International Journal Dental and Medical Sciences Research
Volume 5, Issue 2, Mar - Apr 2023 pp 826-828 www.ijdmsrjournal.com ISSN: 2582-6018

Adrenal Ganglioneuroma - A rare tumor

Date of Submission: 10-04-2023 Date of Acceptance: 22-04-2023

ABSTRACT :- A 30 years old female presented with incidental findings of large mass in Right adrenal gland reported on a CT Scan abdomen as Benign Right adrenal adenoma. Further investigations revealed it to be a non functioning (non secreting) tumor.Because of the large size of the tumor, we decided to operate on her. Laparoscopic adrenalectomy was done. Histopathological examination of the specimen suggested it to be a Gangleoneuroma of the Right adrenal gland.

I. INTRODUCTION :-

Gangleoneuromas are benign & well differentiated tumors of the sympathetic nervous system. It consists of mature ganglionic cells & schwann cells. They can arise from the sympathetic chain anywhere in neck, mediastinum, retroperitoneum & adrenal glands (1). Posterior mediastinum is the most common location, followed by the retroperitoneum (2).

Ganglioneuroma of the adrenal gland is very rare (1). These tumors are usually asymptomatic & mostly detected incidently on the imaging studies.

The prevelance of adrenal incidentaloma is 0.2% in young patient, 3% in 5th, & 7% seventh decade (3) , (8).

Mostly these ganglioneuroma are hormonallyinactive & non functioning.

Imaging characteristic of adrenal Ganglioneuromas are variable, so a pre-operative diagnosis is very difficult. A definitive diagnosis can be made only by histopathological examination. (3)

In this case report we present a case of a 30 year old female patient with a adrenal tumor found incidently on a CT Scan.

II. CASE:-

A 28 years old lady presented with H/O-frequent micturition for 1.5 years. She was investigated for that.Her CT scan abdomen revealed exophytic isodense lobulated mass arising from the laterlal limb of right adrenal gland with mild to moderate in homogenous enlargement. No calcific/fat density foci within the mass.Smooth indentation on upper pole of right kidney without infiltration, findings are suggestive of benign adrenal mass, probably adenoma.The size of adrenal mass, was 6.3 x 5.0 x 3.8 cm (TR X CC X AP). Her routine blood investigations were normal. Her serum cortisol was 3.20 mcg/dl (Ref-0.056-63.44). Morning serum cortisol was 10.13 mcg/dl (Ref-4.30-22.40), serum aldosterone was 32.0 ng/dl, free plasma metanephrin was 58.00 ng/l (Ref-7.90-88.70). patient was operated & laparoscopic rigth adrenalectomy done.

Post operated histopathological report turned out to be Ganglioneuroma - Schwannian stroma dominant - maturing type.

III. DISCUSSION:-

Ganglioneuroma are benign tumors originating from neural crest cells & arising from great sympathetic chain. These tumors extend from the skull bone to the retroperitoneum and they rarely presents in the adrenal glands (4)(5). They usually present in the young age group, and are discovered incidently or due to non - specific mass effects symptoms (6).

On histology, it consists of mature schwann cells & ganglion cells with fibrous stroma (7)(9).

Ganglioneuroma is a member of neurogenic tumor group.It differs from other neurogenic tumors in its benign potential (10).

These non functional & non symptomatic tumors are usually detected incidently on USG or CT Scansand referred to as incidentaloma.

The pathology of adrenal incidentaloma may vary and differential diagnosis includes - adenoma, myo-lipoma, cyst, lipoma, pheochromocytoma, adrenal carcinoma, metastatic cancer, tuberculoma (3)

Ganglioneuroma can coexist with pheochromocytoma or paraganglioma and can be functional with high level of catecholamines or corticotrophin - releasing hormones (11).

Ganglioneuromas generally occur in young age group but can be seen among the age group 40-50 (12)(13).

Ganglioneuroma most frequently found in the posterior mediastinum & then in retroperitoneum.

Adrenal gland is the rare site for Ganglioneuroma (14).

Adrenal Ganglioneuroma is usually non functional & asymptomatic. Occasionally, composite tumors with pheochromocytoma are seen & can be functional (15,16).

Ganglioneuroma can also produce &

secrete other hormones such as catecholamines, VIPs, testosterones, although without causing any symptoms, indicating it's pleuripotency of precursor cells (7,17,18).

The macroscopic characteristics of adrenal Ganglioneuroma are an encapsulated mass with a firm consistency & a solid homogenous, greyish – white cut surface. HPE shows mature ganglion cells & schwann cells among a fibrous stroma.
Histologically they can be classified in two groups -Mature & Maturing (3)

According to analysis - Ganglioneuromas are characterised by reactivity for S-100, vimentin, synaptophysin, neural markers (16).

The precise diagnosis of adrenal Ganglioneuroma by imaging modalities before the surgery is difficult.
Adrenal Ganglioneuroma is described as a well circumscribed tumor with lobular shape & low alternation.

For tumor size large than 5 cm, heterogenocity & calcification may suggest malignancy (19).
PET Scans can help in making a diagnosis between adrenal Ganglioneuroma & ACC or Metastasis.

Recent studies recommended that non secreting adenal incidentalomas larger than 6 cm or with suspicious feature of malignancy or imaging study should be treated by adrenalectomy (20,12,8,21,10). Papavramidis et al reported that adrenal Ganglioneuroma should be treated by adrenalectomy & prognosis is excellent. The role of laparoscopic adrenalectomy in these case is well established due to the inherent advantages of the laparoscopy (22).

REFERENCES:-

[1]. Hayim Gilshtein et al : Ganglioneuroma of the Adrenal Gland: A rare Tumor in a Rare location. Case report in oncology 2012;5:0487-489.
[2]. Oliver J, fernandez A, Aguilera A etal. Ganglioneuroma adrenal : dilema clinico quirurgico acerca de un hallazgo fortuito, Endocrinal Nutr, 2013; 60:e37-e40.
[3]. Adas et al. Ganglioneuroma presenting as an adrenal incidentaloma: a case report. J Med CaseRep2014;8:131.
[4]. Linos D et al, Adrenal ganglioneuromas: incidentalomas with misleading clinical & imaging features. Surgery 2011; 149:99-105
[5]. Sucandy et al: Ganglioneuroma of the Adrenal gland and retroperitoneum: a case report. N Am JMed Sci 2011;3:336-338.
[6]. Hayes FA et al : Clinical Manifestation of Ganglioneuroma. Cancer. 1989.; 63 : 1211-1214.
[7]. Geoerger B et al : Metabolic activity and clinical features of primary ganglioneuromas, Cancer 2001, 91:1905-1913.
[8]. Grumbach MM, et al : Management of clinically inapparent adrenal mass (" incidentaloma"). Ann Intern Med 2003, 138(5):424-429.
[9]. Enzinger FM et al: Ganglioneuroma. In Soft Tissue Tumors 3rd edition. St.Louis: Mosby-YearBook; 1995:929-964.
[10]. Joshi W : Peripheral neuroblastic tumors: pathologic classification based on recommendations of international neuroblastoma pathology committee (modification of Shimada classification). Pediatr Dev Pathol 2000, 3:184 - 199.
[11]. Sahani S, et al: Ectopic corticotropin-releasing hormone (CRH) syndrome from metastatic small cell carcinoma: a case report & review of the literature. Diagn Pathol 2010, 8:56.
[12]. Qing Y, et al : Adrenal ganglioneuromas: a 10-year experience in a Chinese population . Surgery 2010, 147:854-860.
[13]. Rhondeau G, et al: Clinical and biochemical features of seven adult adrenal ganglioneuromas. J Clin Endocrinol metab 2010,95:3118-3125.
[14]. Jain M et al: Retroperitoneal ganglioneuroma: report of a case diagnosed by fine- needle aspiration cytology, with review of the literature. Diagn Cytopathol 1999, 21:194-196.
[15]. Charfi S, et al: Composite pheochromocytoma associated with multiple endocrine neoplasia type 2B. Ann Pathol 2007, 28:225-228.
[16]. Sasaki S1, et al: Large adrenal ganglioneuroma. Intern Med 2012,51:2365-2370.
[17]. Koch CA, et al: Adrenal ganglioneuroma in a patient presenting with severe hypertension & diarrhea. Endocr Relat Cancer 2003, 10:99-107.
[18]. Kogner P: Neuropeptides in neuroblastomas and ganglioneuromas. Prog Brain Res 1995, 104:325-338.
[19]. Fassnacht M, et al: Adrenal tumors: how to establish malignancy? J Endocrinol Invest 2004,27:387-399.
[20]. Yamaguchi K et al : Two cases of ganglioneuroma .Urology 2006,67:622.e1-

e4.
[21]. Maweja S et al: Adrenal ganglioneuroma. A neoplasia to exclude in patients with adrenal incidentaloma. Acta Chir Belg 2007, 107:670-674.
[22]. Papavramidis TS, et al: Retroperitoneal ganglioneuroma in an adult patient: a case report and literature review of the last decade. South Med J 2009, 102(10):1065-1067.