CASE REPORT

Case report of a rare incidentaloma of the adrenal gland—Schwannoma

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Abstract

The Schwannoma is a benign growth of the nerve sheath cells most commonly seen in the vestibulocochlear nerve. Its prevalence in the adrenal gland is 1-3%. Here we discuss a case that presented as an incidentaloma of the right adrenal gland in a young male patient who had vague abdominal symptoms and a normal hormonal profile. He underwent an excisional biopsy of the right adrenal gland due to the large size of the lesion (more than 4cm). The histopathology report helped to establish the diagnosis of Schwannoma. Incidentaloma is defined as a lesion of the adrenal gland encountered on any radiological investigation carried out for symptoms that are not associated with adrenal pathologies. After discovering such lesions, it is imperative to perform radiological and hormonal investigations in an organised manner to plan further management of such cases.

Keywords: Schwannoma, Adrenal Incidentaloma, Adrenocortical Adenoma, Adrenocortical Carcinoma, Adrenalectomy.

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Introduction

The Schwannoma is a benign growth of cells in the nerve sheaths of peripheral and cranial nerves known as Schwann cells which were discovered by a German physiologist, Theodore Schwann, in the nineteenth century. These cells fulfil the role of primary glial cells in the peripheral nerves and form the myelin sheath around axons outside CNS. These tumours most commonly arise in the head, neck, and extremities with the most common site being the vestibulocochlear nerve.[1] The frequency of presentation in the adrenal gland is extremely low as only 1-3% of all Schwannomas occur in the retroperitoneum and about 0.2% of all adrenal tumours are Schwannomas.[2,3,4] Adrenal Schwannoma usually presents as Incidentaloma on a radiological investigation. The only way to reach a definitive diagnosis is excisional biopsy and histopathology with immunohistochemistry.[5] Here, we present a case of adrenal Schwannoma which was diagnosed incidentally in the urology outdoor patient department of a tertiary care hospital in Lahore. Consent was obtained from the patient to publish this case.

Case Report

A 24-year-old male presented to the outdoor patient department (OPD) of Lahore General Hospital, Lahore in January 2022 with the complaint of right loin pain for one year. The pain was episodic with sharp character. There was no haematuria, nausea, or hypertension associated with it. The patient had lost 5kg in the past one year. His family history was unremarkable. Physical examination gave no positive findings. Ultrasound KUB showed a supra-renal mass on the right side; CT-KUB with contrast confirmed it as right adrenal mass of 10x9x9 cm in size. [Fig. 1]. On the biochemistry panel his serum cortisol, plasma metanephrine, serum testosterone, and urine VMA levels were in the normal range. A right open adrenalectomy was performed by the head of the urology department 21 days after his presentation in the OPD. During surgery, the adrenal mass was noted to be about 10x10x10 cm in size and it was densely adherent to the right renal artery, inferior vena cava, right psoas muscle, and liver. The mass was encapsulated, grey-brown and homogenous on the cut section. [Fig. 1]. Surgery was performed under general anaesthesia with endotracheal intubation, while an epidural was also applied. The sample was sent for histopathology in the hospital's lab. Postoperatively, the patient was shifted to the Surgical

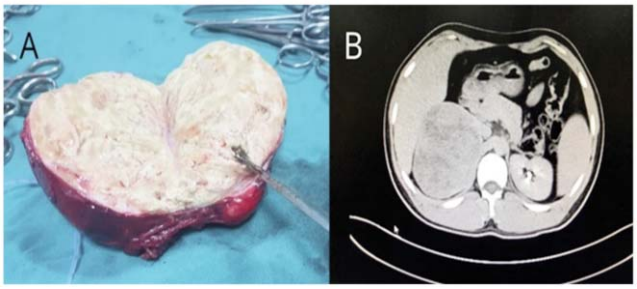

Figure-1: Image A shows the resected specimen of the right adrenal mass which was sliced, it shows a homogenous light brown colour with a capsule surrounding it. Image B shows the CT-KUB with a homogenous right adrenal mass.

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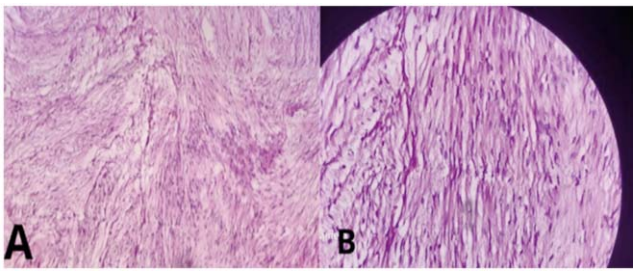

Figure-2: Image A shows H&E stained section (100x) containing Verocay bodies. Image B demonstrates H&E stained section (40x) having hypocellular and hypercellular areas.

Intensive Care Unit (SICU) and was kept there for three days. He was given one transfusion of whole blood in the SICU. The rest of his recovery was uneventful. The histopathology report marked the lesion as a Schwannoma, which showed elongated spindle cells with palisading to form Verocay bodies. [Fig. 2]. The patient was subsequently shifted to the ward from where he was discharged home on the sixth postoperative day after the removal of his drain. He came back for the follow-up a week after discharge when his stitches were removed. He exhibited complete recovery on his follow-up after one month.

Discussion

Incidentaloma is defined as a tumour of the adrenal gland greater than 1cm in diameter, discovered during a radiological investigation performed for reasons other than an adrenal pathology. A stricter definition by the European Society of Endocrinology and European Network for the Study of Adrenal Tumours (ESE/ENSAT) also exclude lesions observed during investigations for hereditary syndromes and extra-adrenal tumours from being labelled as incidentalomas.[6, 7]

The prevalence of these lesions has increased with time due to an increase in the number and improvement in the quality of radiological investigations over time. This is compounded by the fact that the prevalence of incidentalomas on CT has risen from 0.6-1.3% in studies between 1982 and 1986 to 4.8-5% from 2006 to 2008.8 It is also noteworthy that the risk of incidentaloma increases with age and it peaks in the fifth decade of life. It is rare before the age of 30, and if present it is commonly an adrenocortical carcinoma (ACC). This, and the fact that an incidentaloma of more than 4cm in diameter can also be an ACC, makes it imperative that such a lesion be investigated properly before an intervention is planned as our patient also had both these risk factors.[8]

As we know, the adrenal gland consists of Zona Glomerulosa, Zona Fasciculata, Zona Reticularis, and medulla, and these zones chiefly produce Mineralocorticoids (e.g. Aldosterone), Glucocorticoids (e.g. Cortisol), Adrenal Androgens (e.g. Estrogens) and Catecholamines (e.g. Epinephrine) respectively. The lesions in these regions are mainly adenomas of any part of the cortex which is mostly inactive but 10% are functional which may cause Autonomous Cortisol Secretion (Z. Fasciculata) or Hyperaldosteronism (Z. Glomerulosa). The malignant lesion of the adrenal cortex is known as adrenocortical carcinoma whose prevalence is about 2% which may cause symptoms of hypersecretion in all regions of the adrenal cortex. The lesion of the adrenal medulla is called Pheochromocytoma.[8] A Schwannoma is a very rare adrenal incidentaloma and a little more than 40 cases have been reported so far. It is mostly a benign tumour arising from the Schwann cell of the nerves innervating the adrenal medulla, and gradually compressing the adrenal cortex as it enlarges. As the retroperitoneum is a wide space, the tumour usually enlarges to a great extent before it is symptomatic, and, hence, large-sized Schwannomas of the adrenal gland are observed on presentation.[9] The workup of incidentaloma comprises biochemical and radiological investigations. For autonomous cortisol secretion check, the Endocrine Society recommends that two out of the three following investigations should be performed: Dexamethasone suppression test (DST), late-night salivary cortisol (LNSC) levels, and 24-hour urine free cortisol levels (UFC).[8] In this case, serum cortisol level was performed which is not a good investigation to rule out ACS. To rule out Pheochromocytoma, 24-hour urine fractionated Metanephrine levels or plasma Metanephrine should be performed as was done in this case.[8] However, this can be skipped if the lesion is less than 10 Hounsfield units on plain CT.[10] If the patient exhibits hypertension or hypokalaemia, then plasma Aldosterone: Renin ratio should be checked.[6] In cases where a female patient shows signs of virilization or a male shows signs of feminization, it is important to check the levels of sex hormones; these patients should be checked for adrenocortical carcinoma.[6,8] In Schwannoma, this biochemical profile should be within the normal range.

A CT without contrast should be initially performed to look for size, site, and density of the lesion. A schwannoma is a well-circumscribed and homogenous lesion with a mean density of around 30 HU. On MRI it is seen as hypo-intense in T1 and Hyper-intense and heterogeneous in T2 weighted images.[11]

Surgical excision is recommended for lesions more than 4cm in diameter. Our patient did not require any pre-

operative preparation as his pre-operative workup was normal. According to the European guidelines, it is suggested that patients with tumours less than 6cm in diameter, but without local invasion should be offered laparoscopic adrenalectomy.[8] However, this patient had a larger lesion so an open adrenalectomy was performed. Histopathology of a Schwannoma shows two distinct patterns with highly cellular Antoni A region containing Verocay bodies and less cellular Antoni B region and is positive for S100 protein on immunohistochemistry.[11]

Conclusion

A Schwannoma is an extremely rare occurrence in the Adrenal gland and presents with a normal biochemical profile in patients. It can cause symptoms by mass effect and should be surgically removed if it is more than 4cm in size, because of the malignant potential of large Adrenal lesions.

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