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A Case of ruptured dermoid

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Abstract : Teratomas are bizarre neoplasms derived from embryonic tissues that are typically found only in the gonadal and sacrococcygeal regions of adults. Retroperitoneal teratomas are rare and present challenging management options. Rupture of retroperitoneal dermoid is a rare complication. We report here a rare case of a histologically unusual extragonadal retroperitoneal dermoid in a male patient with rupture detected on computed tomography, MRI during the workup of abdominal pain.

Keyword :Extragonadal teratoma, retroperiteum, Rupture, histological analysis

1. Introduction:

Dermoid cysts (benign cystic mature teratomas) are congenital tumors consisting of derivatives from the ectoderm, endoderm and mesoderm germ cell layers. A teratoma is considered to be a nonseminomatous germ cell tumour and is typically located in either the sacrococcygeal region or in the gonads. We present a rare case of a massive retroperitoneal tumor with rupture in a 27 -year-old patient, which was treated successfully with surgical resection.

2. Case History:

A 27 years male Presented with abdominal pain, abdomen distension for one month duration with no associated history of vomiting, haematemesis, malena or altered bowel habit, loss of appetite and weight, jaundice. His general physical examination revealed averagely built & nourished, active male with no jaundice, pallor, cyanosis, clubbing and lymphadenopathy. Abdominal examination revealed a soft, diffuse, intra-abdominal, non-tender, vague palpable mass in umbilical and right hypochondrial region. There was no organomegaly. Bowel sounds were normal. Rest of the physical examination was unremarkable. Spine examination was normal with no spine tenderness or deformity. Laboratory investigations were within normal limits. USG abdomen showed 15 * 11 cm well defined heteroechoic mass lesion in midline. Also multiple heteroechoic lesions noted in right hypochondrial and peripancreatic region with largest measuring 5.3 * 5.4 cm in epigastric region. (FIGURE 1) Bilateral hydroureteronephrosis noted.

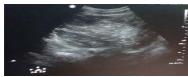


FIGURE 1 USG

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CECT abdomen showed 22 * 9.9 cm well defined fat (HU -35) filled lesion with calcification (HU +1000) and soft tissue in lower abdomen and pelvis. Multiple similar lesions without calcification and soft tissue also noted in peritoneum largest measuring 7.8 * 6.6 cm.(FIGURE 2&3)

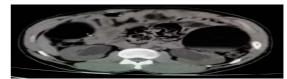


FIGURE 2 CT



FIGURE 3 CT

MRI abdomen revealed shows evidence of well defined fat containing lesion measuring 3.5 *2.1 cm in segment 8 of right lobe of liver. Evidence of large well defined heterointense lesion in the pelvis measuring 10.2(anteroposterior)*13.1 (transverse) 20.4(craniocaudal) predominantly containing fat and soft tissue component (with CT correlation, calcification also noted within the lesion).The lesion is seen compressing the bladder causing dilatation of ureteral and pelvicalyceal system of both kidneys.

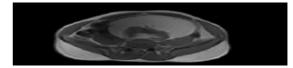


FIGURE 4 - T1 MRI



FIGURE 5 - T2 MRI

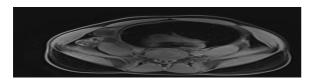


FIGURE 6 - FAT SUP MRI

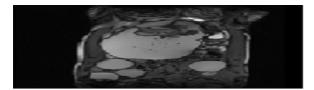


FIGURE 7 - CORONAL MRI

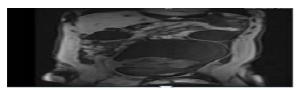


FIGURE 8 - FAT SUP CORONAL MRI

With the above findings, a provisional diagnosis of dermoid cyst pelvis with possible rupture and peritoneal seedling was made. Aspiration of the lesion showed yellow color fluid with hair. (FIGURE 9)



FIGURE 9 ASPIRATE

2.1 Operative findings:

Patient was taken up for laparotomy and excision of dermoid cyst under epidural anesthesia.

2.2 Intraoperative findings:

Multiple loculated cystic lesions in the peritoneal cavity with largest measuring 20*25 cm extending from pelvis to 6 cm above umbilicus. Cyst wall adhesion with greater omentum, bladder, rectum, right ureter noted with bilateral hydroureter. The cyst is separated from adhesions, punctured and contents are aspirated and removed. Contents include skin, hair, pultaceous material. Hemostasis achieved, DT placed and fixed to skin. Wound closed in layers.(figure 10 & 11) Specimen sent for histopathological examination.



FIGURE 10 - INTRA OP



FIGURE 11

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2.3 HPE examination:

HPE examination of cyst wall shows stratified squamous epithelium with lamellar keratinization, hair follicles, foci of calcification lobules of mature fat with focal areas showing mucinous glands and respiratory epithelium and single foci of glial tissue and mature cartilage.

Feature consistent with mature cystic teratoma.

The patient was followed up till 6 months & is doing good. Follow up ultrasound showed no evidence of recurrence.

3. Discussion:

The word teratoma is derived from the Greek word teraton, meaning monster, and was used initially by Virchow in the first edition of his book on tumours, which was published in 1863. Since mature cystic teratomas are composed of all three germ cell layers, the term 'dermoid' is a misnomer. Mature cystic teratomas are thought to arise from primordial germ cells. Macroscopically there are two types: Cystic teratoma: usually benign, contains yellowish liquid material, composed of fully developed tissue. Solid teratoma: generally malignant, have a varied aspect, formed of fibrous, fatty, cartilaginous and bone tissue consists of immature embryonic tissue . Retroperitoneal dermoid is rare and usually develops, in childhood. Most of the retroperitoneal tumors in childhood are cystic and benign. The order of frequency of teratoma localization is : Ovarian, Testicular, Anterior Mediastinal, with retroperitoneal localisation occurring least of all. Symptoms of Retroperitoneal Teratoma RPT) are variable. Spontaneous rupture of cystic retroperitoneal teratomas is a rare occurrence probably because of the thick encasing capsule. Rupture or perforation of the cyst may give rise to peritonitis. Peritonitis resulting from a chronically leaking dermoid cyst is characterized by multiple small white peritoneal implants, diffuse or focal omental Infiltration and inflammatory masses involving the omentum and bowel and dense adhesions and variable ascites that simulate carcinomatous or tuberculous peritonitis. Clement et al and Achtari et al reported chemical peritonitis following cystic fluid spillage. The patients needed further surgeries to treat the complications. The incidence of chronic granulomatous peritonitis after rupture or leakage of cystic fluid is also extremely rare. Intra-abdominal peritoneal seedlings, adhesions and/or masses are frequent sequelae. In most such cases, abdominal seedlings are essentially of mature neuroglial elements and long-term survival rate is good. Awareness is the key to diagnosis and appropriate management. The treatment of choice for retroperitoneal dermoid cvsts is complete surgical excision. The main reason for this is the worsening symptoms associated with the increasing mass effect as the tumour continues to grow. Malignant change has also known to occur.

Testicular ultrasound is necessary to rule out a coexisting testicular germ cell tumor in male patients. This is a necessary step since 50% of men with retroperitoneal tumors also have testicular carcinoma in situ, a precursor for testicular germ cell tumors. Long-term care also involves advising patient to follow up with annual CT to detect relapse at an asymptomatic phase. **4.Conclusion**:

Teratomas have distinctive imaging characteristics that allow for their easy identification, but histological examination is considered the current standard for definitive diagnosis and assessment of malignant potential. Extragonadal retroperitoneal dermoid should also be considered as a differential in a male presenting with abdomen pain. Rare complication of rupture should be considered. Optimal management avoids unnecessary repeated major surgeries and provides a good long-term outcome with minimal complications.

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