several problems related to its diagnosis, genetic aspects, especially in the absence of a family history. And the criteria of malignancy, given the non-existence to date of a certain & universal criterion that judges the malignancy of the tumor. We present through this article the exceptional case of "bilateral familial pheochromocytomas with strong suspicion of malignancy".

Clinical Case: A 29-year-old lady, hypertensive since 4 years, admitted for exploration of severe secondary hypertension. She reports a triad of menard and severe lumbar pain, the somatic examination shows hypersensitivity of the flanks, a BP of 240/120 mmhg. the abdominal CT scan confirmed by an MRI show a right adrenal mass of 5 cm & another left of 1 cm, of suspicious appearance (irregular contours, areas of necrosis, heterogeneity, spontaneous density at 35 HU, a wash out at 33% with presence of L4 spinal angioma and peri-aortic lymph nodes. urinary methoxylated derivatives (UMD) returned high. The genetic study was positive VPL (on various radiological tests, there was just a spinal angioma) We retained the diagnosis of familal bilateral pheochromocytoma. In front of the clinical & radiological signs of malignancy, we decided to do: a right total adrenalectomy & a left partial adrenalectomy. Unfortunately, the blood pressure didn't drop, and UMD were still positive; the anatomopathological study shows a pheochromocytoma with a PASS score estimated at 3 (benign!) Faced with conflicting clinical-radiological and histological data, we decided to total adrenalectomy without lymph node dissection and to closely monitor the progress. the post Op blood pressure was normalized and the UMD returned negative.

Conclusion: Malignant familial bilateral pheochromocytoma is a very rare & very difficult entity to diagnose, manage & monitor. A good management requires serious collaboration between: endocrinologist, radiologist, urologist, pathologist

Adrenal

ADRENAL CASE REPORTS

Bilateral Pheochromocytoma Due to Von Hippel Lindau With Adrenal-Sparing Adrenalectomy in a Child

Kelly Hilk, BS[1], Anurag K. Agrawal, MD[2], Nicola Cadenas, MS[3], Sanziana Roman, MD[3], Priya S. Srivastava, MD[3], Kevin Yen, MD[3], Maya Beth Lodish, MD[3].

[1]Wayne State University School of Medicine, Detroit, MI, USA, [2]University of California of San Francisco, San Francisco, CA, USA, [3]University of California San Francisco, San Francisco, CA, USA.

Background: More than 40% of pediatric pheochromocytoma or paragangliomas have associated underlying genetic germline mutation. **(1) Clinical Case:** We present an 8-year-old male who arrived the emergency department with hypertension to 170/115. MRI of the abdomen revealed bilateral well demarcated adrenal masses with central necrosis. Urine metanephrines showed elevated normetanephrine of 15244 µg/24 hr (reference range, 58 - 670 µg/24 hr) and normal urine metanephrines. Urinary vanillylmandelic acid was mildly elevated 35 mg/gCr and homovanillic acid was normal. MIBG scan revealed increased radiotracer activity correlating to the bilateral adrenal masses without evidence of metastasis. Diagnosis of bilateral pheochromocytomas was made. Genetic testing revealed a novel, heterozygous, pathogenic variant of *VHL* tumor suppressor gene, consistent with Von Hippel-Lindau syndrome. Perioperative blockade was achieved with prazosin, amlodipine, and metoprolol. Due to low likelihood of metastasis in pheochromocytomas due to VHL, adrenal sparing bilateral adrenalectomy was attempted and resulted in 15% sparing of left adrenal gland vs radial bilateral adrenalectomy. **(2) Clinical Lessons:** 1. Endocrine etiologies of hypertension, although rare, are important causes of hypertension in the pediatric population. 2. Genetic testing prior to surgical intervention could determine surgical course and preservation of adrenals. 3. A multidisciplinary approach to care and referral to a center with experienced surgery, oncology, nephrology, endocrinology, anesthesiology, critical care and genetics is crucial to maximizing outcomes with pheochromocytoma. **Reference:** 1. NGS in PPGL (NGSnPPGL) Study Group, Toledo RA, Burnichon N, Cascon A, Benn DE, Bayley JP, Welander J, Tops CM, Firth H, Dwight T, Ercolino T, Mannelli M, Opocher G, Clifton-Bligh R, Gimm O, Maher ER, Robledo M, Gimenez-Roqueplo AP, Dahia PL. Consensus Statement on next-generation-sequencing-based diagnostic testing of hereditary phaeochromocytomas and paragangliomas. Nat Rev Endocrinol. 2017 Apr;13(4):233–247. 2. King KS, Prodanov T, Kantorovich V, Fojo T, Hewitt JK, Zacharin M, Wesley R, Lodish M, Raygada M, Gimenez-Roqueplo AP, McCormack S, Eisenhofer G, Milosevic D, Kebebew E, Stratakis CA, Pacak K. Metastatic pheochromocytoma/paraganglioma related to primary tumor development in childhood or adolescence: significant link to SDHB mutations. J Clin Oncol. 2011 Nov 1;29(31):4137–42.

Adrenal

ADRENAL CASE REPORTS

Biotin Supplementation Creates the Misleading Diagnosis of Secondary Adrenal Insufficiency

Hooman Motahari, MD, Soumya Thumma, MD, Lakshmi Menon, MD.

University of Arkansas for Medical Sciences, Little Rock, AR, USA.

Introduction: Biotin (vitamin B7) is a water-soluble vitamin and an essential cofactor for the metabolism of fatty acids, glucose, and amino acids. Cases of biotin interference with laboratory testing have been described, most of which involve interference with thyroid function tests. Interference with gonadal steroids, adrenal, and pituitary hormones are rare. We report a case of T3 thyrotoxicosis in which biotin supplementation created the appearance of secondary adrenal insufficiency (AI).

Case: A 66-year-old woman was referred for the evaluation of low TSH. She had chronic fatigue, low libido, and dizziness on standing. Vitals were stable with BP 135/64 mmHg and BMI 23.5. No evidence of mucosal or cutaneous hyperpigmentation. Laboratory evaluation revealed low ACTH <5 (7.2–63.3 pg/mL), low morning cortisol 3.8 and high DHEA-S 174 (13–130 ug/dL). TSH was low at 0.32 (0.32–5.60 uIU/mL) with normal prolactin and