CASE REPORT

Primary retroperitoneal teratoma in a young woman: a case report

Nadeem Iqbal,[1] Mahnoor Aitzaz Khan,[2] Ibrahim Khalil,[3] Adnan Ali,[4] Haris Aleem,[5] Nadeem Bin Nusrat[6]

Abstract

Teratomas are usually seen in gonads but they do occur in other extra gonadal regions such as sacrococcygeal region, mediastinum, head and neck, and retroperitoneum. Rarely in the retroperitoneal area, such tumours mostly develop in the pararenal area and usually on the left side. They have bimodal presentation at the age of six months and then in early adulthood. They originate from the germ cells that have failed to migrate to normal anatomical destinations. Many of such patients are diagnosed incidentally. Here, we report a case of symptomatic primary retroperitoneal mature teratoma in a young lady managed at Pakistan kidney and Liver Institute, Lahore.

Keywords: Benign Mature Cystic Teratoma; Kidney; Surgery; Retroperitoneal teratoma.

DOI: 10.47391/JPMA.7188

Submission completion date: 28-07-2022
Acceptance date: 07-01-2023

Introduction

Teratomas are deemed one of the most familiar congenital tumours and comprise tissues originating from pluripotent embryonic cells.[1] They are usually seen in gonads but they do occur in other extra gonadal regions such as sacrococcygeal region, mediastinum, head and neck, and retroperitoneum.[1-3] In the context of retroperitoneal area, the bulk of such tumours develop in the pararenal area and mostly on the left side as in this case. They have bimodal presentation at the age of six months and then in early adulthood.[2-4]

Studies relevant to such cases have estimated that teratomas in the retroperitoneal area occur in up to 4% of the retroperitoneal neoplasms.[2-4] They originate from the germ cells that have failed to migrate to normal anatomical destinations. Germ cells are totipotent cells that go through differentiation into different germ layers.[4] Many of such patients are diagnosed incidentally. However, at other times, symptomatic patients do come with abdominal pain, or a mass or lump in the abdominal region is observed in these patients.[4-5] It is pertinent to note that a majority of these mature teratomas are benign in nature; however, malignant degeneration may occur in few instances.[3-5] Here, we are reporting a case of symptomatic primary retroperitoneal mature teratoma in a 28-year-old female managed at our centre.

Case Report

A 28-year-old married woman was referred from a peripheral health centre to the Pakistan Kidney and Liver Institute Lahore, for an incidental finding of the left suprarenal mass in October 2021. On history taking, she did not complain of any symptoms in the past such as abdominal bloating, nausea/vomiting, or pressure effects leading to early satiety, pain, gastroesophageal reflux, or flank pain. There was no history of haematuria, lower urinary tract symptoms, weight loss or fever. She had negative history of abdominal surgery or genitourinary interventions. Her family history was negative for any genitourinary malignancies or other types of cancers.

She was stable vitally and afebrile. On abdominal examination, no tender areas were noted. There was no visceromegaly or abnormal lymph nodes anywhere in the body. Later on, an abdominal ultrasound study was undertaken which revealed a large left supra renal cyst. No other abnormalities were noted at the time of the study.

A CT of the abdomen and pelvis with and without contrast was obtained, which revealed a mass in the left supra-renal region (Figure 1A). It was a large cystic lesion (10cm in size), and had soft tissue density, fat density, and calcific components. These features were suggestive of mature cystic teratoma (germ cell tumour). Additionally, this lesion was effacing and mildly compressing the anterior left renal parenchyma, separate from the left adrenal gland, abutting the body and tail of the pancreas and the greater curve of the stomach. On computed tomography of the chest, no active pulmonary pathology was noted. Her biochemistry investigations such as renal function tests and serum electrolytes were in normal ranges. Tumour markers such as CEA (6.95 ng/mL) and

[1,6]Department of Urology, Pakistan Kidney and Liver Institute, Lahore, Pakistan, [2,3,5]Final Year MBBS Student, Shifa College of Medicine, Islamabad, Pakistan, [4]Final Year MBBS Student, Pakistan Kidney and Liver Institute, Lahore, Pakistan.

Correspondence: Nadeem Iqbal. Email: dr_nadeemiqbal84@yahoo.com
ORCID ID. 0000-0001-7154-9795

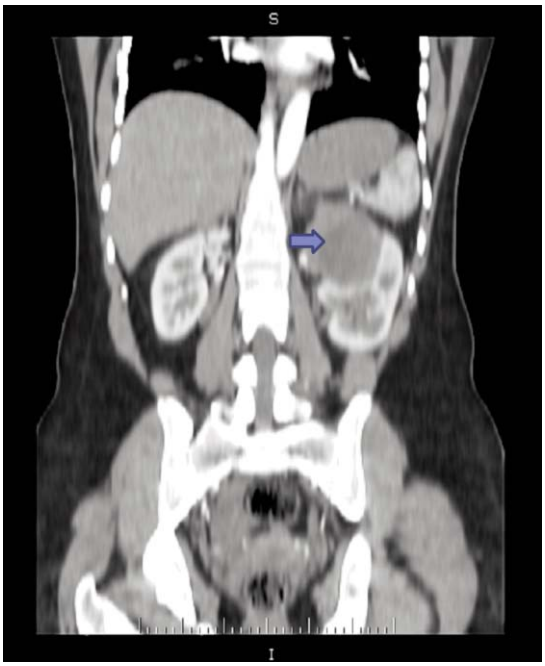

Figure-1A: Left suprarenal site of the lesion.

AFP (2.41 ng/mL) were normal. A non-functional suprarenal lesion was evidenced by lab and clinical picture (24 hour urinary metanephrines were negative). Computed tomographic guided biopsy was taken by interventional radiologists, that had an impression of a Mature Cystic Teratoma.

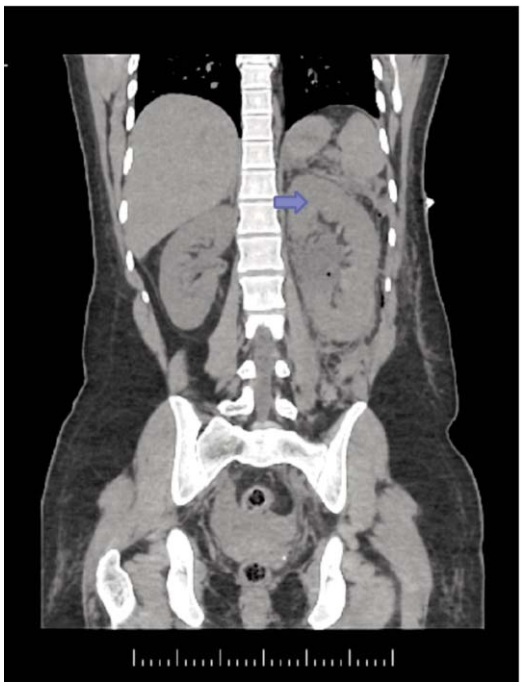

Figure-1B: Left suprarenal site of the lesion.

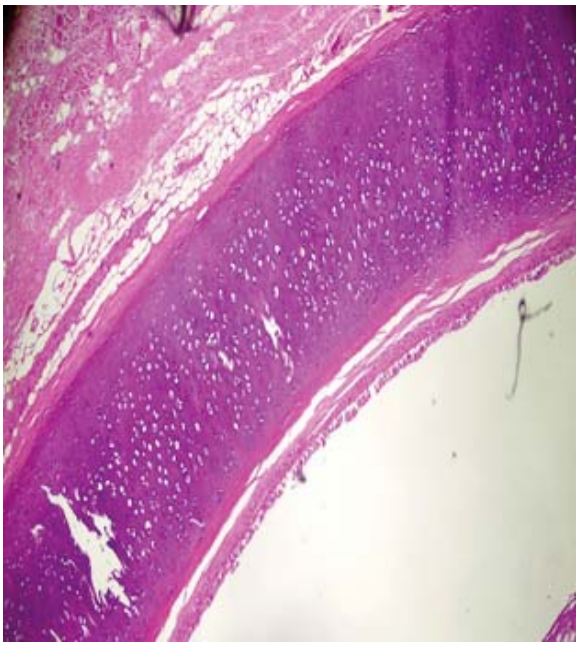

Figure-2A: 200x magnification showing respiratory epithelial lining with underlying cartilage resembling bronchial tissue.

The patient was scheduled for a left open suprarenal cyst excision. Per-operatively, there was an enlarged left supra renal mass which was badly adherent to the surrounding structures, including the bowel, pancreas, and spleen. It was an arduous task to separate it from the surrounding organs. The patient had a good post-operative recovery and was discharged on the fifth post-operative day.

The final histopathology report revealed multiple cystic areas. Some sections had lining of skin with sebaceous glands. While others had denuded lining with histiocytic infiltrate. Others were lined by respiratory epithelium (Figure 2A), urothelium, and columnar epithelium. The underlying tissue was variable; at places it was the smooth muscle tissue of gut with ganglion cells, while other areas showed neural and adipose tissue; adrenal tissue areas of inflammation with foreign body type giant cells reaction was also evident at places (Figure 2B). There were few dilated blood vessels lined by adipose tissue and giant cells. No evidence of malignancy was observed. No renal tissue was identified after extensive sampling. There was no evidence of malignancy or immature tissue in any of the sections examined. Hence, a final diagnosis of a mature cystic teratoma was made. On a follow-up CT scan after the surgery, the previously observed left supra renal mass was not visible (Figure 1B). The patient was doing well at her six-month follow-up.

The patient's consent was taken for publishing her case.

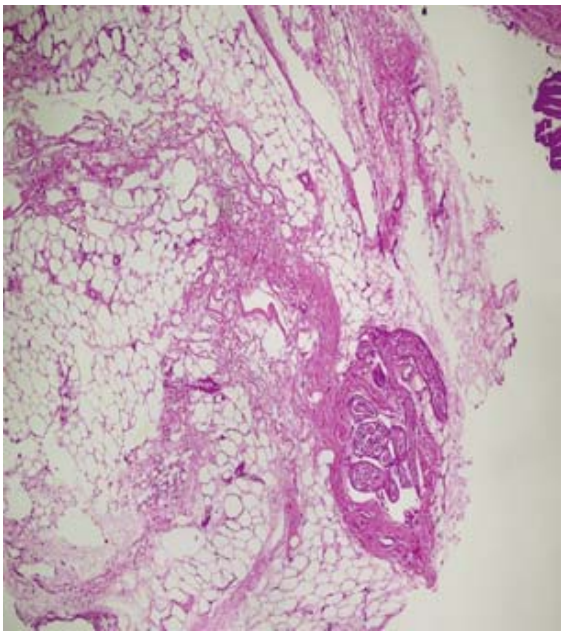

Figure-2B: 200x magnification showing adipose tissue with collection of nerves and blood vessels.

Discussion

Primary retroperitoneal tumours are rare tumours and originate from germ cells that have failed to migrate to their normal locations. Germ cells are totipotent cells which means that they have the potential to undergo differentiation into different germ layers.[3-4] It can be a difficult task to differentiate primary retroperitoneal tumours from gonadal malignancy. So, it is vital to exclude gonadal primary with the help of clinical examination and imaging before labelling a teratoma as primary retroperitoneal tumour.[2-5] Primary retroperitoneal tumour can manifest itself as a solitary lesion, while metastasic growths emanating from gonads may result in multicentric lesions. Primary retroperitoneal tumours are twice as common in females as compared to males.[4-5] Primary retroperitoneal tumours commonly occur in the retroperitoneum in proximity to the upper pole of the left kidney. Generally, they are asymptomatic, however, they may manifest symptoms such as abdominal pain or distension, nausea, vomiting, or compression.[5-6] Back pain, genitourinary symptoms, and lower extremity swelling may also occur.[5-7] In our case, the patient was asymptomatic.

In case of malignant teratomas (0.2%-2%) there is potential to metastasise to the lung and lymph nodes. Only a few cases may have raised serum AFP levels owing to lack of specific tumour markers for teratomas.[6-7] Radiological diagnosis of such tumours need contrast enhanced CT scan or by a magnetic resonance imaging scan. Imaging facilitates in outlining the tumour relationship with nearby structures, which is vital in mapping out the plan of the surgery. Definitive diagnosis is made after surgical resection of the lesion wherein the specimen is sent for histopathological diagnosis.

Macroscopically, cystic teratomas contain mature tissue and sebaceous material which are benign; whereas solid looking teratomas comprise immature embryonic tissue in addition to cartilaginous, bony fatty, and fibrous components, and are usually malignant.[5-7]

The main goal of the treatment plan is complete surgical resection. However, at times, complete surgical resection can be a daunting task owing to the intricate location of the tumour and proximity to crucial structures. In case of incomplete resection of the tumour, there is likelihood of malignant transformation of teratomas. Despite complete surgical resection malignant teratomas frequently recur. Five-year survival rates may reach 100% in case of teratomas as against 67% in cases of malignant tumours.[6-8]

It is worth noting that disease-free survival relies upon the thoroughness of tumour removal, hence, surgical resection in low-burden tumour can be of much value.[7-8] In case of residual mass left after surgery, the malignant transformation of a mature teratoma to a sarcoma or carcinoma takes place in almost up to six percent of patients.[7-10] Therefore, it is imperative to continue stringent follow-up of such patients with an annual CT imaging to pick silent progression of the residual disease.

Conclusion

Primary retroperitoneal teratomas are one of the rarest tumours being diagnosed in adults. Most of the time they are asymptomatic and are detected incidentally on imaging. However, surgical resection is required not only for definitive diagnosis but also for the treatment of the tumour. Most importantly, patients need stringent yearly follow-up for detection of recurrence.

Disclaimer: None.

Conflict of Interest: None.

Source of Funding: None.

References

1. Hoang VT, Trinh CT, Le TB, Le TK. Recurrence of retroperitoneal mature cystic teratoma in an adult: a case report. Radiol Case Rep 2019; 14: 692-6.
2. Peyvandi H, Arsan F, Alipour-Faz A, Yousefi M. Primary retroperitoneal mature cystic teratoma in an adult: A case report. Int J Surg Case Rep 2016; 28: 285-8.
3. Paradies G, Zullino F, Orofino A, Leggio S. Rare extragonadal teratomas in children: complete tumour excision as a reliable and

essential procedure for significant survival. clinical experience and review of the literature. Ann Ital Chir 2014; 85: 56–68.
4. Tojal A, Santos N, Vicente J, Carrilho N, Casimiro C. Retroperitoneal mature cystic teratoma involving the left adrenal gland in a 75-year-old male: a case report and literature review. J Surg Case Rep 2020; 2020: rjaa451.
5. Patil PS, Kothari P, Gupta A, Dikshit V, Kamble R, Kekre G, et al. Retroperitoneal mature cystic teratoma in a neonate: a case report. J Neonatal Surg 2016; 5: 15.
6. Sasi W, Ricchetti GA, Parvanta L, Carpenter R. Giant mature primary retroperitoneal teratoma in a young adult: report of a rare case and literature review. Case Rep Surg 2014; 2014: 930538.
7. Banthia R, Yadav P, Bharti A, Lal H. Mature cystic teratoma presenting as suprarenal mass. BMJ Case Rep 2020; 13: e237734.
8. Mathur P, Lopez-Viego MA, Howell M. Giant primary retroperitoneal teratoma in an adult: a case report. Case Rep Med 2010; 2010: 650424.
9. Tiu A, Sovani V, Khan N, Hooda S. Primary retroperitoneal mature cystic teratoma (dermoid cyst) in a 51- year-old male: case report and historical literature review. SAGE Open Med Case Rep 2017; 5: 2050313X17700745.
10. Rathod J, Patel S, Barot KS, Naik SH, Bhatt R, Chotaliya J. Massive primary retroperitoneal immature teratoma in an adult: A case report. Int J Surg Case Rep 2021; 81: 105775.