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RESEARCH ARTICLE

PRIMARY ADRENAL GANGLIONEUROMA: A CASE REPORTAND REVIEW OF THE LITERATURE

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ABSTRACT

An Adrenal Ganglioneuroma (AGN) is an extremely rare, hormonally silent benign tumor. It is common for AGNs to be erroneously diagnosed, considered as other adrenal malignancies. Therefore it is imperative that, in work-up of adrenal incidentalomas, other less common aetiologies such as ganglioneuroma should be considered, and the latter should be suspected in adrenal mass on imaging without associated hormone hyper production. Here, the present authors report a case of giant adrenal ganglioneuroma in a 25-year-old female presenting with vague right side abdominal pain, after abdominal computed tomography (CT) scanning, the patient underwent an open right adrenalectomy and histopathological examination further confirmed the lesion as giant AGN, which measured 16 cm * 13 cm* 10 cm.

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INTRODUCTION

Ganglioneuroma is an exceptional, differentiated and benign tumour arising from primordial neural crest cells (1). They are mostly localized in the posterior mediastinum and retroperitoneal region; pure adrenal ganglioneuromas are especially rare (2). Ganglioneuromas comprise about 0–6% of all of adrenal incidentalomas (3). When the diameter of AGN extends over 6 cm, such size will be called "Giant" (4).These tumors are commonly asymptomatic and hormonally silent even when the lesion is of considerable size (5).The imaging characteristics of adrenal GN are inconstant and some are very similar to other adrenal tumors such as adrenocortical carcinoma (ACC) and pheochromocytoma (6-7). Therefore it is very common that AGN is misdiagnosed preoperatively, this leads to AGN being labelled as "a clinical & surgical dilemma" entity for surgeons and causes unwarranted delay in management. This obstacle can be overcome by detailed history & examination, timely diagnosis by means of endocrine evaluation and imaging leading to accurate management in the form of surgery thus enabling a definitive diagnosis to be made by histological examination.

In this case report, we present the case of a 25-year old female patient incidentally discovered to have a large adrenal ganglioneuroma on computed tomography (CT) scanning during an evaluation of her right lower-quadrant pain & final diagnosis was confirmed by histopathological examination.

Case presentation: A 25-year old married female patient was admitted to the Urology department with complaints of nonspecific, non- colic, intermittent left flank pain. She had pain for six months. She had no symptoms of headache, nausea, vomiting, diarrhoea, blurred vision, palpitation or chest distress, and there was no loss of appetite or weight.Physical examination revealed no abnormal mass, tenderness or rebound tenderness in the abdomen and the results of routine laboratory tests were all found to be within the normal range.Further evaluation included an abdominal ultrasound demonstrating a heterrogenous suprarenal cyst on right side of size approximately 13 x 10 cms. Contrast enhanced CT abdomen & pelvis was done which reported a hypo dense SOL in right hypochondrium (32 HU) of size 10.8x 13.4x15.7 cm with minimal enhancement in late venous phase, margins of the lesion were well maintained with surrounding structures, right side adrenal was not separately seen (Fig.1).

There was no evidence of abnormal nodes or an enhanced softtissue mass. Retroperitoneal lymph node involvement was not detected, and there was no evidence of invasion to the adjacent tissues or organs. An endocrine evaluation consisting of urine catecholamine and cortisol levels and a 1mg overnight dexamethasone suppression test was normal. With these results, it was not possible to determine whether the adrenal tumor was benign or malignant and the patient was symptomatic, the decision was taken to proceed with right side adrenalectomy, intraoperatively a large solid heterogeneous mass was found which had pushed right kidney inferiorly and inferior vena cava ventrally, multiple feeding vessels were present, the mass was removed in toto and specimen sent for histopathology (Fig. 2). The postoperative period was uneventful and she was discharged on postop day 5. Histopathology reported the largest diameter of the tumor as 16 cm with negative surgical margins, microscopically it demonstrated scattered ganglion sheets in dense schwannian stroma & mature ganglion cells. Immunohistochemistry was positive for S-100 confirming the diagnosis of adrenal ganglioneuroma schwannian dominant maturing type (Fig.3). The patient is doing well & no recurrence was detected during the one-year follow-up visits.

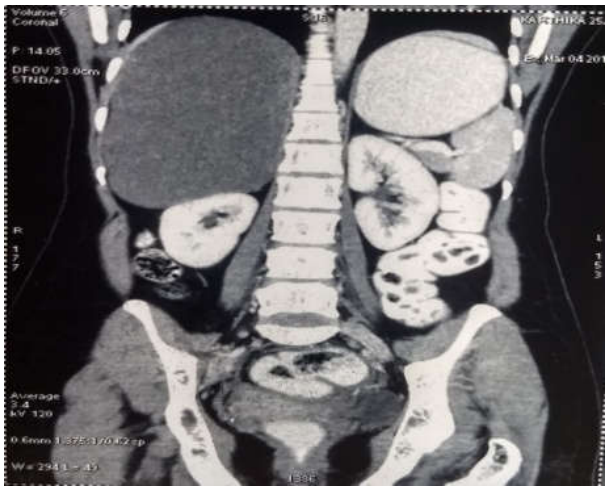

Figure 1. A hypo dense SOL in right hypochondrium (32 HU) of size 10.8x 13.4x15.7 cm

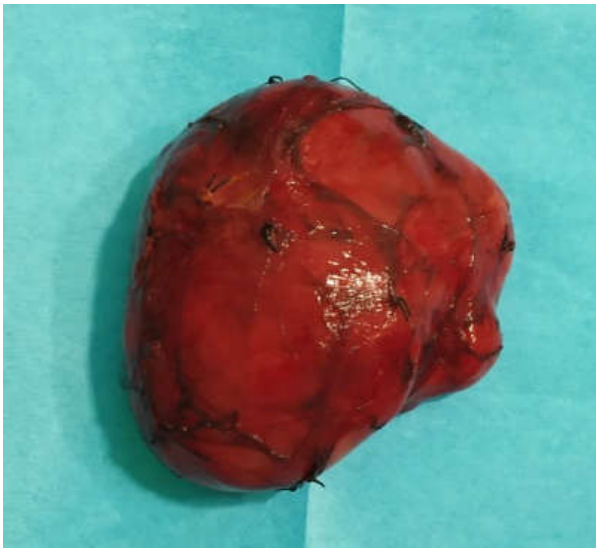

Figure 2. A large solid mass was removed in toto and specimen sent for histopathology

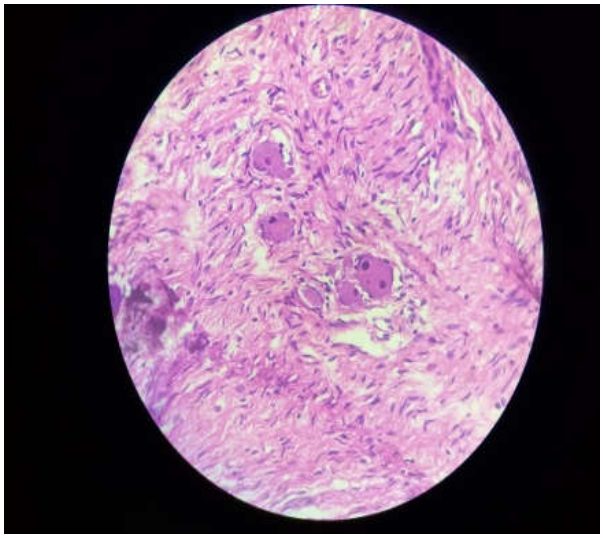

Figure 3. IHC was positive for S-100 showingschwannian dominant maturing type

DISCUSSION

Ganglioneuroma (GN) arises from the neural crest cells —the sympathetic ganglia and the adrenals. It is composed of Schwann cells, ganglion cells and fibrous tissues (8).Ganglioneuroma is a very erratic benign tumor and unusually found in the adrenal gland (2, 9). In Indian scenario there is a dearth of literature regarding case reporting of rare entities. Only a handful case of Adrenal Ganglioneuroma (AGN) was reported (10-13). AGN usually occurs in patients ranging from age 10 to 40 (4). In the present case too, the patient belongs to the younger age group. Adrenal Ganglioneuroma (AGN), most commonly involves the right adrenal gland (14). While Adas *et al* (9) report a case of AGN involving left adrenal gland. In present study, AGNs of patient occurs in right adrenal gland as well. Majority of adrenal ganglioneuroma do not demonstrate any clinical manifestations, However a minority of patients exhibit slow growing abdominal mass or abdominal pain (4). In the present case, the patient complains of dull & intermittent abdominal pain. Changwen Zhang *et al* (15) concluded that no gender bias is seen in AGN.Komai Y *et al* (16) reported the largest AGN with 22 cm in size. In the present case, the largest dimension of lesion was 16 cm.

The majority of patients with AGN is hormonally silent; rarely cause symptoms of diarrhea, virilization and hypertension (14). In the present case too, patient did not secrete hormone & did not have symptoms of hypertension or virilization.A minority of cases of AGN reported in literature produce hormones such as catecholamines, vasointestinal peptide and androgens resulting in symptoms like weakness, diarrhea, hypertension and virilization (17-18). The preoperative diagnosis of AGN uniquely based on CT & MRI is very challenging (9).Qing *et al* (6) stated that the misdiagnosis rate of adrenal GN on CT and MRI before surgery is 64.7%. The imaging characteristics of adrenal GN on CT and MRI have been well defined withthe most noticeable imaging finding of an AGN is a count of <40 HU (14).However in our case, the CT value was 32 HU. Tumor size is very significant to conclude the prognosis for adrenal masses, size larger than 6 cm, seems to be a forecast of malignancy in 25% cases (19) & open surgery is recommended for giant AGN (4).

However Zografos GN *et al* (20) stated that irrespective of size laparoscopic adrenalectomy became a standard curative treatment for AGN because of low invasion and benign nature. Abraham *et al*(21) reported that the largest AGN resected by laparoscopic approach was 17 cm in size with excellent prognosis. In the present case report, the patient underwent open adrenalectomy.

Conclusion

This case report presents an extremely rare entity of adrenal ganglioneuroma (AGN). A preoperative identification of AGN can be a daunting assignment, particularly in asymptomatic cases. Therefore, early careful evaluation is done to rule out the other possible differential diagnosis by means of endocrine tests, radiological imaging and treatment are extremely significant and the gold standard method to reach a definitive diagnosis is histological evaluation. The prognosis of AGNs is excellent.& the treatment is complete surgical resection without the need for chemotherapy or radiotherapy because of the benign nature of the lesion. This case has been reported for its rarity and to illustrate the point that when preoperative diagnosis is not possible, histological examination provides the final answer.

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