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Case Report

Pathology Section

Adrenal Ganglioneuroma: A Rare Incidentaloma

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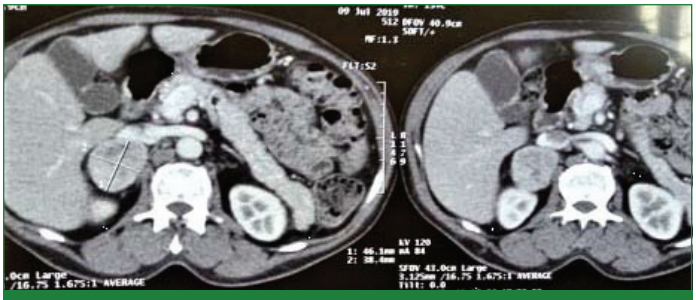
ABSTRACT

Adrenal ganglioneuroma is a very rare tumour of sympathetic nervous system that originate from neural crest sympathogonia which are completely undifferentiated cells of the sympathetic nervous system, constituting 20 to 30% of all Ganglioneuromas (GNs). GNs are generally asymptomatic and usually do not secrete hormones and most often detected incidentally on routine imaging tests during work up. The tumour could cause some complications, if it becomes large enough to press against the adjacent organs. Ganglioneuroma generally present as solitary mass which are painless and grow slowly. The common sites of GN are posterior mediastinum (41%), retroperitoneum (37%), adrenal gland (21%) and neck (8%). Histopathologically, ganglioneuroma is entirely composed of ganglion cells and schwannian stroma and does not contain neuroblasts, intermediate cells or mitotic figures. Preoperative diagnosis of GNs is difficult and cannot be made on radiological findings alone, thus histopathological examination is required in order to confirm the diagnosis of GN. Adrenal ganglioneuroma is a rare sympathetic tumour which originated from adrenal medulla. Prognosis is good after removal of the tumour, adjuvent therapy is not required and there is no recurrence. This report is of a rare case of adrenal ganglioneuroma in a 66-year-old male from Bihar came with complaints of bipedal oedema for seven days, white coloured urine and burning pain during micturition for three months. The patient was evaluated for chyluria, routine investigations were normal but ultrasound and Computed Tomography (CT) scan of abdomen revealed non-specific solitary right adrenal mass measuring 44.8×38.2 mm. Biochemical investigations showed increased catecholamines and metanephrines in plasma. The excised mass was sent for histopathological examination which showed features of adrenal ganglioneuroma and same was confirmed by immunohistochemistry.

Keywords: Catecholamines, Metanephrines, Sympathetic chain tumour

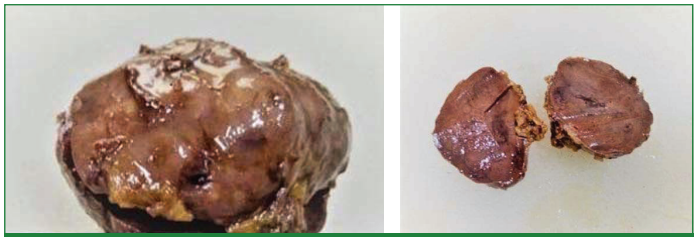
CASE REPORT

A 66-year-old male came in Urology Surgical Outpatient Department with complaints of bipedal oedema for seven days, white coloured urine and burning pain during micturition for three months. The patient was addicted to tobacco chewing for 40 years. There was no past history of diabetes, hypertension and thyroid. He had undergone bilateral hydrocoele surgery two years back. On general examination, no abnormality was found. Systemic examination did not show any abnormality in chest, cardiovascular system and abdomen. Per rectal digital examination showed grade three prostate enlargement. Hematological investigation did not show any abnormality. Biochemical investigations showed increased catecholamines and metanephrines in plasma. Prostatic specific antigen test was not done. Being evaluated for chyluria, ultrasound abdomen revealed incidental right adrenal mass measuring 44.8×38.2 mm. There was no obvious flow or necrosis or calcification. The patient underwent CT abdomen which revealed a tumour in right adrenal gland which was well-defined diffusely enhancing mass lesion of size 44.8×38.2×41.6 mm [Table/Fig-1] detected in right adrenal gland. The differential diagnosis of adrenal mass comprises of adenoma, pheochromocytoma and adrenal cancer. It is generally challenging to obtain a precise differential diagnosis of adrenal GN prior to surgery. The provisional diagnosis was of pheochromocytoma. The mass was excised and sent for histopathological examination.



[Table/Fig-1]: CT scan showing right adrenal mass of 44.8×38.2×41.6 mm.

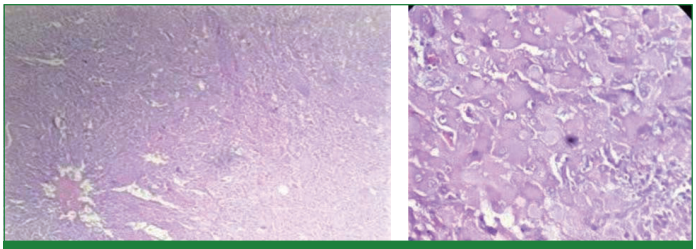
Gross Examination showed excised right adrenal mass measuring 4.5×3.8×4 cm [Table/Fig-2]. The majority of the tumour edge was smooth and the boundary is clearly defined. Cut surface showed well-circumscribed mass which is firm with resilient texture and brownish in colour [Table/Fig-3]. On microscopic examination, low power showed moderate cellularity with variable loose stroma [Table/Fig-4]. On high power, the cells showed admixture of ganglion cell and schwann cells [Table/Fig-5]. On the basis of morphology, the diagnosis of Ganglioneuroma was made. On immunohistochemistry, the ganglion cells show positivity for synaptophysin [Table/Fig-6a], neuron specific enolase [Table/Fig-6b] and schwann cell showed positivity for S100 [Table/Fig-6c], and final diagnosis of adrenal ganglioneuroma was made. Concomitantly, the patient was medically treated for filariasis and prostatomegaly. On follow-up, the patient was found free from symptoms. No recurrence of the tumour was found in six-month follow-up.



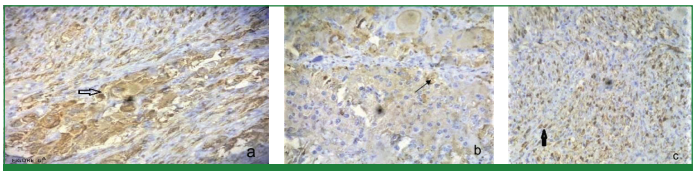
[Table/Fig-2]: Excised adrenal mass measuring 4.5×3.8×4.1 cm.
[Table/Fig-3]: Cut surface showing well circumscribed, firm, resilient texture, brownish in colour. (Images from left to right)

DISCUSSION

The GN is tumour of the sympathetic chain and it may be found in base of skull to the neck, posterior mediastinum, retroperitoneum



[Table/Fig-4]: On microscopic examination low power shows moderate cellularity with variable loose stroma (H&E; 10X).
[Table/Fig-5]: Section showing admixture of ganglion cell and schwann cell. (H&E; 40X). (Images from left to right)



[Table/Fig-6]: a) Immunohistochemistry showing ganglion cell showing positivity for neuron specific enolase (40X); b) Immunohistochemistry showing ganglion cell showing positivity for synaptophysin (40X); c) Immunohistochemistry showing 100 positivity for schwann cell (40X). (Images from left to right)

and rarely adrenal gland [1-3]. Since, adrenal ganglioneuroma is not very common and usually silent, they are most often detected during routine workup for other condition [4]. These are benign and well-differentiated. Adrenal tumours are found incidentally in 1–10% of abdominal CT scans; of these, 1% to 6% are GN [5]. Ganglioneuroma are slow growing tumour which is infrequent and originate from primitive neural crest cells and are composed of schwann cells and ganglion cells. GN's are rarely found in the adrenal gland [6,7]. Although GN is generally considered to occur more frequently in young people, some recent studies have shown that, it may also be seen between the ages 40 and 50 [8,9]. Due to the lack of significant clinical symptoms and signs, preoperative definitive diagnosis is very difficult, similar to previous studies the present case was incidentally found during ultrasound and CT scan of abdomen which was unrelated to the patient symptoms of chyluria, bipedal oedema and prostatomegaly. GN are generally non-secreting, but about 37% of tumour can secrete catecholamines, their metabolites and vasoactive intestinal peptide. In the present case, catecholamines and its product metanephrines were increased. In case of hormone secreting tumours, patients may experience diarrhea due to vasoactive intestinal peptide release, sweating or high blood pressure due to catecholamine release [10]. The patient had increased level of catecholamines and metanephrines which was subsided after excision of the mass.

Histopathologically, the schwann cells appear mixed together with ganglion cells that exhibit abundant eosinophilic cytoplasm and large vesicular nuclei with prominent nucleoli; the ganglion cells may be isolated or may form small aggregates. In the present case, similar findings of ganglion cell mixed with Schwann cell were found. Immunohistochemistry reveals that the ganglion cells are positive for synaptophysin, neuron-specific enolase, CD56 and the neuron-specific protein 9.5, and the Schwann cells are positive for S100 [11]. In the present case, ganglion cell is positive for synaptophysin and neuron specific enolase and Schwann cell is positive for S100. Prognosis after surgical resection seems to be excellent and there is no recurrence or need for adjuvant therapy.

CONCLUSION(S)

Adrenal lesions are uncommon surgical specimen and generally found during radiological imaging procedure. Careful clinical evaluation and endocrine tests raise the suspicion of ganglioneuroma. Histopathological examination with the help of immunohistochemistry make the definitive diagnosis. Hence, Ganglioneuroma can be considered as a differential diagnosis of incidental adrenal mass lesion.

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