ layers and are typically located in the gonads. Retroperitoneal teratomas are a rare clinical entity, usually found in children.
The diagnosis of this kind of tumors is a challenge, it is a rare entity in adults and surgical excision is the only definitive treatment.

Palavras-chave:
Adrenalectomia
Neoplasias Retroperitoneais/cirurgia
Neoplasias Retroperitoneais/diagnóstico por imagem
Teratoma

Teratoma Retroperitoneal: Um Diagnóstico Diferencial de Adenoma Adrenal

R E S U M O

Caso clínico de uma paciente de 28 anos que apresentava dor dorso-lombar esquerda com alguns meses de evolução. Realizou estudo ecográfico que identificou uma lesão na glândula suprarrenal esquerda, confirmada por tomografia computorizada .
Foi realizada adrenalectomia esquerda laparoscópica. Intraoperatoriamente identificou-se um nódulo na parede abdominal posterior, independente da glândula suprarrenal e do rim esquerdos. O exame histopatológico revelou um teratoma cístico maduro.
Os teratomas são tumores congénitos que derivam das três camadas germinativas, localizados habitualmente nas gónadas. Os teratomas retroperitoneais são raros e geralmente são detectados em crianças.
O diagnóstico destes tumores é um desafio, são uma entidade rara nos adultos e a sua excisão cirúrgica é o único tratamento definitivo.

* Autor Correspondente / Corresponding Author.
E-Mail: monicalaureano@sapo.pt (Monica Laureano)
Centro Hospitalar de Leiria
Rua das Olhalvas
2410-197 Leiria

http://dx.doi.org/10.1016/j.rpedm.14.1.CC170093

Introduction

Teratomas are uncommon neoplasms derived from the three germ cell layers. They are congenital and usually arise from the gonads. Extragonadal teratomas tend to occur in midline structures as the anterior mediastinum, retroperitoneum, sacrococcygeal region and pineal gland.[1]

Primary retroperitoneal teratomas account for only 1% - 11% of retroperitoneal neoplasms.[2] They occur mainly in infancy and childhood and are rare in adults.[3] They constitute less than 4% of all extra-gonadal teratomas with only partly described in adults.[4]

We report a case of a retroperitoneal teratoma that was mistaken by an adrenal adenoma in the diagnostic workup.

Case Report

A 28-year-old female with known diabetes mellitus type 1 presented with a left dorso-lumbar pain for a few months, a mild hypertension and a difficult diabetic control. Abdominal ultrasound revealed a 3 cm nodular formation, apparently on the left adrenal gland. Abdominal computed tomography (CT) confirmed a nodule arising from the left adrenal gland, well delimited, with 25x38 mm, homogenous, suggestive of an adrenal adenoma (Fig.1).

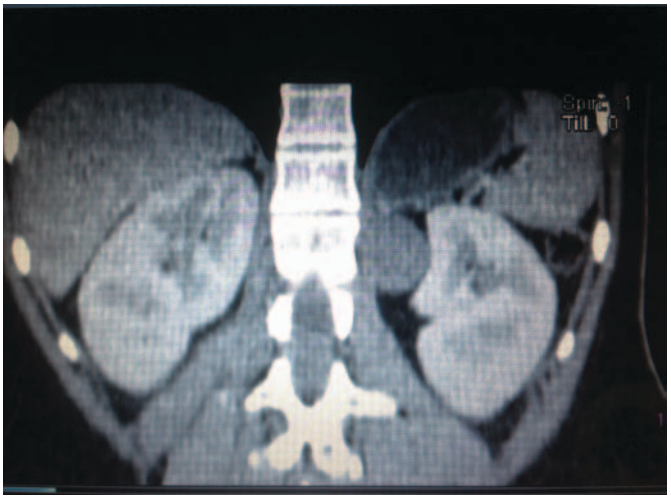

Figure 1. Coronal CT scan cut showing a nodule arising from the left adrenal gland

The biochemical markers used to identify a functioning adrenal tumor showed a slightly elevated plasma cortisol 30.31 ug/dL (N: 4.3-22.4) with the other analysis within normal values (adrenocorticotropic hormone (ACTH) 19.9 ng/L (N: <46), androstenedione 3.04 ng/mL (N: 0.5-4.7), dehydroepiandrosterone sulfate 37.4 µg/dL (N: 25.9-460.2), rennin 1.78 ng/mL/h (N: 0.8-2.5), aldosterone 0.8 ng/dL (N: 1-16), vanilmandelic acid 1.60 mg/24 h (N: 1.8-6.60), urinary metanephrines 425 µg/24 h (N: 329-1263) urinary cortisol 12.3 µg/24 h (N: < 43.0).

Since the nodule was near 4 cm, the patient had back pain and mild hypertension, a laparoscopic adrenalectomy was performed. Subsequently, a nodular lesion was identified, localized posteriorly to the adrenal gland, in the posterior abdominal wall, with the same characteristics described on the CT. The tumor and adrenal gland were excised and bagged removed.

The adrenal gland had macroscopic and microscopic normal aspects. The tumor measured 3.5x3.0x2.5 cm, encapsulated, with smooth surface. Histopathological examination revealed a mature cystic teratoma with multiple tissues such as pseudo stratified ciliated epithelium, mucous glands and cartilage (Fig.s 2,3 and 4).

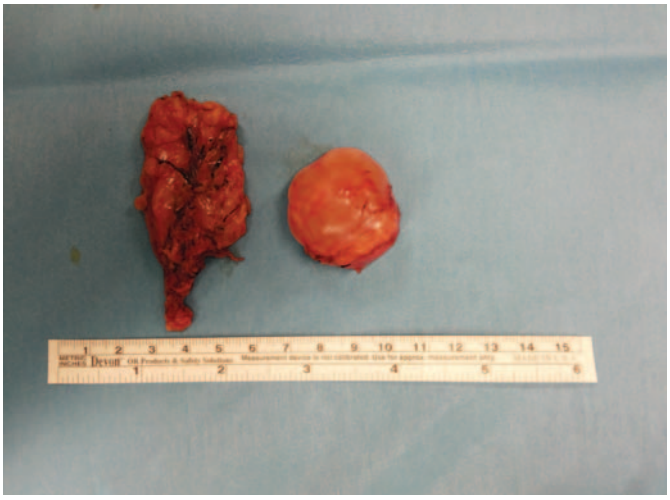

Figure 2. Specimens picture: adrenal gland on the left and tumor on the right

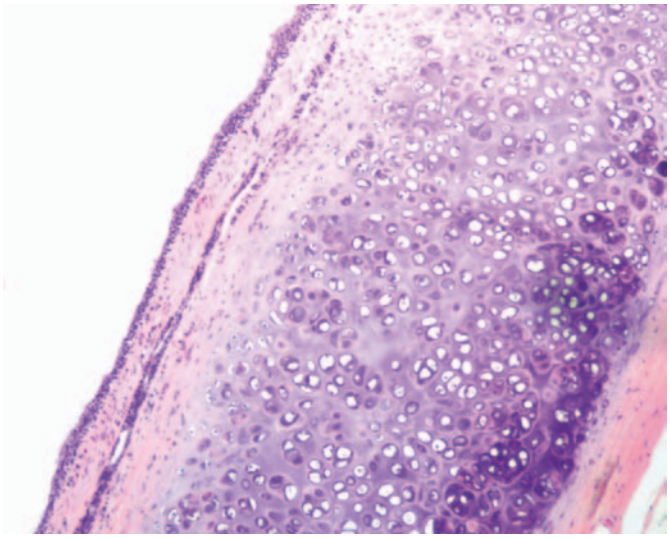

Figure 3. Microscopic aspect of teratoma with pseudostratified epithelium and cartilage

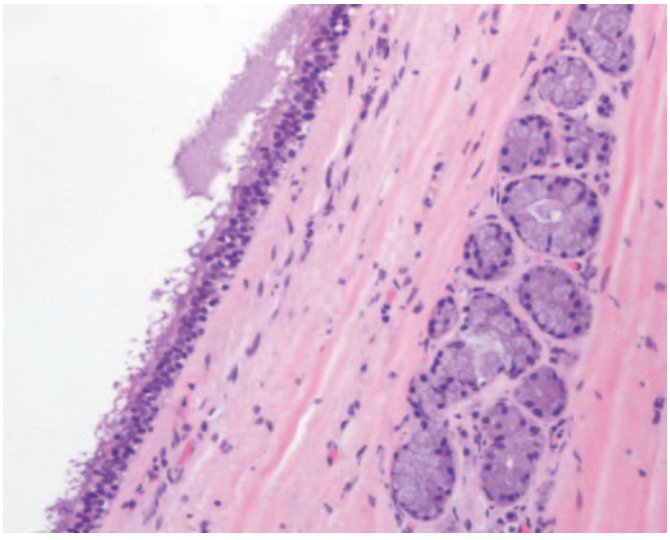

Figure 4. Microscopic aspect of teratoma with mucous glands

As retroperitoneal teratomas are usually secondary tumors, the abdominal CT and chest X-ray images taken prior to surgery were re-examined for evidence of a primary tumor. However, no lesions were found apart from the described one, confirming the diagnosis of a primary retroperitoneal teratoma.

Postoperative course has been uneventful with discharge on the third post-op day. Three-years follow-up has been done without any signs or symptoms of relapse, with no hypertension and controllable glycaemia.

Discussion

Teratoma is a germ cell tumor derived from totipotential cells, which comprises tissues originating from more than one germ cell layer, usually all three, and virtually giving rise to all tissues. Most teratomas are found in gonads. Nevertheless, many extragonadal sites have been reported, including the mediastinum, sacrococcygeal region, pineal gland and retroperitoneum.[5]

Retroperitoneal teratomas are rare in adults and most often located near the upper pole of the kidney, mostly on the left side.[6]

Retroperitoneal teratomas are usually asymptomatic except when compression of the surrounding structures occurs. Patients with compressive symptoms may present with back pain, gastrointestinal or genitourinary symptoms.[6] Our patient symptoms probably resulted from lesion growth, compressing the paravertebral muscular fibers or nerves and also from the compression over the kidneys pedicle.

The differential diagnosis of retroperitoneal teratomas include ovarian tumors metastasis, renal cysts, adrenal tumors, retroperitoneal fibromas, retroperitoneal sarcomas, hemangiomas, xanthogranulomas, enlarged lymph nodes and perirenal abscesses.[7,8]

Teratomas are classified into mature and immature teratomas based on the maturation of their constituents. Mature teratomas, like the one in our patient, are benign, commonly found in females and highly variable on histology. Being made up of well differentiated parenchymal tissue, they can be solid, cystic or mixed. Mature cystic teratomas may even consist of partially to complete well-developed organ systems. In contrast, immature teratomas are solid and made up of immature (undifferentiated) parenchymal tissue. They can be possibly benign, possibly malignant or frankly malignant. Any constituent can undergo malignant transformation and benignity cannot be ascertained until specimen examination, therefore, these tumors must be surgically removed.[9-11]

The majority of retroperitoneal teratomas are secondary neoplasms with male preponderance. Primary retroperitoneal teratomas are extremely rare neoplasms and pediatric age groups are more involved, with female incidence twice that in male. Such teratomas rarely occur in adults and are often mistaken for other adrenal neoplasms[6] which happened with our patient.

Surgical excision, both by open surgery or by laparoscopy, is the mainstay of treatment and confirms the diagnosis. Laparoscopic excision is feasible and offers the advantage of being less morbid and ensuring an early return to work.[12,13] The prognosis is excellent with nearly 100% survival rate after complete excision.[9]

Regardless of the benign nature of mature teratomas, malignant transformation is a possibility and, therefore, close follow-up is required.[14] There are no consensus guidelines of follow-up on this kind of tumors in the literature. We did clinical examination and annual ultrasound with no signs of relapse so far.

Responsabilidades Éticas

Conflitos de Interesse: Os autores declaram a inexistência de conflitos de interesse na realização do presente trabalho.
Fontes de Financiamento: Não existiram fontes externas de financiamento para a realização deste artigo.
Confidencialidade dos Dados: Os autores declaram ter seguido os protocolos da sua instituição acerca da publicação dos dados de doentes.
Consentimento: Consentimento do doente para publicação obtido.
Proveniência e Revisão por Pares: Não comissionado; revisão externa por pares.

Ethical Disclosures

Conflicts of Interest: The authors have no conflicts of interest to declare.
Financing Support: This work has not received any contribution, grant or scholarship.
Confidentiality of Data: The authors declare that they have followed the protocols of their work center on the publication of data from patients.
Patient Consent: Consent for publication was obtained.
Provenance and Peer Review: Not commissioned; externally peer reviewed.

References

1. Gatcombe HG, Assikis V, Kooby D, Johnstone PA. Primary retroperitoneal teratomas: a review of the literature. J Surg Oncol. 2004;86:107-13.
2. Wang RM, Chen CA. Primary retroperitoneal teratoma. Acta Obstet Gynecol Scand. 2000;79:707-8.
3. Gupta R, Reddy Y, Gupta R, Welsh JS, MAdani A, Hosmane NS. Primary benign cystic retroperitoneal teratoma in an asymptomatic adult. Biochem Biophys J Neutron Ther Cancer Treat. 2013;1: 5-7.
4. Lukanovic A, Patrelli TS. Retroperitoneal mass with ischiorectal fossa extension: diagnosis, clinical features and surgical approach. A literature review starting from a rare clinical case of primary retroperitoneal dermoid cyst. Eur J Gynaecol Oncol. 2010;31:709-13..
5. Mardi K, Sharma S, Sood S. Mature cystic teratoma involving the left adrenal gland with complete colonic wall formation in a 24-year-old female: A rare case report. Clin Cancer Invest J. 2015; 1; 47-9.
6. Lambrianides AL, Walker MM, Rosin RD. Primary retroperitoneal teratoma in adults. Urology. 1987;29:310-2.
7. Pantoja E, Llobet R, Gonzalex-Flores B. Retroperitoneal teratoma: a historical review. J Urol. 1976;115:520-3.
8. Pandya JS, Pai MV, Muchhala S. Retroperitoneal teratoma presenting as acute abdomen in an elderly person. Indian J Gastroenterol. 2000;19:89-90.
9. Khiyami A, McHenry CR. Retroperitoneal cystic teratoma masquerading as an incidentally discovered adrenal mass. Endocr Pract. 2011;17:e130-4. doi: 10.4158/EP11183.CR.
10. Bhatti A, Al-Hindi H, Azzam A, Amin T, Abu-Zaid A. Mature (benign) cystic retroperitoneal teratoma involving the left adrenal gland in a 22-year-old male: a case report and literature review. Case Rep Oncol Med. 2013;2013:610280. doi: 10.1155/2013/610280.
11. Yadav R, Kataria K, Balasundaram P, Karak AK. Mucinous cystadenocarcinoma arising in an ectopic kidney simulating a retroperitoneal dermoid cyst: a rare tumour presenting as a diagnostic dilemma. Malays J Pathol. 2013;35:95-8.
12. Chen JC, Khiyami A, McHenry CR. Retroperitoneal cystic teratoma masquerading as an incidentally discovered adrenal mass. Endocr Pract. 2011;17:e130-4. doi: 10.4158/EP11183.CR.
13. Castillo OA, Vitagliano G, Villeta M, Arellano L, Santis O. Laparoscopic resection of adrenal teratoma. JSLS. 2006;10:522-4.
14. Okulu E, Ener K, Aldemir M, Isik E, Irkkan C, Kayigil O. Primary mature cystıc teratoma mimickıng an adrenal mass in an adult male patient. Korean J Urol. 2014;55:148-51. doi: 10.4111/kju.2014.55.2.148.