

Adrenal Intravascular Papillary Endothelial Hyperplasia

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ABSTRACT

Aim: We describe the rare case of a woman with adrenal intravascular papillary endothelial hyperplasia (IPEH) or Masson's tumor. We present relevant background information on IPEH and our case report, as well as describe a workup and treatment plan for the lesion.

Introduction: Intravascular papillary endothelial hyperplasia (IPEH) or Masson's tumor is a rare lesion, i.e., predominantly found in the oral cavity, head, and neck.¹⁻⁴ To our knowledge, only six adrenal IPEH cases have been reported in the literature to date.⁴

Case report: A 49-year-old woman originally presented to us with an incidentaloma found on computed tomography scan. Due to the eventual size increase of the mass, a laparoscopic left adrenalectomy was performed. Pathologic evaluation of the mass revealed a 3 cm IPEH arising in a hemangioma within the adrenal gland.

Conclusion: Before the diagnosis of IPEH is considered as the etiology for an adrenal incidentaloma, it is crucial to rule out more common tumors. Serological and radiographic studies are critical to the workup.

Clinical Significance: The IPEH is a very interesting lesion of the adrenal gland, however, it is exceedingly rare and thus exclusion of more common and serious lesions must be done prior to the diagnosis of IPEH.

Keywords: Adrenal tumor, Adrenalectomy, Case report, Laparoscopic.

How to cite this article: Holmes C, Akhras A, Schneider A, Yacoub JH, Picken M, Kabaker A. Adrenal Intravascular Papillary Endothelial Hyperplasia. *World J Endoc Surg* 2017;9(1):16-19.

Source of support: Nil

Conflict of interest: None

INTRODUCTION

Originally described by Pierre Masson in 1923, Masson's tumor, now known more accurately as intravascular

papillary endothelial hyperplasia (IPEH), is predominantly found in the oral cavity, head, and neck.^{1,5,6} Only six cases occurring in the adrenal gland have been reported to date, with our case being the 7th.⁴ The IPEH lesions exist in 3 forms: Primary, secondary, and extravascular. Primary IPEH is known to develop within normal blood vessels, most commonly medium-sized vessels, and the secondary variety arises from a vascular malformation (e.g., hemangiomas, arteriovenous malformations).²⁻⁶ The extravascular type, which is the least common, develops from a hematoma.⁵

The pathogenesis of IPEH is not currently known, however, the pattern of growth and the association with other lesions has suggested to some authors that IPEH lesions are not neoplasms, but the result of a reactive process.^{1,2,5,6} The majority of IPEH lesions are found in soft tissue and arise from a thrombosed muscular artery.^{2,4-6} When IPEH lesions are evaluated histopathologically, multiple "fronds and papillae lined by endothelial cells fill up cystic spaces".⁵ The lesions are characterized by blunted papillary projections with hyalinized cores that extrude into vessel lumens (Figs 1A to D).^{2,5} In IPEH, papillary projections lack pleomorphism, anaplasia, increased mitotic activity, necrosis, and disorganization; all of which distinguish IPEH from malignant angiosarcomas and Kaposi's sarcomas.⁵

Patients with IPEH of the most common sites present with asymptomatic growth. However, if the lesion is not superficial, it is often found incidentally on imaging for nonspecific complaints.^{2,4,5} The only described treatment for IPEH is excision, and the vast majority of intra-abdominal IPEH lesions are excised prior to the diagnosis due to the concern for malignancy or as part of a treatment algorithm. Subsequent microscopic examination reveals the true diagnosis.²⁻⁵

CASE REPORT

A 49-year-old female presented to the clinic with an incidental 4.2 cm left adrenal mass initially discovered 9 years prior to presentation on a computed tomography (CT) of the abdomen and pelvis. The mass was followed with serial imaging and displayed significant interval growth of over 1 cm in maximal dimension. The patient was subsequently referred to endocrinology for further evaluation and a very thorough hormonal workup was completed including plasma aldosterone, renin, potassium,

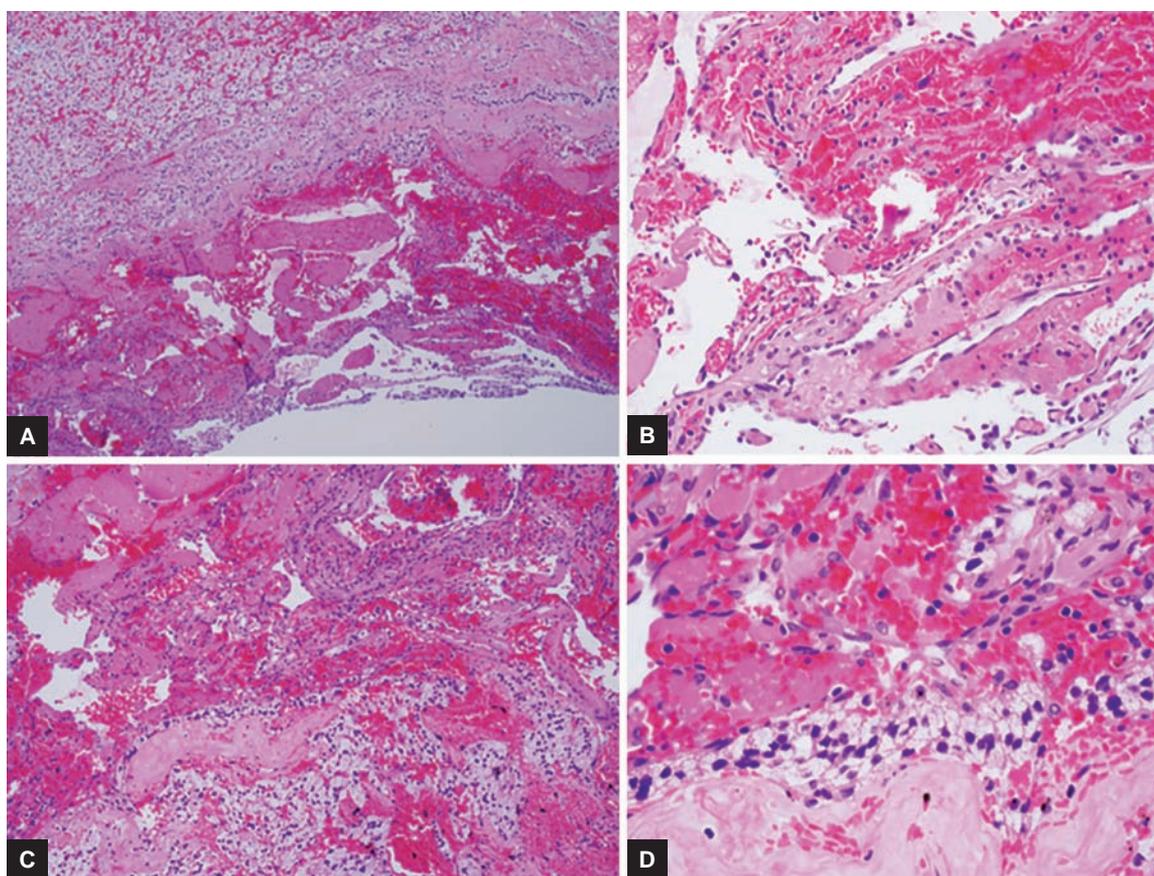
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Figs 1A to D: (A) Relatively circumscribed but unencapsulated and variably cellular lesion is seen located within adrenal cortex (H&E, original magnification 60×); (B) a pseudopapillary arrangement admixed with extravasated red blood cells and fibrin is seen (H&E, original magnification 125×); (C) pseudopapillae are lined by prominent flat, spindle to oval cells, resembling endothelial cells (H&E, original magnification 250×); and (D) at high magnification, flattened spindle cells lining the vascular spaces are seen. All these lining cells were strongly positive for CD31 and CD34 (not shown) (H&E, original magnification 500×)

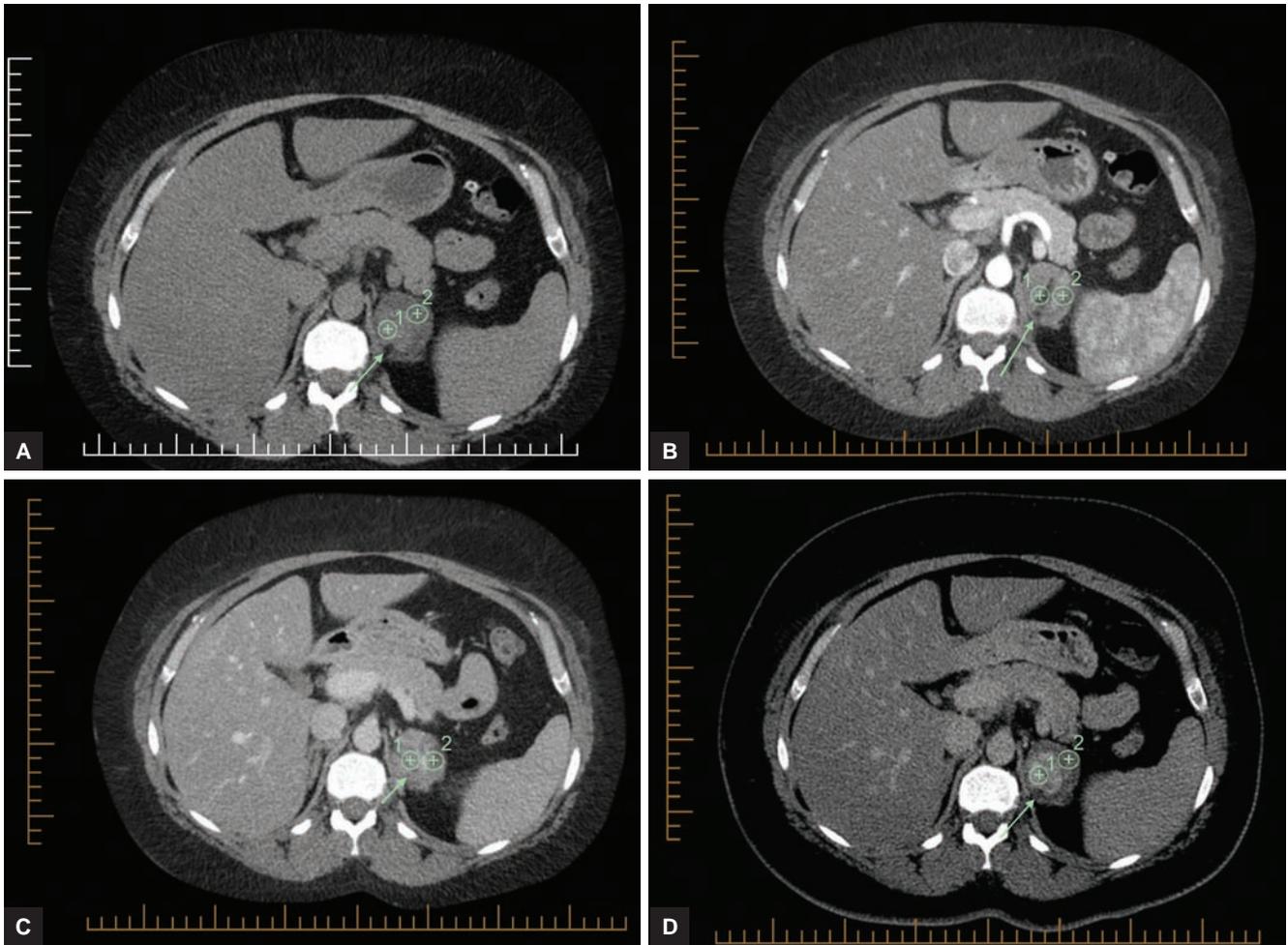
adrenocorticotropic hormone, dehydroepiandrosterone, testosterone, fractionated metanephrine levels, and 24-hour urine creatinine, cortisol, fractionated catecholamines and metanephrines. All these tests returned normal and the patient was referred for a surgical consultation. The patient's medical history consisted of essential hypertension, morbid obesity, nephrolithiasis, diverticulitis, diverticulosis, gastroesophageal reflux disease, iron deficiency anemia, and mononeuritis of the upper limb. Her surgical history was significant for a cesarian section and sigmoid colectomy. Her comorbid conditions were well-controlled with the following medications: Benazepril-hydrochlorothiazide, ibuprofen, metoprolol succinate, omeprazole, potassium chloride, and zolpidem. Of note, the patient does not belong to a known kindred of von hippel-lindau, multiple endocrine neoplasia, neurofibromatosis type 1, Li-Fraumeni or Beckwith--Wiedemann, however, does have a family history of thyroid malignancy in addition to pancreatic cancer, esophageal cancer, diabetes, stroke, and heart disease. The patient had a 20 pack-year smoking history, and no history of drug use. Her examination was consistent with a morbidly obese

woman with no abdominal masses, hernias, or tenderness. Besides obesity, she had no evidence of potential hormonal abnormalities on physical examination. She reported no recent weight loss. Given her thorough functional work-up to date, no additional serology or urine studies were ordered, however, a repeat CT scan of the abdomen and pelvis with and without contrast was obtained in three phases to better characterize the left adrenal mass.

The scan revealed persistent growth of the mass, which possessed indeterminate imaging characteristics (Figs 2A to D). Due to the size increase of the mass and the indeterminate imaging, a laparoscopic left adrenalectomy was recommended. The procedure was completed without incident and the intraoperative findings revealed an encapsulated mass involving the left adrenal gland, which easily separated from the surrounding organs and was removed *en bloc*. Pathologic evaluation of the mass revealed a 3 cm IPEH arising in a hemangioma (Figs 1A to D).

DISCUSSION

The IPEH lesions are rarely found in an intra-abdominal location and are an exceedingly rare etiology of an



Figs 2A to D: (A) Left adrenal nodule is demonstrated on precontrast images; (B) arterial phase; (C) portal venous phase; and (D) three-minute delayed phase. The lesion demonstrates heterogeneous appearance with two primary components. The larger more lateral component has low attenuation on the precontrast images demonstrating hyperenhancement on the arterial phase with washout on the portal venous and delayed phases (ROI 2). This is consistent with the adenoma portion of the lesion. The smaller more medial component has soft tissue attenuation on the precontrast images and demonstrates heterogeneous nodular hyperenhancement on the arterial phase with progressive fill-in and persistent enhancement on the delayed images (ROI 1). This component demonstrates an enhancement pattern, i.e., compatible with a hemangioma. The overall size of the nodule is $4.7 \times 3.2 \times 4.2$ cm. The size of the medial component is $2.5 \times 2.2 \times 3.1$ cm.

adrenal mass.³⁻⁶ In our case, an incidentaloma found on imaging warranted workup for much more common and potentially life-threatening etiologies.^{2,7,8} The evaluation of an adrenal incidentaloma has been well-described and must include a functional workup as well as an appropriate dedicated cross-sectional imaging study. The decision to proceed with surgical excision is based on the risk of malignancy and the results of the functional workup. Usually, any functional mass warrants excision, otherwise its size and imaging characteristics are used to determine the malignant potential. Each case must be evaluated on an individual basis, but in general, nonfunctional masses less than 4 cm with benign characteristics on imaging may be followed with serial imaging and laboratory evaluation.⁶⁻⁸ For nonfunctional lesions in the range of 4 to 6 cm with benign imaging characteristics, short interval monitoring is appropriate. Adrenal masses greater than 6 cm should be excised

after hormonal workup.^{6,7} If the lesion appears to have worrisome imaging characteristics, regardless of the size, then the excision should be considered. This patient's lesion was originally under 4 cm, however possessed a few indeterminate imaging features. The treating physicians at the time recommended a short interval follow-up imaging study, however, the patient was lost to follow-up until several months prior to her presentation.

Radiologic imaging of adrenal masses, specifically density or Hounsfield units (HU) assessment provides critical information in clinical decision making.⁸ The HUs are a measure of attenuation coefficient of X-rays as they pass through tissues. Different tissue types with different attenuation will have different Hounsfield values, with distilled water being zero.⁸ Based on this scale, tissue composition of structures, like masses, for instance, can be inferred from CT images.⁸ The etiology of adrenal masses is often determined with minimal error utilizing

HUs. Adrenal adenomas are defined as measuring <10 HUs; and pheochromocytomas are measured at >100 HUs.^{7,8} In our patient's case, imaging revealed that the mass measured had two main components that behaved differently, (Fig. 2). The lateral peripheral component had low attenuation with HU near zero and demonstrated early arterial hyperenhancement and washout. These characteristics are consistent with an adrenal adenoma. The medial aspect, however, had soft tissue attenuation with HU around 35 and had nodular heterogeneous enhancement on the arterial phase with progressive fill-in and persistent enhancement on the delayed phase. On the 3-minute delayed sequence, it had homogenous enhancement matching that of the blood vessels (blood pool). While this appearance is neither typical and nor specific when seen in the adrenal gland, in other organs, this pattern of enhancement would have suggested a hemangioma. Given the presence of these two components and the atypical appearance of the medial component, this lesion was considered "indeterminate" on the CT.

Biochemical evaluation of masses is critical to the treatment algorithm.⁷ Functional adrenal masses, such as pheochromocytomas and aldosteronomas must be ruled out prior to the operative intervention to avoid the associated morbidities of operating on nonoptimized patients.⁷ Testing for evidence of functional adrenal masses is important not only in narrowing the differential, but also critical for operative planning and preoperative management.^{1,7} Our patient had a complete biochemical workup and was found to have a nonfunctional adrenal mass, however, the change in size and indeterminate imaging characteristics guided our management toward resection of the mass for a tissue diagnosis.

CONCLUSION

The diagnostic assessment of an incidental adrenal lesion requires workup for many malignant and benign diseases.^{1-3,7} Before the diagnosis of IPEH is considered,

it is crucial to eliminate common tumors from the differential. Common lesions, such as cysts, carcinomas (primary or metastatic), and adenomas of the adrenal gland should first be considered.^{2,7} Serological and radiographic studies are crucial to the workup of incidentalomas regardless of which treatment algorithm is followed.

CLINICAL SIGNIFICANCE

The IPEH is a very interesting lesion of the adrenal gland, however, it is exceedingly rare and thus exclusion of more common and serious lesions must be done prior to the diagnosis of IPEH.

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