MATURE CYSTIC TERATOMA OF LEFT ADRENAL GLAND DIAGNOSED AS MYELOLIPOMA

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Abstract

A 32 year old woman who presented with fever, sore throat and abdominal pain that was evolving from last 7 months. A huge mass was palpable in left hypochondrium and left lumbar region. All baselines were within normal range. On ultrasonography, it appeared as well defined diffusely echogenic area in epigastric region likely arising from adrenal/pancreas, while CT scan findings reported a large heterogenous fat containing mass in left supra renal region. On the basis of CT findings, diagnosis of adrenal myelolipoma was made, exploratory laparotomy was done, the adrenal mass was excised and sent for histopathology. The biopsy report has shown that microsection of lesion were composed of benign squamous and pseudostratified epithelium, fat cartilage and glial tissue. Postoperative recovery of patient was uneventful.

Keywords: extragonadal teratoma, retroperitoneal teratoma, adrenal myelolipoma, mature cystic teratoma of adrenal gland

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The teratoma are nonseminomatous germ cell tumors which are composed of parenchymal tissue mostly derived from all three germ layers.[1]

These are well differentiated tumors and affect gonads and paraxial (midline) structures most of the time. Extragonadal sites are sacrococcegeal, mediastinal, retroperitoneal and pineal region and account for 15% of the total.[2] The least common of these tumors are retroperitoneal teratomas[3] and among those, primary mature cystic teratoma of adrenal gland is extremely rare.[3,4] Primary retroperitoneal teratoma is reported in population under 30 years of age and only 10% are reported above this age.[2] Adrenal teratomas are usually benign tumors with no functional symptoms but it's a great challenge for a surgeon to diagnose them because they usually mimic myelolipomas, angiomyolipoma or liposarcomas, however, adults may show some malignant transformations. A literature review was done from the cases reported on internet from 2000-2018 and it showed 29 adult cases and 6 pediatric cases of adrenal teratoma. A study shows that majority of the reported cases were females.[5] Most of the cases are asymptomatic until pressure symptoms started after the increase in tumor size affecting neighboring structures. Surgery is required to make a histopathological diagnosis[6] and is the mainstay treatment option. Successful surgery resulting in the complete removal of tumor has shown excellent prognosis with nearly 100% five-year survival rate.[7] Here we are reporting a case of mature cystic teratoma/ dermoid cyst of left adrenal gland in 32-years female who presented with nonspecific upper abdominal pain for 7 months and heaviness for 4 months with off and on fever spikes.

CASE PRESENTATION

A thirty-two year old married female with off and on dyspepsia was referred because of incidental finding of mass on ultrasound of abdomen and pelvis. On examination, a mass measuring 9x10cm was found in left lumbar region. The mass was firm in consistency and had ill-defined borders. Repeat ultrasonography was done and it showed a well-defined diffusely echogenic area in epigastric region which seems to be arising from pancreas extending up to left lumbar

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region abutting the left kidney. It measured 109 × 90 × 76mm. Few internal echogenic foci were also seen.

CT scan of abdomen was advised and it was suggestive of large heterogenous fat containing mass in left supra renal region. Left adrenal gland was not visualized. The mass had internal calcifications and area of soft tissue attenuation. All these findings were suggestive of adrenal myelolipoma.

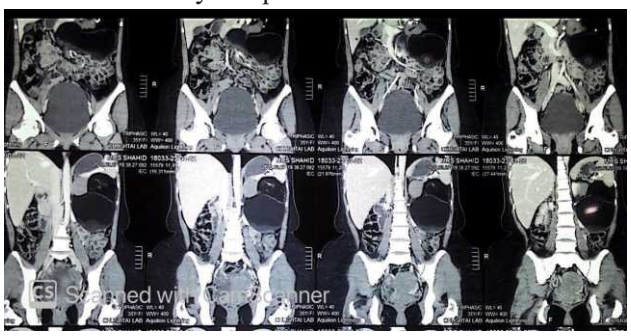

Figure 1: *The coronal view of CT abdomen showing large heterogenous fat containing mass in left supra-renal region*

Blood investigations had shown serum cortisol, dihydroepiandrostenedione (DHEA SO 4), plasma metanephrines and nonmetanephrines and serum electrolytes within normal ranges. So a provisional diagnosis of adrenal myelolipoma or adenoma was made and elective procedure of exploratory laparotomy with left adrenalectomy was planned.

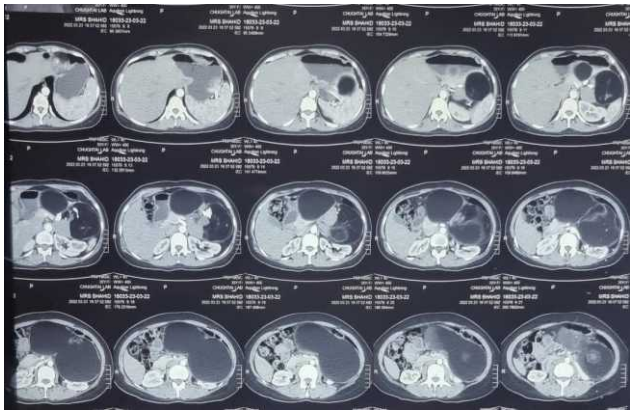

Figure 2: *Axial view of CT scan shown in Figure 1*

Intraoperatively a huge heterogenous mass of 10×15 cm was found which was well encapsulated and multilobulated. It was arising from left adrenal gland, splaying the transverse colon and mesocolon, pushing stomach superiorly, abutting lower border of pancreas, compressing left renal vein and causing its cavernous dilatation. The mass had its blood supply from multiple feeding vessels. Left kidney was spared. All abdominal viscera were intact.

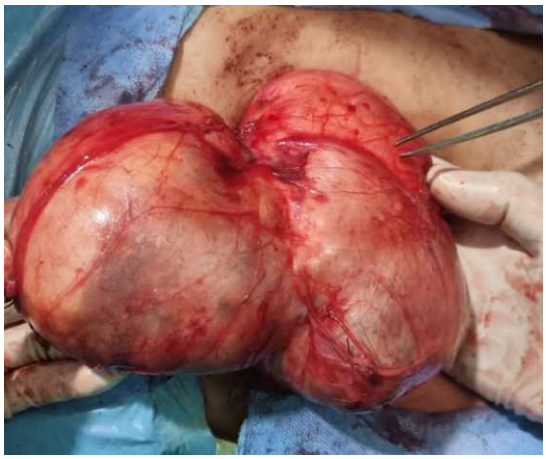

Figure 3: *Intraoperative picture of tumor*

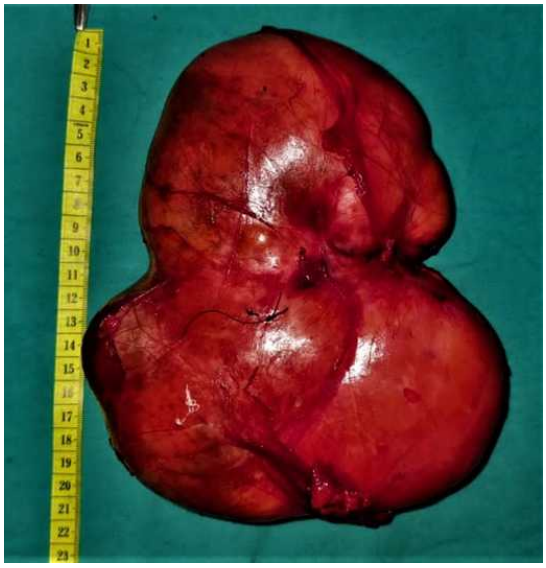

Figure 4: *The picture of tumor after resection*

The mass was resected along with intact capsule and left adrenal gland. Grossly, the capsule was intact. No surface deposits were present. On slicing, the cyst was filled with cheesy material and hair follicles with areas of fat, hemorrahge and some calcifications can be seen. Maximum thickness of capsule was 1cm. Representation sections were taken and submitted as follows: A) Medial and lateral margins B) Anterior resection margin C) Deep resection margin D) Resected margin E) Superior resection margin F) inferior resec-

tion margin G-K) Representative margins.

Histopathological findings showed benign squamous epithelium, fat, cartilage, pseudostratified epithelium, glial tissue and benign cartilage.

DISCUSSION

Teratomas arise from uncontrolled proliferation of pluripotent cells: germ cells and embryonal cells. The site and time of presentation of tumor is greatly dependent on its origin. Extragonadal teratomas arise generally from embryonic cell source.[8] They can be solid cystic or mixed tumors. In-addition, teratomas can be mature or immature depending upon the maturation of parenchymal tissues present in them.

Mature cystic teratomas (aka dermoid cyst) have partially or completely matured tissues e.g. bone, muscle, thyroid etc. and they have less likely chance of becoming malignant. However, in calcified teratomas malignancy should be ruled out as 12.5% of calcified teratomas are malignant.[9]

Most of the retroperitoneal teratomas are secondary benign tumors which are unusual and accounts for about 11% of all retroperitoneal tumors. They typically occur in pediatric age group, however, adults are affected in their 30s and 40s.[10]

Clinically adrenal teratomas are usually asymptomatic until these tumors bulk up and press the surrounding structure causing pain and obstruction.

Most of the time the patients present with nonspecific lumbar or flank pain, however, obstructive symptoms involving gastrointestinal genitourinary tracts and lymphatic vessels are not uncommon.[10] It's very rare for these tumors to present as abscess or acute peritonitis which is usually due to trauma or secondary infections history and examination of such patients provides vague information, investigations play a vital role in making diagnosis.

Some retroperitoneal teratomas secrete tumors markers like alfa fetoproteins and carcinoembryonic antigens. It's very helpful to detect these markers in patients to monitor treatment efficacy and relapse.[11]

Radiological investigations are the best options in making diagnosis prior to surgery. USG helps differentiating between solid and cystic lesions but it is less helpful in diagnosing retroperitoneal diseases. CT and MRI scan offer better view of anatomy and differentiation of soft tissues.

On computed tomographic scans, teratomas appear as heterogenous masses of fatty tissue with calcifications and magnetic resonance imaging shows teratoma as intensely highlighted area around the tumor component.

However, definitive diagnosis always requires histopathological evaluation. Pathological criteria for benign lesions were met in this case (1) absence of other similar lesions in other parts of body (2) absence of other similar lesions in other parts of body (3) normal serum levels of AFP and hCG (4) absence of recurrence on long-term follow-up.[1]

The diagnostic criteria of this reported case is similar to the previously reported cases.

Surgery is the only treatment available. These tumors are highly resistant to radio-and chemotherapy. Surgery can be done by open and laparoscopic methods but this is highly dependent on size of tumor, site of tumor, surgeon skills and patient's choice. However, laparoscopic surgeries have shown short postoperative hospital stays, less SSIs and little need of analgesia.

All patients diagnosed with retroperitoneal teratomas should have a testicular ultra sound scan at the time of discharge to rule out coexisting germ cell tumors in gonads. Studies have shown that approximately 50% of patients with retroperitoneal teratomas have testicular carcinoma in situ.[13-15]

CONCLUSION

Retroperitoneal teratomas, specifically adrenal teratomas are very rare tumors especially in adult age. These usually present with pressure symptoms on surrounding areas. Radiological investigations are best option for diagnosing these tumors. Complete surgical removal provides almost complete cure with very low recurrence rate. All patients diagnosed with these teratomas must have their testicular ultrasound done as approximately 50% cases had shown association with testicular carcinoma in situ.

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