Case Report

Large retroperitoneal dermoid cyst in a 55 year old lady: a rare case report

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ABSTRACT

Retroperitoneal cysts are usually rare asymptomatic lesions but may present with unspecific symptoms depending on the size, location and complications such as hemorrhage, infection, or rupture. A 55-year-old lady presented with complaints of non-obstructed umbilical hernia. CT scan of abdomen and pelvis was done suggestive of a large cystic non-enhancing thin walled lesion of size 27×15 cm in right lumbar region. Intraoperatively, there was a large mesenteric cyst from retroperitoneum of size 25×15 cm, extending from right iliac fossa up to right hypochondrium. A confirmatory diagnosis of dermoid cyst was made following histopathology. Retroperitoneum is a rare site for dermoid cyst. Retroperitoneal cysts which develop within the retroperitoneal space are rare intra-abdominal tumours with an incidence of 1 per 1,40,000. The incidence of recurrence for retroperitoneal cysts is higher than with other forms of cysts because their proximity to major blood vessels and other organs makes them difficult to completely excise. Surgical resection is indicated to establish a diagnosis and prevent eventual complications. Complete excision of tumour is necessary due to the risk of malignancy. After complete surgical resection the 5 years survival rate is nearly 100%.

Keywords: Dermoid, Retroperitoneal, Teratoma

INTRODUCTION

Mesenteric and retroperitoneal cysts are usually rare asymptomatic lesions. Mature cystic teratomas, also known as dermoid cysts, are neoplasms composed of tissues from at least two of the three germ layers.1,2 Epidermoid cysts that develop in the retroperitoneal space are quite rare.2 They are usually discovered incidentally during investigation of irrelevant illnesses or during routine abdominal ultrasonography.3 Retroperitoneal cysts are mostly asymptomatic but may present with unspecific symptoms depending on the size, location and complications such as hemorrhage, infection, or rupture.4 They are asymptomatic in one-third of patients.5,6 Contrast-Enhanced Computed Tomography (CECT) of the abdomen and pelvis is used to confirm the diagnosis.7,8 Surgical excision of the whole cystic mass is the choice of treatment. In this case report we present a benign cystic retroperitoneal dermoid cyst in the right lumbar region in a 55-year-old healthy female patient who presented with a 2-month history of asymptomatic non-obstructed umbilical hernia.

CASE REPORT

A 55-year-old lady presented with chief complaints of swelling over the umbilical region since two months associated with vague dull-aching pain. The swelling increased in size on exertion and reduced spontaneously on lying down.
There were no associated complaints of pain, fever, vomiting, constipation, diarrhoea, passage of blood in urine or stools, breathlessness, loss of weight or appetite and no history of other swellings in the body.

There was history of previous surgery done for right sided complex ovarian cyst 15 years ago, details of which were unavailable. Patient was not a known case of diabetes mellitus, hypertension, heart disease, asthma or any other major illnesses.

She was not a known alcoholic, smoker or drug addict. There is history of attaining menopause 10 years ago and bears three children presently. On physical examination, there was evidence of a reducible uncomplicated umbilical hernia with a defect of three centimeters. On deep palpation, diffuse fullness was felt in right lumbar region extending up to right iliac fossa, non-tender and firm to touch. Per vaginal and per rectal examination had no abnormal findings.

Routine laboratory investigations were within normal limits. She underwent an ultrasonography of abdomen and pelvis that was suggestive of a large complex ovarian cyst, most likely dermoid cyst of size $20.5 \times 10.5$ cm with a non-obstructing umbilical hernia and evidence of cholelithiasis.

Following differential diagnosis following CECT were made including large ovarian cyst, mesenteric cyst CA-125 - 9.3 [within normal range].

Following ultrasonography, a contrast enhanced Computed Tomography [CT] scan of abdomen and pelvis was done suggestive of a large cystic non-enhancing thin walled lesion of size $27 \times 15$ cm in right lumbar region extending up to right iliac quadrant, crossing the midline, with no mass effect on ureters, no solid component or fat within the lesion (Figure 1 to 4).

Figure 1: CECT image of abdomen and pelvis showing location of cyst.

Figure 2: CECT image of abdomen and pelvis showing extent of cyst.

Figure 3: CECT image of abdomen and pelvis showing cyst with relation to surrounding structures.

After complete anaesthesia fitness and workup, patient underwent an elective exploratory laparotomy.
Intraoperatively, there was evidence of a large thin walled mesenteric cyst arising from retroperitoneum of size 25×15 cm, extending vertically from right iliac fossa upto right hypochondrium and xiphisternum with no clear point of origin.

Cyst was carefully dissected and separated from lateral and posterior abdominal wall. Retroperitoneal vessels and right ureter were identified and left intact. Cyst wall was incised and opened. There was evidence of abundant greyish white sticky pultaceous material with approximately 50 ml of serous fluid. Cyst was completely evacuated (Figures 5 to 9). Total weight of cyst contents was four kilograms.

Figure 4: Removal of pultaceous material from cyst.

Figure 5: Extent of cyst wall.

Figure 6: Cyst wall after removal of all pultaceous content.

Figure 7: Gross appearance of pultaceous content removed from cyst weighing 4 kilograms.

Figure 8: Thinned out cyst wall.
Figure 4 to 8 shows intraoperative pictures of dermoid cyst excision.

The thin wall of the cyst was separated completely, excised and sent for histopathological examination. No enlarged lymph nodes were found. Peritoneal lavage followed by peritoneal drain insertion was done. Umbilical hernia defect was also closed by anatomical repair. Patient tolerated the procedure well.

On histopathological analysis (Figure 9-11), gross examination cyst wall with pultaceous material was noted. Microscopic examination, cyst wall lined by stratified squamous epithelium with underneath hair follicles and sebaceous glands present.

On the basis of gross and microscopic examination, a confirmatory diagnosis of dermoid cyst was made. Cyst fluid showed no evidence of any pathogens. Postoperatively intraperitoneal drain was removed on the sixth day. Patient was discharged on the eighth day after an uneventful postoperative course.

DISCUSSION

Dermoid cyst is a cystic teratoma that includes a mix of developmentally mature and solid tissues. Retroperitoneum is a rare site for dermoid cyst. Retroperitoneal cysts which develop within the retroperitoneal space are rare intra-abdominal tumours that originate from an embryologic error during development. Other rare sites are mediastinum, sacrococcygeal region and central nervous system.

Adult retroperitoneal dermoid cyst commonly affects females (15-40 years of age). There is 25% chance of malignancy. Differential diagnosis of retroperitoneal cysts includes retroperitoneal sarcoma, hydatid cyst, ovarian tumor, mesenteric cyst, and renal tumors.

Contrast enhanced computed tomography scan is the investigation of choice for diagnosis and to assess the extent of the cyst preoperatively. Extensive use of computed tomography for diagnosis of abdominal and retroperitoneal malformations has enhanced the detection rate for these lesions.

Serum tumor markers (AFP, CEA, CA 19-9) level should be measured in suspicious patients of malignancy. They can be helpful in diagnosis, monitoring disease and detecting relapse. Definitive diagnosis is made by histopathological examination. Whenever possible symptomatic cyst should be excised completely while adjacent vital structures should be preserved. Complete resection of the tumor is curative. Marsupialization or draining of the cyst usually results in a recurrence. Spillage of cyst contents may lead to infection or recurrence.

Confirmatory diagnosis of the lesion is often not possible from ultrasonography and CT scan. Since it is impossible to entirely rule out cystic malignancies, surgical resection and histopathologic examination is required for definitive diagnosis.

The sole symptom reported by our patient was vague abdominal pain associated with umbilical hernia, following which a differential diagnosis of a large mesenteric cyst / complex ovarian cyst was done on ultrasonography of abdomen and confirmed with contrast enhanced CT scan for which patient was electively operated. Intraoperatively a rare finding of a large retroperitoneal cyst was observed and completely excised with its pultaceous material. Histological examination revealed features of a dermoid cyst. The lesion was adjacent to retroperitoneal great vessels and right ureter, which required special attention during operation and was technically demanding. Limitations of this study were: In case of colorectal malignancies, the stage wise distribution of the disease (in percentage) observed by other authors is shown.

CONCLUSION

Retroperitoneal location of an epidermoid cyst, outside the presacral region is extremely rare. Diagnostic imaging is helpful in making the diagnosis, although histopathologic examination is definitive in the final analysis of the lesion. Therefore, surgical resection is
indicated to establish a diagnosis and prevent eventual complications, such as hemorrhage, infection or rupture. Complete excision of tumor is necessary due to increased risk of malignancy.

After complete surgical resection the 5 years survival rate is nearly 100%. Diagnosis requires a high degree of suspicion especially when clear diagnosis on CT scan cannot be made regarding the origin and extent of such cysts.

Surgery is the gold standard for the diagnosis and treatment of retroperitoneal dermoid cysts. Successful treatment of a benign retroperitoneal dermoid cyst depends on appropriate diagnosis, careful operative technique and adequate management of the underlying pathology. This study shows the unique presentation and advances in knowledge regarding management of such rare cases.

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REFERENCES
