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Case Report

Unusual appearance of an adrenal ganglioneuroma

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ABSTRACT

Ganglioneuromas are rare tumors that occur spontaneously or arise from a poorly differentiated neuroblastic tumor. Although they are typically described in the pediatric population, they can occur in adults. Ganglioneuromas are often discovered incidentally and their typical imaging appearance, although non-specific, is that of a well-defined solid mass. We are presenting a case of a fat-containing adrenal lesion in a 53-year-old male. The extensive lipomatous changes within the lesion led to the presumption that it represented an adrenal myelolipoma. Pathology revealed a ganglioneuroma with extensive lipomatous changes. This is an uncommon presentation of an adrenal ganglioneuroma mimicking an adrenal myelolipoma. The diagnosis of an adrenal ganglioneuroma raises the possibility of syndromic associations for which patients may undergo genetic testing. We provide a review of typical imaging features of an adrenal ganglioneuroma and provide insight into the situations in which a ganglioneuroma can be suggested as a diagnostic consideration.

Case report

Ganglioneuromas (GN) are rare tumors that may occur spontaneously or arise from a poorly differentiated neuroblastic tumor. Adrenal GNs are often discovered incidentally and their typical imaging appearance, although nonspecific, is that of a well-defined solid mass.

A 53-year-old male with no significant past medical history presented to the emergency department with left lower quadrant pain. The patient reported having fevers and chills and was tachycardic at the time of presentation. No significant laboratory abnormalities were identified. A computed tomography (CT) scan of the abdomen and pelvis with intravenous contrast revealed acute sigmoid diverticulitis and the patient was subsequently treated with conservative

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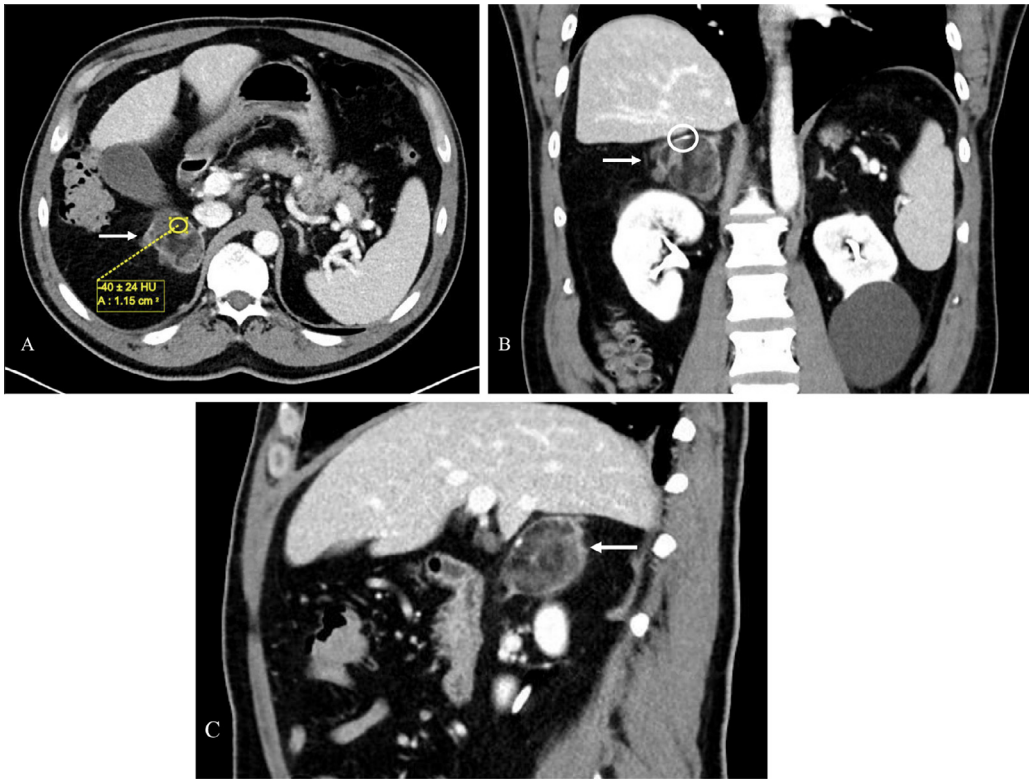

Fig. 1 – Fifty-three-year-old male with incidental right adrenal mass. Axial (a), coronal (b), and sagittal (c) images from contrast enhanced CT scan of the abdomen and pelvis demonstrate a 5.4 cm right adrenal mass (arrows) with a significant volume of macroscopic fat (as indicated by the region of interest on Fig. 1a with -40 internal Hounsfield units), calcifications (circle on Fig. 1b), and a lobulated contour and peritumoral fat stranding. There were intact fat planes between the right adrenal mass and surrounding structures.

medical management. Incidentally, a 5.4 cm right adrenal lesion containing macroscopic fat and peripheral calcifications (Fig. 1) was found. The lesion also had a lobulated contour and perilesional fat stranding. A nonurgent surgical consultation was recommended given the size of the lesion and the presence of atypical imaging features. Adrenal myelolipoma was raised as a potential diagnosis due to the presence of macroscopic fat.

Given the size and the atypical appearance of the adrenal mass, the patient elected to proceed with a laparoscopic right adrenalectomy. Surgery was performed 6 months after the initial discovery of the adrenal lesion. Operative notes remark that the lesion was "rather adherent to surrounding tissue and somewhat inflamed but did not appear malignant." The operation was uneventful, and the patient had a benign postoperative course.

Surgical pathology revealed that the excised lesion was consistent with a GN with extensive lipomatous changes. The resected adrenal gland (85 g) had a well-circumscribed nodule (5.1 × 3.9 × 3 cm) within the adrenal medulla with a tan yellow, soft, and fatty cut surface (Fig. 2a). Microscopically, the tumor was composed of a mixture of Schwann cells and mature ganglion cells, diagnostic features of a GN (Fig. 2c). In addition, there was extensive admixed mature adipose tissue (Fig. 2b). The possibility of an adrenal collision tumor was eliminated as no other distinct histology was present.

Given the association of GNs with hereditary adrenal tumor syndromes such as Multiple Endocrine Neoplasia type 2, succinate dehydrogenase complex iron sulfur subunit variants, and Neurofibromatosis type I the patient was referred to the Genetics Clinic and has met with a genetics counselor. At this time, the patient is considering whether to proceed with genetic testing and the possibility of a syndromic association remains unknown. The patient is otherwise feeling well and is currently asymptomatic.

Discussion

GN is a rare tumor composed entirely of ganglion cells and Schwannian stroma [1]. A GN may occur spontaneously or may be formed by the spontaneous maturation of a neuroblastoma. A neuroblastoma begins as a morphologically undifferentiated or a poorly differentiated neuroblastic tumor [2]. The relative frequency of spontaneously occurring GN and those arising from a neuroblastoma is unknown. The median age at diagnosis is approximately 7 years, although it may occur in all age groups [1,3]. The most common locations are the posterior mediastinum (41.5%), retroperitoneum (37.5%), adrenal gland (21%), and neck (8%) [1,4]. Rarely, GNs are found in the spermatic cord, heart, or gastrointestinal tract [4].

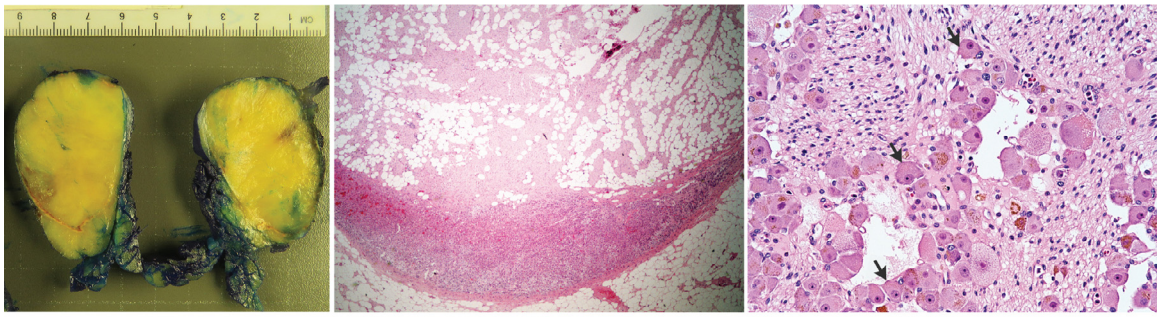

Fig. 2 – Fifty-three-year-old male with incidental right adrenal mass. Cut surface of the adrenal gland with a well-circumscribed tan-yellow soft tumor in the medulla. Notice the rim of compressed adrenal cortex (a). Low power microscopic view of the tumor shows a rim of adrenal cortex adjacent to the tumor which demonstrates extensive lipomatous changes (hematoxylin & eosin, x 20) (b). Higher power view shows numerous ganglion cells (arrows) and schwann cells (c). Some ganglion cells show the accumulation of brown, granular intracytoplasmic pigment representing Nissl substance (hematoxylin & eosin, x 200) (c).

A study of 49 patients with a primary GN found that a GN may present with metabolic activity such as increased secretion of catecholamines and/or metaiodobenzylguanidine uptake [5]. Rarely, GN secretes sufficient quantities of vanillylmandelic acid or homovanillic acid to manifest with symptoms of catecholamine excess, such as flushing [6]. The tumor is most commonly identified incidentally and patients are typically asymptomatic at the time of discovery [3]. However, a GN may result in coughing, abdominal pain, or dyspnea secondary to mass effect [1].

Differentiating a GN from other nonhyperfunctioning adrenal tumors such as adenoma, adrenocortical carcinoma, myelolipoma, or hemangioma may be challenging [3]. A study performed to characterize the appearance of an adrenal GN on CT imaging found that their "typical imaging characteristics" include solid attenuation and postcontrast enhancement, which are nonspecific. Calcifications may be present in a GN while vascular invasion was found to be an uncharacteristic feature [3]. No single defining characteristic has been found to differentiate a GN from an adrenocortical carcinoma. Extensive adipose tissue in GNs is uncommon; moreover, its location in the adrenal gland is unusual as GNs have been reported to more frequently arise in the posterior mediastinum and retroperitoneum [7–10]. A primary diagnostic consideration for an adrenal lesion containing macroscopic fat is an adrenal myelolipoma. Differentiating a GN from an adrenal myelolipoma may be possible based on the presence of macroscopic fat, but discerning the two entities may be challenging in the context of a lipomatous GN.

Our patient presented with an indeterminate right adrenal mass with adrenal myelolipoma as a diagnostic possibility due to the significant amount of macroscopic fat. However, the mass measured over 4 cm (Fig. 1). According to the American College of Radiology (ACR) guidelines, an adrenal mass between the size of 1 and 4 cm with no diagnostic benign imaging features (macroscopic fat, cyst, hemorrhage, nonenhancement, calcifications, or density <10 Hounsfield units) which demonstrates stability for 1 year or longer is most likely benign with no additional work-up required. If an adrenal mass is greater than 4 cm in size without benign diagnostic features and the patient does not have a history of cancer, surgical resection (without biopsy) is recommended to treat a possible primary adrenocortical carcinoma [11].

In summary, GNs are usually asymptomatic masses commonly found within the posterior mediastinum and retroperitoneum, and less commonly arise from the adrenal medulla [12]. The presence of fat is unusual and lipomatous GN is an extremely rare variant first reported in 1999 by Hara et al [8]. Patients diagnosed with a GN carry a good prognosis after surgical resection [12]. We presented a case of a rare adrenal tumor with atypical imaging features which was proven to represent an adrenal GN with an extensive mature adipocytic component following surgical resection.

Patient consent

Patient have no objection to any of the above and give permission for the same.

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