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Case Report:Primary retroperitoneal mature cystic teratoma (dermoid cyst) in the right lumbar region of a 45-year-old female

Bhushan M. Warpe[1], Shweta Joshi-Warpe[1],Netaji R. Patil[2], Rahul Sakpal[3] , Abhay Y. Desai[4]

Associate Professors' (Pathology)[1], Consultant (Radiology)[2], Assistant Professor (Pathology)[3], Associate Professor (General surgery)[4]at B.K.L. Walawalkar Rural Medical College & Hospital, Shree-kshetraDervan, District-Ratnagiri, 415606, State-Maharashtra, India

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ABSTRACT:A 45-year-old female patient presented with chief complaints of swelling in the right lumbar region, noticed since two years and pain since two months. Per abdomen examination confirmed a lump of around 7x6 cm in size
in the right lumbar region. Contrast enhanced computed tomography (CECT) abdomen revealed a 7.5x6.5x3 cm well-defined, heterogeneously hypo-attenuating cystic mass with focal areas of calcifications and fat attenuation in retroperitoneum. The patient was taken up for exploratory laparotomy and a tumor was found in the retroperitoneum and was excised with due care. Histopathological examination was confirmatory of Primary retroperitoneal mature cystic teratoma (dermoid cyst). This is an uncommon benign tumour that has been suggested to originate from embryonic error during developmental stages. The postoperative stay was uneventful.

Keywords: Dermoid, Retroperitoneal cysts, Teratoma, Lumbar

I. INTRODUCTION:

Retroperitoneal mature cystic teratomas (RMCT) are called dermoid cysts when they are composed of well-differentiated derivations from at least two of the three germ layers (ectoderm, mesoderm, endoderm) [1]. 10%-20% of RMCT occur after thirty years of age. Adult RMCT commonly affects females aged, 15-40 years [2].
On contrast enhanced Computed Tomography (CECT) scan, the presence of hypo-attenuating fat within the cyst and calcifications in the cyst wall is highly suggestive of cystic teratoma [1]. Surgical excision of the cystic mass is the treatment of choice [1,2]. Histopathology (HPR) is the 'gold standard' for definitive diagnosis. In this case report, we discuss a RMCT in the right lumbar region of a middle aged female.

II. CASE REPORT:

A 45-year-old perimenopausal lady presented with complaints of progressive swelling over the right lumbar region noticed since last two years, associated with vague dull-aching pain since past two months. She was previously operated with Caesarean section (LSCS), 20 years back. Patient did not have history of addiction, diabetes mellitus, hypertension, asthma or any other majorillness.

On deep palpation, diffuse fullness with firm consistency was felt in right lumbar region, measuring 7x6 cm and was advised CECT. Her cervical PAP smears showed bacterial vaginosis infection.
Routine laboratory investigations were within normallimits. Her serum markers were within normal levels. CEA: 1.2 ng/ml, CA-125: 23.2 U/ml, AFP: 7.9ng/ml (higher part of normal range), CA19-9: 22.3 U/ml, β-HCG: 2.3 mIU/ml.

She underwent a CECT scan of abdomen and pelvis which was suggestive of a RMCT (Fig. 1C). CECT revealed a 64x54x85 mm sized well-defined, heterogeneously enhancing, oval-shaped cystic-mass at right lumbar region. The growth showed patchy calcification within the growth and had peripheral wall calcification. It was abutting the right-kidney and right-psoas muscle. The growth showed a hypodense area of -60 Hounsfield-units (fat component). It displaced the right ureter laterally.

After workup, patient underwent an elective exploratory laparotomy. Intra-operatively, the cyst was carefully dissected and separated from lateral and posterior abdominal wall. Retroperitoneal vesselsand right ureter were spared. Cyst wall was removed intact and sent for histopathological H&E examination. The RMCT cyst measured 7.5x6.5x3 cm and had retroperitoneal fat on the outer cyst surface giving it an irregular appearance (Fig. 1A). On cut section, the cyst was thick-walled firm and thin-walled at places which was filled with thick yellow fluid. It

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was multi-loculated with largest cyst measuring 6cm diameter and smallest cyst measuring 1cm diameter. There were grey-white areas separating the locules. Yellowish areas were seen, denoting fat with greasy texture at places. No bone, tooth or hairs were noted (Fig. 1B). No enlarged lymph nodes were found. No ascites noted. Cyst fluid showed no evidence of any pathogens on culture studies.

Microscopic examination: Cyst walls were variably lined by pseudo-stratified ciliated columnar (respiratory) epithelium (Fig. 2B), low cuboidal to glandular columnar epithelium (Fig. 2C). The sub-epithelial fibrous tissue shows sweat glands, mucus glands, nerve bundles, muscle, adipose tissue and blood vessels with chronic mononuclear infiltrate (Fig. 2A, 2D). No stratified squamous epithelium or hair follicles or sebaceous glands were seen in our case.

On the basis of gross and microscopic examination, a confirmatorydiagnosis of primary retroperitoneal mature cystic teratoma (dermoid cyst) was made.

Postoperatively intra-peritoneal drain was removed on the sixth day. Patient was discharged on the seventh day after an uneventful postoperative course and advised follow-up.

III. DISCUSSION:

In 1769, retroperitoneal tumor (RT) was first described by Morgagni [3], while urography was first used to diagnose RT in 1937 [4]. A 82-year-old female was the oldest reported case with RT [5].

In 1987, RT was first diagnosed onCT scan and with the advent of MRI, cases are getting discovered incidentally nowadays.

The differential diagnosis of a primary retroperitoneal mass could be of two types: solid and cystic.

Cystic retroperitoneal lesions are rare intra-abdominal tumors with an incidence of 1 per 1,40,000 cases in surgery section. They can be either neoplastic or non-neoplastic. Refer Table 1 [6-8].

Table 1: Retroperitoneal masses – differential diagnoses[6-8]

Non-neoplastic cystic RT	Neoplastic cystic RT	Fat-containing primary retroperitoneal masses	Primary retroperitoneal masses with calcification
Mature teratoma	Lymphangioma	Teratoma	Teratoma
Cystic mesothelioma	Müllerian cyst	Lipoma	Malignant fibrous histiocytoma
Mucinous cystadenoma	Epidermoid cyst	Well-differentiated liposarcoma	Dedifferentiated liposarcoma
	Lymphocele	Dedifferentiated liposarcoma	Ganglioneuroma
	Pancreatic pseudocyst		Paraganglioma
	Hematoma		
	Urinoma		

Immature teratomas are usually solid masses comprised of undifferentiated tissues which may include fat and calcification. Mature teratomas are mainly cystic masses comprised of mature tissue elements belonging to ectoderm, mesoderm and endoderm components. They are accepted to arise as vestiges of the wolffian and müllerian ducts or from pronephric/mesonephric tubules [9].

The anterior mediastinum, retroperitoneum, presacrum, coccygeal region, intracranium, neck and abdomen are the extragonadal sites for RMCT [10].

The risk of malignancy in untreated RMCT increases with age, male sex, presence of immature tissues and solid components. Many serum markers such as AFP, CA-125, CA 19-9, CEA, β-HCG were studied in RT, but these were more associated with malignancies arising from the teratoma [1,10]. Elevated AFP levels were found in 50% of immature teratomas and 6% in mature teratomas [9,10]. These markers are evaluated pre-operatively to exclude other germ cell tumors,

Plain abdominal radiographs are first-line investigations to diagnose RMCT but are largely non-specific.Ultrasound has limited sensitivity in detecting RMCT which makes CECT-scan the imaging modality of choice. CECT-scan findings indicative of primary RMCT include well-circumscribed fluid component, hypo-attenuating fat and calcifications which were found in our patient [1,2,4].

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Surgical resection followed by histopathologic examination is required for definitive diagnosis of RMCT [1,2].Laparoscopic surgery is the preferred approach, especially when the cyst is well-circumscribed, smaller on imaging studies [9].

With excision of RMCT, prognosis is excellent. However, if left untreated, condition complicates due to obstruction of nearby viscera or metastasis from malignancies arising from RT. Post-operative follow-up is recommended to detect early recurrence.

IV. CONCLUSION:

CECT is the imaging modality of choice for RMCT. Complete excision of tumor is the treatment of choice. Histopathology is the gold standard in definitive diagnosis. IHC is not mandatory to diagnose RMCT.

Conflict of Interests:
The authors declare that there is no Conflict of Interests.

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FIGURES:

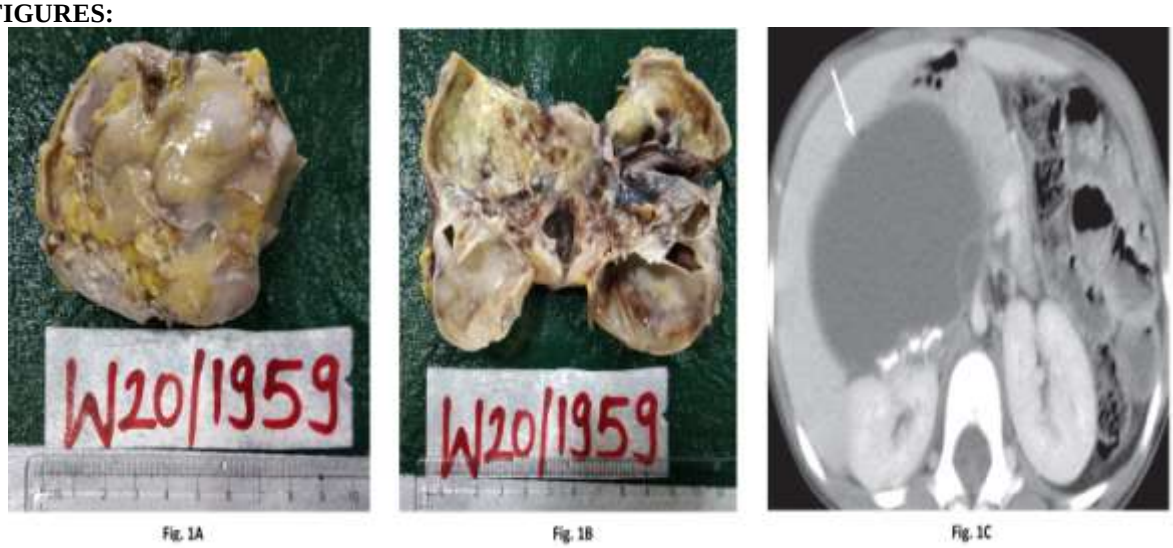

Fig. 1: Gross photograph: Irregular external cystic surface with adherent fat (1A); Cut section of cystic retroperitoneal tumor showing grey-white area and variably thickened cyst wall with fat within a cyst locule (1B);
CECT abdomen shows a well-defined hypoattenuating mass with internal septa and calcifications in the right lumbar region abutting the right kidney(arrow) (1C).

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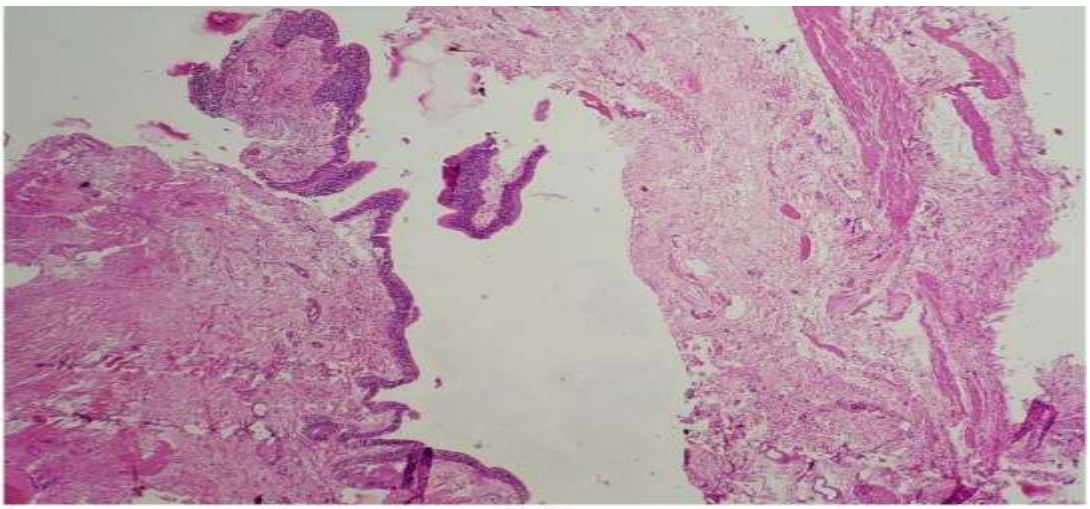

Fig. 2A

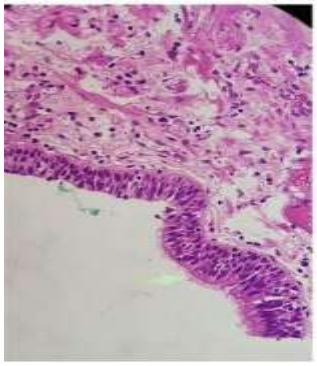

Fig. 2B

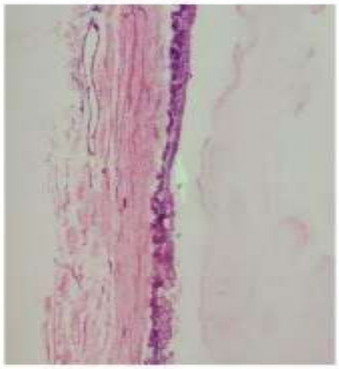

Fig. 2C

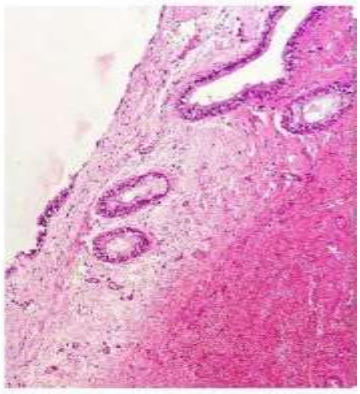

Fig. 2D

Fig. 2: Photomicrographs of Primary retroperitoneal mature cystic teratoma-2A showing cyst wall with variably layered lining epithelium, subepithelial tissue containing muscle, fat, glands, blood vessels, nerve bundles (H&E, ×40); 2B showing cyst wall lined by pseudostratified ciliated columnar epithelium with subepithelial mononuclear infiltrate (H&E, ×400); 2C showing lining columnar epithelium with surrounding extracellular mucin (H&E, ×400); 2D showing subepithelial benign glands in fibrovascular stroma (H&E, ×400).