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General Medicine

ADRENAL GANGLIONEUROMA- AN UNUSUAL PRESENTATION

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ABSTRACT Often found incidentally as a part of imaging (hence incidentaloma) or on evaluation for a specific complaint , adrenal masses can range from being just adenomas to malignant metastasis or pheochromocytomas thus warranting evaluation. comorbids with history of abdominal pain for a year with acute gastroenteritis for 2 days.USG picked up a right suprarenal mass12.7x 9 x12.2 cm.CECT Abdomen revealed central necrosis and non visualization of the adrenal gland: suggesting adrenal tumor ganglioneuroma / cortical carcinoma.Hormonal workup including 24 hour metanephrine test was normal.USG guided biopsy reported ganglioneuroma.Patient underwent adrenalectomy.

Adrenal ganglioneuromas are rare, hormonally silent tumors which may resemble malignancies.Appropriate evaluation by endocrine tests , imaging and histopathology is crucial for a diagnosis.

KEYWORDS : incidentaloma, adrenal mass, ganglioneuroma

INTRODUCTION

An adrenal mass can be found as part of evaluation for a specific complaint related to adrenal pathology or as an incidental finding on imaging for unrelated issues (incidentaloma).Most masses are either adenomas, myolipomas , malignant metastsasis, carcinomas or pheochromocytoma hence warrant investigation.This unexpected diagnosis has become increasingly frequent and a common problem in clinical practice owing to increasing availability of high resolution imaging procedures.

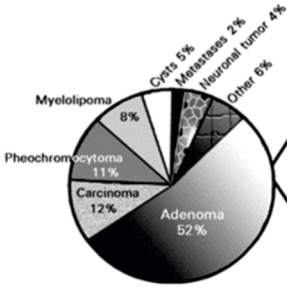


Fig 1 – Histological Diagnosis Of Adrenal Incidentalomas(2)

CASE REPORT

A 29 year old male with no previous co-morbids presented with fever , low grade, intermittent for 4 days and loose stools , about 3 episodes since 2 days , watery in consistency not associated with blood or mucus and 2 episodes of vomiting consisting of food particles.He also reported vague pain in the right lumbar region since 1 year.Patient also noticed weight loss over the last two months which he was unable to quantify.He had no other symptoms.On examination vitals were stable, general examination was normal and systemic examination revealed diffuse tenderness over the right hypochondrial and lumbar regions , otherwise normal.Routine lab investigations showed hemoglobin 11.9 , peripheral smear showed normocytic normochromic anemia.Other investigations were unremarkable with normal blood sugars, liver and renal function tests and urine analysis after which he was started on antibiotics and iv fluids.

Ultrasound abdomen showed a well defined heterogenoes lesion measuring 12.7x 9 x 12.2 cm in the right suprarenal region with minimal internal vascularity and mild splenomegaly.Fever panel sent reported negative.CT abdomen showed a well defined heterogenous lesion with lobulated margins in the right suprarenal region measuring 12.1x 14.2 x 9.3 cm : no areas of fat / calcification within.Post contrast the lesion showed delayed enhancement with hypodensity in the centre suggesting necrosis and non visualization of the adrenal gland suggestive of adrenal tumor ganglioneuroma / cortical carcinoma.Thyroid function tests and serum cortisol was within normal limits ; 24 hour urine metanephrine and normetanephrine tests were negative.

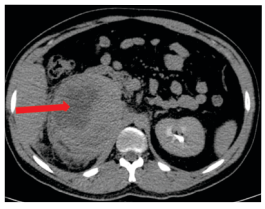
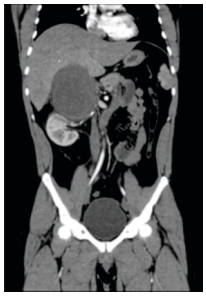


Fig 2 a) CT- contrast image of adrenal incidentaloma (axial)

b) coronal



Endocrine and surgical oncology consult was obtained ; USG guided trucut biopsy done reported back as ganglioneuroma.Patient subsequently underwent adrenalectomy.

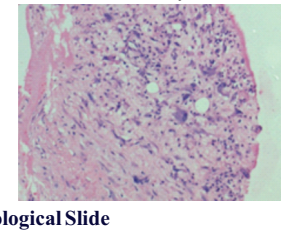


Fig 3 Histopathological Slide

DISCUSSION

A ganglioneuroma is a rare , differentiated and benign neoplasm arising from primordial neural crest cells of sympathetic ganglia or adrenal medula.On histology, it is composed of mature schwann cells and ganglion cells with fibrous stroma. Ganglioneuromas belong to neurogenic tumor groups that include ganglioblastoma and neuroblastoma.

They are non –functional and non symptomatic masses detected incidentally are called incidentaloma and can grow aggressively at times().They commonly present in the 4th or 5th decade of life.Their main locations include posterior mediastinum,retroperi toneum,adrenal gland and neck.Twenty percent are located in the adrenal medulla(3).

Although described well on imaging, they can mimic primary or secondary malignancy and precise diagnosis is difficult without histopathological exam and PET CT.Even though lesions over 4.5 cm predict malignancy, this need not be the case often.Recent studies recommend that non secretory incidentalomas larger than 4 cm or with suspicious features of malignancy on imaging should be treated with adrenalectomy(4).Surgically treated patients have excellent prognosis(5).A few cases of recurrence have occurred.Long term follow up is recommended

CONCLUSION

Adrenal ganglioneuromas are rare, hormonally silent benign tumors but can sometimes resemble other adrenal malignancies.Adequate evaluation by endocrine tests and imaging is necessary.Histopathology examination can make a definite diagnosis.Prognosis is good with surgical excision.

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