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CASE REPORT

Primary Retroperitoneal Teratoma Presenting As Diabetes Mellitus In A Child

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ABSTRACT

Primary retroperitoneal teratoma is a rare entity which has a distinctive imaging appearance. To our knowledge, this is the first reported case of teratoma causing diabetes mellitus in a 9-year old boy who was put on insulin. On clinical examination, there was a large tender mass with a well defined margin, caudally in the epigastrium. Ultrasound of abdomen revealed a huge retroperitoneal multiseptated predominantly cystic mass in the upper abdomen. Plain abdominal computed tomography revealed a large well defined cystic mass in the same region. Contrast enhanced abdominal CT revealed a mass with peripheral rim enhancement. It has multiple septa which also enhanced. Clefts containing fat were present adjacent to the septa. Laparotomy with tumour resection was performed. Pathology of the mass disclosed a retroperitoneal cystic teratoma.

Key Words: Ultrasound, CT scan, Retroperitoneal teratoma, Diabetes mellitus.

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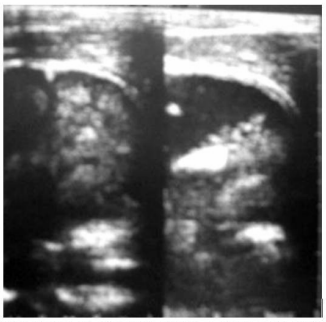
Introduction

A teratoma is a primary germ-cell neoplasm that is composed of well-differentiated tissue derived from embryonic germ cell layers [1]. Primary retroperitoneal teratomas are extremely rare tumours, representing less than 10% of all primary retroperitoneal tumours [2]. It is a benign neoplasm that can occur in many locations, but are most commonly seen in the gonads, head and neck, the anterior mediastinum, the sacrococcygeal area, the retroperitoneum, and the central nervous system (CNS). Teratomas have also been reported throughout the gastrointestinal (GI) tract and associated organs, including the caecum, rectum, and pancreas. Teratomas have been described using all imaging modalities, but the specificity for diagnosis of fat and calcifications makes CT the modality of choice. We present this case because of its rarity in causing diabetes mellitus.

Case Report

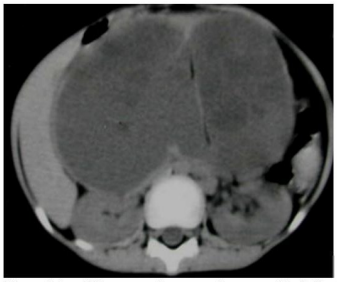
A 9-year-old boy presented with upper abdominal swelling and intermittent abdominal pain since 20 days. There was a history of diabetes mellitus being diagnosed two years earlier, and of being put on insulin. There was no history of weight loss and jaundice. The external genitalia including testis were normal. Physical examination revealed a large tender mass with a well defined margin, caudally in the epigastrium. Abdominal ultrasound showed a well defined large retroperitoneal heterogeneous multiseptated predominantly cystic mass, measuring 11.0 cm x 6.0 cm,

located in the upper and mid abdomen. The solid components were seen attached to its anterior wall. No calcifications were noted within it [Table/Fig 1].



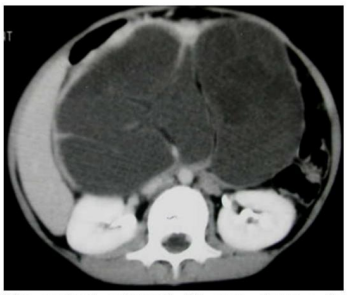
(Table/Fig 1) Ultrasound of abdomen shows a large well defined heterogeneous multiseptated predominantly cystic mass measuring 11.0 cm x 6.0 cm in size located in the upper and mid abdomen. The solid components were seen attached to its anterior wall. No calcifications were noted within it.

Abdominal CT confirmed a huge well defined heterogeneous cystic mass in the central abdomen extending from epigastrium till the infrarenal level [Table/Fig 2].



(Table/Fig 2) The plain CT scan shows a large well defined heterogeneous predominantly cystic mass in central abdomen extending form epigastrium to infrarenal level. It measures 15.0 cm x 9.0 cm in size. No calcification noted within the mass.

Contrast enhanced abdominal CT revealed a mass with peripheral rim enhancement. It has multiple septa which also enhanced. A few lobulated hyperdense foci were also seen within the mass. The multiple clefts of hypodensity (HU -20 to -40) of fat value adjacent to the septa, were noted. There was no evidence of calcification [Table/Fig 3].



(Table/Fig 3)The contrast enhanced CT scan shows a large well defined heterogeneous predominantly cystic mass with peripheral rim and multiple septal enhancement in central abdomen extending from epigastrium to infrarenal level. A few lobulated hyperdense foci are also seen within the mass. There is evidence of multiple cleft of hypodensity(HU of –20 to –40) with fat value adjacent to the septa. There was no calcification noted within the mass.

A diagnosis of retroperitoneal teratoma was made. MR imaging was not done in our case. Ultrasound guided fine needle aspiration cytology smears showed plenty of inflammatory cells, cyst macrophages, debris, few multinucleated giant cells, and nucleated and anucleated squamous cells. A diagnosis of infected cyst or teratoma was made.

Exploratory laparotomy revealed a huge retroperitoneal, well encapsulated teratoma of 11.0 cm in diameter. The cyst was located posterior to the duodenum and pancreas, and extended from the right upper to the left upper quadrant of the abdomen. All surrounding viscera, including pancreas, stomach and duodenum were pushed anteriorly, and the portal vein and the hepaticoduodenal ligament were displaced laterally. Excision of the cyst was performed, and pressure on the pancreas and other surrounding structures, was relieved.
Histological examination demonstrated an encapsulated grayish brown cystic mass containing yellowish white cheesy material, hair and cartilage. The postoperative period was uneventful, and the patient was well when discharged. The patient's random blood sugar returned to normal limits, he does not have to take insulin, and is doing well at follow-up.

Discussion

Teratomas are tumours that contain tissue from the germ layers, that is, the ectoderm, the mesoderm, or the endoderm. These

tumours are thought to arise from pluripotent embryonal cells [1]. Retroperitoneal teratomas are rare tumours, and constitute less than 10% of all primary retroperitoneal tumours [2]. Retroperitoneal teratomas are more common in childhood than at any other time, and they are a rare entity in adults [3],[4]. The incidence of retroperitoneal teratoma in females is twice than in males. Teratomas can occur in any region of the body and in any organ, but they are most commonly found in the paraxial and midline locations [5]. The ovaries and testes are the most common sites of teratomatous growth. Less often, they are found in the sacrococcygeal region, in the head near the pineal body, in the neck near the thyroid, in the anterior mediastinum and in the retroperitoneum. Gonadal teratomas are most often seen in postpubertal patients, whereas extragonadal teratomas usually occur during infancy and in early childhood. The chances of malignant transformation are more common in adults than in children (26% vs 10%) [3],[6],[7].

Abdominal radiography may demonstrate a mass with fat, either with calcification or with bone. Similarly, ultrasound shows uncomplicated fluid and calcification, but it cannot reliably differentiate fat from other soft tissue components [8].

The most characteristic ultrasonographic appearances of mature retroperitoneal teratoma, are heterogeneous lesions containing a well-circumscribed fluid component of variable volume, adipose tissue or sebum in the form of fat-fluid level, and calcification. These findings are well differentiated by CT scan than by ultrasound [8].

Teratomas have been described using all imaging modalities, but the specificity for diagnosis of fat and calcifications makes CT scan the modality of choice. The characteristic signal intensity of fat on MR imaging is high on T1-weighted images, while water demonstrates low signal intensity on T1-weighted images and high signal intensity on T2-weighted images. In addition, MRI may help to delineate the relationship between the tumour and adjacent structures by means of multiplanar images, which is not well demonstrated by the CT scan.

H. Liu et al. has reported that benign teratoma often expands and presses against, rather than encases, the surrounding structures [9].

Retroperitoneal teratomas are usually asymptomatic. When compression of the surrounding structures occurs, patients may have abdominal distension and pain, and nausea and vomiting [10].
In our case, the pressure on the pancreas might have lead to reduction of the production of insulin, thus causing diabetes mellitus.
To our knowledge, no report of teratomas causing diabetes mellitus was found in the English literature.
The postoperative period was uneventful, and the patient was well when discharged, with normal blood sugar, and insulin was stopped.

Conclusion

We present a rare case of diabetes mellitus caused by pressure of a retroperitoneal teratoma on the pancreas.

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