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A Rare Case Of Carcinoid Tumour In Benign Cystic Teratoma Of Adrenal Gland

Dr.R.Mahalakshmi[1],Dr.D.Dhanya Charitha[2], Dr.G.Chandana[3]
1.Professor And Head Of The Department, Dept.Of General Surgery,Rangaraya Medcal College,Kakinada,Andhra Pradesh,India.
2.Postgraduate, Rangaraya Medical College ,Kakinada,Andhra Pradesh,India.
3ipostgraduate,Rangaraya Medical College ,Kakinada,Andhra Pradesh,India.

Abstract:
Teratoma is a solid tumor composed of different histological components and usually involves more than 2 germ layers. Most common sites are gonads, sacrococcygeal, mediastinal, retroperitoneal. We report a case of 26yr old female patient who presented to the OPD with the chief complaint of pain and lump in the right upper abdomen gradually progressing in size to occupy the entire abdomen over a duration of 10 days. CT abdomen revealed a large hypodense lesion with wall calcifications and thin septa – likely arising from retroperitoneum. The patient was posted for surgery and intraoperatively the cyst was found to be arising from the right adrenal gland. Postoperative HPE revealed the features to be s/o mature cystic teratoma with carcinoid features. Postoperative period was uneventful.

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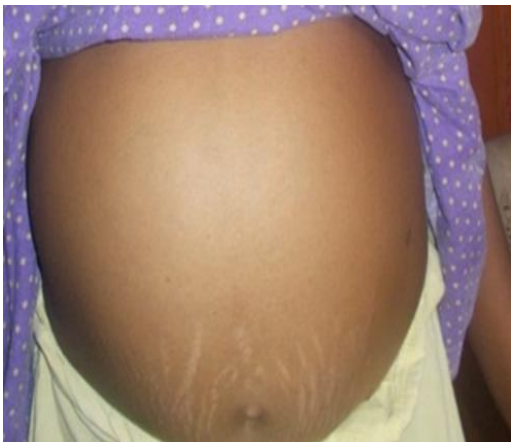
I. Introduction

Teratomas can occur anywhere in the body. They most commonly occur in gonads followed by mediastinum. They also occur in retroperitoneum arising from embryonic cell rests/adrenal glands. They can be solid or cystic or mixed. They can be mature or immature.Immature teratomas have a poor prognosis. They occur in young adults with a predisposition for females in the case of gonadal teratomas. They contain multiple tissue elements derived from the three primitive embryonic layers foreign to the area to which they occur. The following is a case report of Mature cystic teratoma arising from adrenal cell rests with carcinoid features

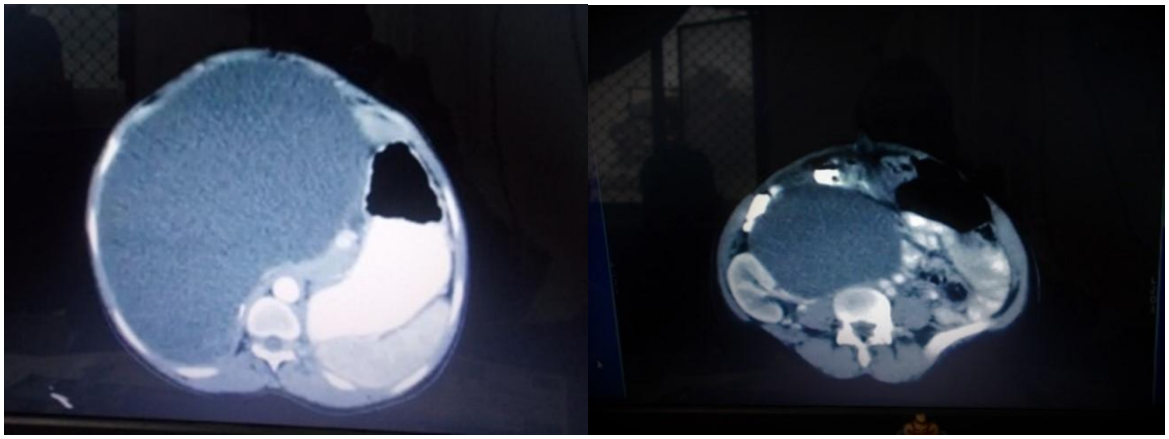
II. Case Report

A 28 year old female presented with vague, dull aching pain in the right upper abdomen for ten days with no other symptoms. On examination, the abdomen is asymmetrically distended, and umbilicus is shifted downward. A vertically oval lump of size 18×15cm is seen right hypochondrium, right lumbar, epigastric and umbilical regions with well defined, rounded borders.It is soft in consistency. The lump is becoming less prominent on the head raising test.It is not moving down with respiration and not falling forward on knee elbow position. On percussion, a dull note is present. Rest of the abdomen is normal. Based on the clinical findings, the provisional diagnosis is right sided retroperitoneal cystic swelling. Routine blood investigations and radiological imaging were done.



ULTRASOUND FEATURES:
A 18×15cm large cystic lesion with internal echoes and peripheral wall calcifications is noted displacing liver upwards and Rt. Kidney laterally .

CECT ABDOMEN.
A large thin walled hypodense lesion with wall calcifications with thin septa in the right side of the abdomen pushing the liver superiorly, right kidney laterally and inferiorly, bowel loops to the left and pancreas to the left-mesenchymal/omental cyst.



She was posted for Laparotomy for excision of the cyst.

INTRAOPERATIVE FINDINGS

A large cystic mass of size 18* 15 cm seen arising from the retroperitoneum pushing duodenum and pancreas anteriorly, liver superiorly ,IVC and aorta medially , kidney inferiorly.contents of the cyst on aspiration are turbid liquid like with pale yellow in colour.Cyst is attached to surrounding structures by flimsy adhesions.Entire cyst is excised taking care not to splill the contents.Specimen is sent for histopathology.

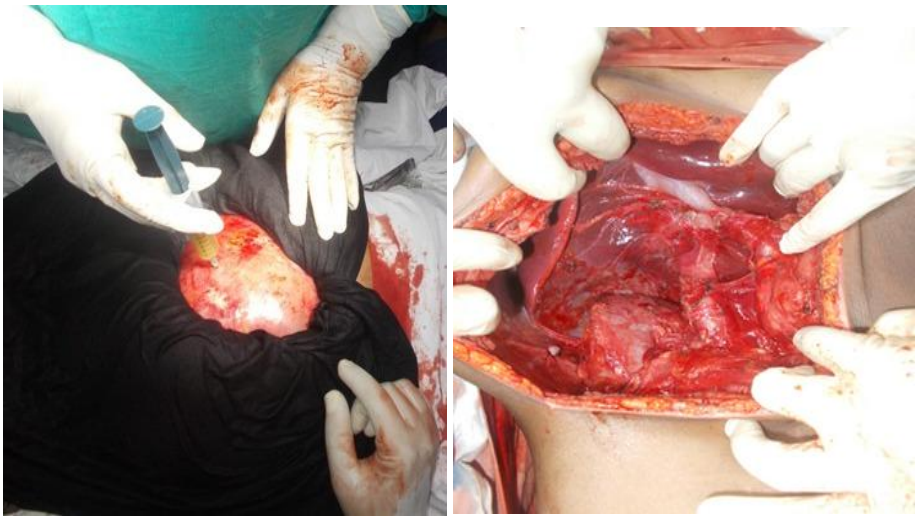
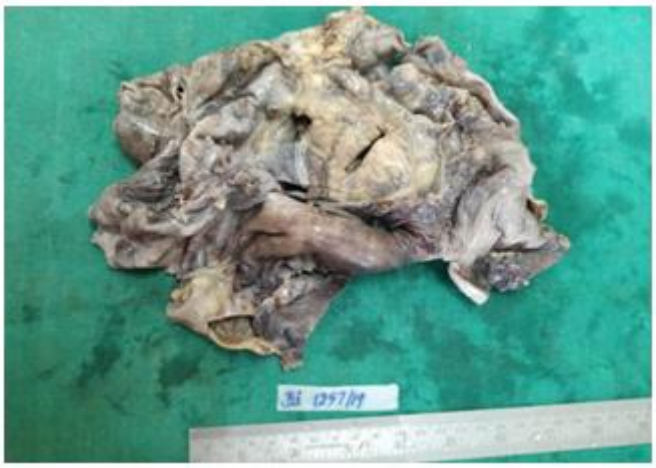


Fig.Intra operative pictures showing cyst and its's contents on aspiration and post operative retroperitoneal structures after removal of cyst.

POST OPERATIVE PERIOD
It is uneventful. Patient recovered well and discharged on 7 th post operative day.

HISTOPATHOLOGY REPORT
GROSS FEATURES :
18x15x5cm unilocular cyst in cut open state with thickness of wall varying from 1.4cm to <0.5cm.
External surface irregular, greyish white& grey brown in colour
The inner surface shows multiple grey white and elevated yellowish patches, the largest measuring 3cm diameter

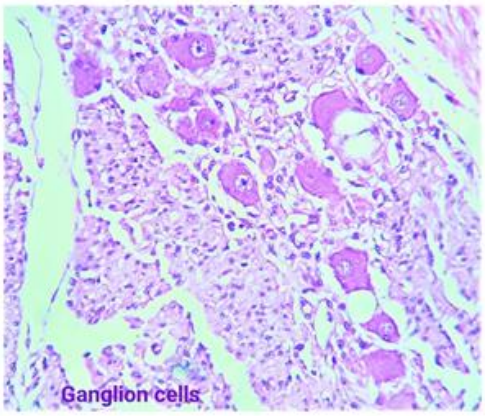
Focal gritty areas – calcifications.

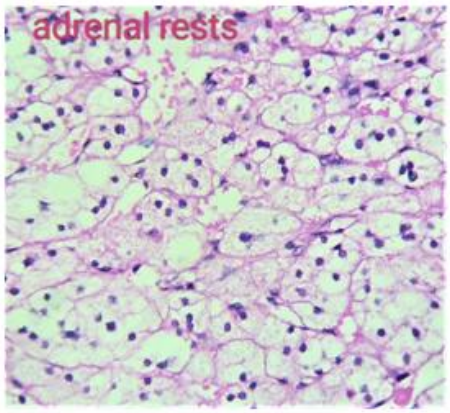


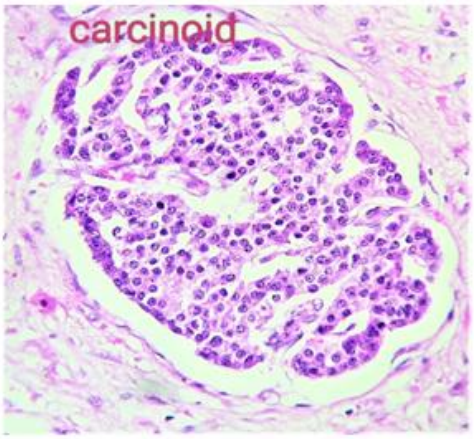
MICROSCOPIC FEATURES:

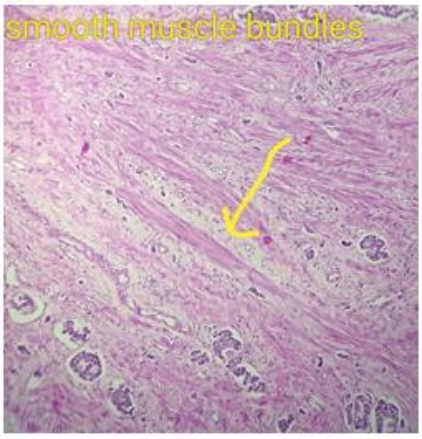
Retroperitoneal cyst lined by flattened cuboidal epithelium. Cyst wall composed of fibro collagenous tissue along with congested blood vessels, hemorrhages,and foci of normal appearing adrenal tissue. There are also large areas of neuroendocrine cells arranged in sheets, cords,acinipattern, and insular pattern .Also, good number of ganglion cells, nerve bundles, and small cystic spaces lined by cuboidal to low columnar epithelium are seen. These features suggestive of Mature cystic teratoma with carcinoid features in the adrenal gland.

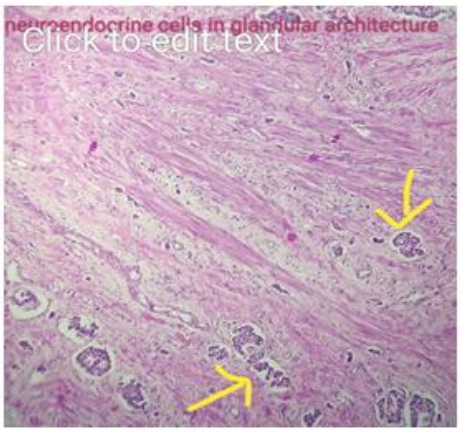
IHC revealed positive immunostaining with neuron specific enolase, chromogranin, and CD 56 and ki67 is 2% with the final report as a Well differentiated neuroendocrine tumor in a benign cystic teratoma of adrenal gland











III. Discussion

Germ cell tumors (GCTs) can be broadly classified into two main categories: seminomatous and nonseminomatous GCTs.Teratomas belong to nonseminomatous GCTs and represent the most common form of all GCTs1. Teratomas are encapsulated neoplasms composed of multiple parenchymal tissues (of varying degrees of differentiation) that are derived from more than one germ cell layer (ectoderm, mesoderm, and endoderm). Generally, teratomas arise from the uncontrolled proliferation of pluripotent cells: germ cells and embryonal cells. The type of pluripotent cell greatly influenced the presentation time and involved the location of teratoma. Teratomas of germ cell sources can be congenital or acquired and are usually found in gonads (testes and ovaries). In contrast, teratomas of embryonic cell sources are always congenital and are usually found in extragonadal locations, such as intracranial, cervical, retroperitoneal, mediastinal, and sacrococcygeal sites[2,3]. According to the area of tumor, teratomas can be classified into gonadal and extragonadal teratomas. Gonadal teratomas are more common, mostly primary neoplasms, mainly in adults, and usually take place in gonads (testes and ovaries)[4].

Conversely,extragonadal teratomas are less common, mostly secondary neoplasms, mainly in infants and young children, and usually take place in sacrococcygeal, mediastinal, retroperitoneal, and pineal gland sites (descending order of frequency)[4]. Furthermore, according to the content of the tumor, teratomas can be classified into solid, cystic, or mixed teratomas. Solid teratomas lack organization and contain only parenchymal tissues. Cystic teratomas contain only sacs of fluid, semifluid, or fat, whereas mixed teratomas contain both solid and cystic components. Besides, according to the epithelial lining and dermal contents of the tumor, teratomas can be classified into epidermoid, dermoid, and teratoid teratomas (cysts). Epidermoid teratomas are lined by stratified squamous epithelium and lack dermal contents. Dermoid teratomas are mostly lined by stratified squamous epithelium and contain various dermal contents such as hair, sweat, and sebaceous glands. Teratoid teratomas are mostly lined by respiratory columnar epithelium and contain sebum [3].

In addition, according to the degree of tumor maturation, teratomas can be classified into mature and immature teratomas. Mature teratomas are generally benign, asymptomatic and more common, among females. They are highly variable on histology and can be solid, cystic, or mixed. They contain different types of parenchymal tissues that are well differentiated. Mature cystic teratomas (AKA dermoid cysts) may have partially to completely well-developed organ systems. On the contrary, immature teratomas are histologically solid teratomas and contain immature (undifferentiated/undeveloped) parenchymal tissues and can be possibly

benign, possibly malignant, or frankly malignant. They are more common among males. Some mature(benign) and immature (possibly benign or possibly malignant) teratomas have an increased tendency to become frankly malignant teratomas, and frankly, malignant teratomas have an increased propensity to metastasize. This group of exceptionally rare teratomas is known as teratomas with malignant transformation(TMT)[5]. Malignant TMT is extremely rare and is usually found in gonadal organs, either testis or ovary. The stratified squamous epithelial components of these teratomas are the ones at an increased risk of undergoing malignant transformations. In addition, teratomas with malignant transformation may produce components of somatic (non-germ cell) neoplasms such as carcinoma, sarcoma, and leukemia [7,8].

Occasionally, a teratoma may contain various components of other germ cell tumor, and hence it is not a pure teratoma per se, but rather it is a mixed germ cell tumor and has malignant nature. In infants and young children, these components are frequently endodermal sinus tumor and choriocarcinoma. A pure teratoma can be benign, however, highly aggressive in its clinical course as in a growing teratoma syndrome (GTS). GTS refers to a rapidly growing pure mature (benign) teratoma that appears during or following chemotherapeutic eradication of malignant components of a nonseminomatous germ cell tumor, and it has normal serum tumor marker levels of alpha-fetoprotein and human chorionic gonadotropin [9]. The vast majority of retroperitoneal teratomas are secondary neoplasms and mostly occur in males [7].

Primary retroperitoneal teratomas are extremely unusual neoplasms accounting for approximately 1–11% of all primary retroperitoneal neoplasms and typically occur in neonates, infants, and children age groups [10]. In adults, these neoplasms commonly present in the third or fourth decades of lives [11]. Primary retroperitoneal teratomas involving adrenal glands are exceedingly uncommon, accounting for only 4% of all primary teratomas and can be mistaken for other histologically related lipomatous adrenal neoplasms [4].They are more common in childhood and rarely occur in adults. Only a few case reports have been documented in literature so far. Clinically, adrenal teratomas usually occur in young women, in the right adrenal gland, and 90% are benign. The majority of cases are asymptomatic, present with nonspecific complaints, or identified incidentally on routine investigations [4].

Teratomas can be diagnosed based on high index of clinical suspicion, routine laboratory, and radiographic investigations [11]. With respect to high index of clinical suspicion, retroperitoneal teratomas involving adrenal glands may present congenitally, or later in life when they grow to massive sizes [12]. Clinical presentations are variable and include nonspecific, abdominal/ flank/ back pain, obstructive gastrointestinal, and genitourinary symptoms, as well as lower limb/ genital swelling due to lymphatic obstruction [11]. They can rarely present with complications such as secondary infections (abscess formation) [13], traumatic rupture leading to acute peritonitis [14] , or malignant transformations [15]. Midline (paraxial) teratoma masses, with restricted mobility, can be easily detected on physical examination [16]. With respect to laboratory investigations, retroperitoneal teratomas can express a diversity of serum tumor markers such as elevated alpha-fetoprotein(AFP), carcinoembryonic antigen (CEA), and CA 19-9. These serum tumor markers are helpful in clinical practice and can be used to monitor successful treatment or detect relapse in patients with specific tumor marker-secreting teratomas. With respect to radiographic investigations, they play valuable roles in the diagnosis of teratomas. Plain radiographs (X-ray) can identify calcified elements in 62% of whereas ultrasound(US) can greatly differentiate between cystic and solid elements. Computed tomography (CT) scans can better distinguish between fat (adipose tissue) and bone(calcified) masses. On the contrary, magnetic resonance imaging(MRI) scans can offer a better resolution of soft tissues, feasible identification of benign and malignant neoplastic features, and, most importantly, superior tumor staging assessment. However, generally, a definitive diagnosis of teratoma demands a histopathological evaluation. Surgical excision of benign (mature) teratoma is required for a definitive diagnosis(by histopathological examination) and remains the mainstay of treatment. Prognosis is fortunately excellent after complete surgical excision with an overall five-year survival rate of nearly 100%. Teratomas are largely resistant to radio- and chemotherapy. Adjuvant radio-and chemotherapy are used only if malignant features of germ cell tumors are identified on histopathological examination. A testicular ultrasound (US) is highly advised to rule out potential coexisting germ cell tumors (GCTs) as approximately 50% of men with retroperitoneal teratomas have testicular carcinomas in situ at the time of diagnosis, which, if left untreated, can develop into testicular germ cell tumor. Well differentiated Neuroendocrine tumours have abundant secretory granules, Chromogranin A and Synaptophysin arranged in well-developed organoid or neuroendocrine shape with nesting, trabecular or gyriform/ serpentine growth patterns. If the neuroendocrine component >30 percent of the tumor, then it is a Neuroendocrine carcinoma(NEC). About 40 percent of NECs contain non-neuroendocrine histology. Carcinoid tumors arising in mature teratomas are tumors with low malignancy potential. Without resection of the tumor, the risk of malignant transformation increases with time.

This case was notable due to the rarity of this occurrence. In this case, there were no features of carcinoid syndrome preoperatively, no hormonal profile done preoperatively and on postoperative followup patient was asymptomatic. In conclusion, primary adrenal teratoma is a rare tumor, which is more common in adults compared with children. In both adults and children, adrenal primary adrenal teratoma usually manifests

as a large adrenal mass, preoperative diagnosis is difficult, and diagnosis can only be confirmed by histopathological examination following surgical removal of the tumor. If features of carcinoid syndrome coexist, evaluation of adrenal hormonal profile is mandatory preoperatively. Surgical excision of benign (mature) teratoma is required for a definitive diagnosis (by histopathological examination) and remains the mainstay of treatment. Prognosis is fortunately excellent after complete surgical excision with an overall five-year survival rate of nearly 100%. Teratomas are largely resistant to radio- and chemotherapy. Adjuvant radio- and chemotherapy are used only if malignant features of germ cell tumors are identified on histopathological examination.

IV. Conclusion

Primary retroperitoneal teratoma involving the region of adrenal gland is exceedingly rare (4% of all primary teratomas), and its occurrence in an adult is exceptionally uncommon. However, it should be regarded in the differential diagnosis in any patient presenting with a flank pain. Carcinoid tumour arising in a mature teratoma is a slow growing malignancy. Approximately 10% of all neuroendocrine tumours are complicated by carcinoid syndrome such as flushing, diarrhea, and valvular heart disease, which typically occurs with pulmonary or hepatic metastasis. The symptoms are attributable to circulating neuropeptides released from the neurosecretory granules of the carcinoid tumour cells, which are no longer sufficiently metabolized by the lungs and liver after metastasis. Histopathological examination of the resected tumor warrants a definitive diagnosis. Surgical excision of mature (benign) teratoma remains the mainstay of treatment with an excellent five-year survival rate of nearly 100%.

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