Radiology Case Reports 12 (2017) 343–347

Available online at www.sciencedirect.com

ScienceDirect

journal homepage: http://Elsevier.com/locate/radcr

Case Report

Extra-adrenal pheochromocytoma at the organ of Zuckerkandl: a case report and literature review

Tarana Gill BA[a,*], *Kalie Adler DO*[b], *Alicia Schrader DO*[b], *Keyur Desai BA*[a], *Joshua Wermers DO*[b], *Nebiyu Beteselassie MD*[b,c]

[a] *University of Missouri-Kansas City School of Medicine, Kansas City, MO, USA*
[b] *Diagnostic Radiology, University of Missouri-Kansas City, Kansas City, MO, USA*
[c] *Department of Radiology, Truman Medical Center, Kansas City, MO, USA*

ARTICLE INFO

Article history:
Received 12 September 2016
Received in revised form
1 December 2016
Accepted 19 December 2016
Available online 21 March 2017

Keywords:
Paraganglioma
Pheochromocytoma
Zuckerkandl
Octreotide
Neuroendocrine

ABSTRACT

Pheochromocytomas and paragangliomas are tumors that occur in characteristic locations and are commonly detected on imaging studies. A correct diagnosis is important because of differences in associated neoplasms, risk for malignancy, and need for genetic testing. In addition, associated complications, including death, can be avoided if appropriately recognized and treated. Here, we report a rare case of an extra-adrenal paraganglioma of the organ of Zuckerkandl.

Introduction

Pheochromocytomas (PCCs) and paragangliomas (PGLs) are rare catecholamine secreting neuroendocrine tumors. The combined estimated annual incidence in the United States is approximately 500-1600 cases per year [1]. Both tumors classically present with paroxysmal attacks of headache, palpitations, and diaphoresis. During these episodes of catecholamine release, blood pressure is typically highly elevated and labile. Eighty-five percent of these masses occur in the adrenal medulla; however, 15% occur extra-adrenally along the sympathetic chain [2]. Most extra-adrenal disease occurs in the subdiaphragmatic region, most commonly within the organ of Zuckerkandl. Because of their varied clinical presentations, imaging, and pathologic appearances, accurate diagnosis can be challenging; however, appropriate and timely diagnosis and treatment is essential in preventing severe complications such as myocardial infarctions, strokes, and death.

Competing Interests: The authors have declared that no competing interests exist.

* *Corresponding author.*
E-mail address: tkgrh2@mail.umkc.edu (T. Gill).

http://dx.doi.org/10.1016/j.radcr.2016.12.009

Case report

A 52-year-old male patient with a known history of coronary artery disease and past myocardial infarction presented to the emergency department with complaints of a 2-month history of nonradiating midsternal chest tightness on exertion. In addition, he endorsed weakness, palpitations, and a 4–5-year history of diffuse abdominal pain. The patient was found to be tachycardic with a labile blood pressure, ranging from 147-183 systolic and 91-133 diastolic. Abdominal examination was unremarkable except for tenderness on palpation of the right upper quadrant and left lower quadrant. The patient was admitted for acute coronary syndrome rule out. Electrocardiogram showed normal sinus rhythm with no ST changes and negative troponins.

Imaging was obtained to further evaluate the abdominal pain. Computed tomography (CT) imaging of his abdomen revealed a heterogeneously enhancing soft tissue mass within the retroperitoneum between the infrarenal abdominal aorta and the inferior vena cava, which measured approximately 4.0 × 4.4 × 4.3 cm. Differential considerations included primary malignancy, metastatic disease, or an extra-adrenal PCC (Figs. 1-4).

Laboratory work revealed an elevated urine 24-hour normetanephrine (5555, normal 122-676), urine metanephrines and normetanephrines (5665, normal 224-832), and 24-hour urine metanephrines were normal (210, normal 90-315). Endocrinology was consulted and ordered a magnetic resonance imaging (MRI) for further evaluation.

Further workup with an MRI of the abdomen with contrast was obtained for further characterization. It revealed the 4.0 × 4.4 × 4.3 cm T1 hypointense, T2 hyperintense heterogeneously enhancing mass within the retroperitoneum along the distal abdominal with subtle scattered areas of restricted diffusion. In combination with clinical findings, it was thought that this could relate to an extra adrenal PCC within the organ of Zuckerkandl. Other differential possibilities included hypervascular metastases.

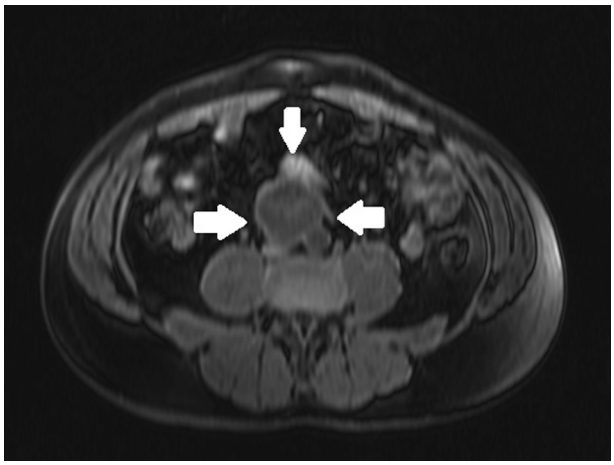

Fig. 1 – Axial unenhanced T1 fat saturation-weighted magnetic resonance (MR) image shows a heterogeneous retroperitoneal mass (arrows) adjacent to the distal abdominal aorta. The mass results in mass effect on the inferior vena cava.

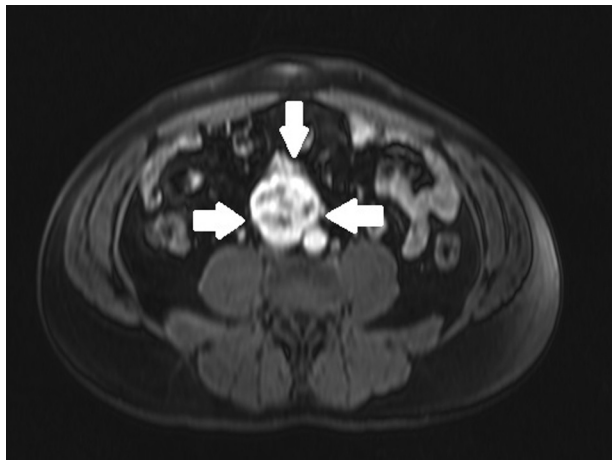

Fig. 2 – Axial contrast enhanced T1 fat saturation-weighted MR image demonstrates a mass (arrows) with peripheral enhancement with central nonenhancement.

A whole body Indium 111 octreotide nuclear medicine scan was then performed along with single-photon emission computed tomography (SPECT) in which a focus of abnormal radiotracer uptake in the mid abdomen in the region of the previously described mass. Although, SPECT/CT is superior to SPECT in terms of diagnostic accuracy and localization of neuroendocrine tumors, that technology is not available at this institution [1]. Thus, through SPECT, the patient was diagnosed with an extra-adrenal PGL. He was then transferred to the surgical intensive care unit for blood pressure control before surgical removal.

The patient later underwent exploratory laparotomy with exploration of the retroperitoneum. A soft brown-tan mass

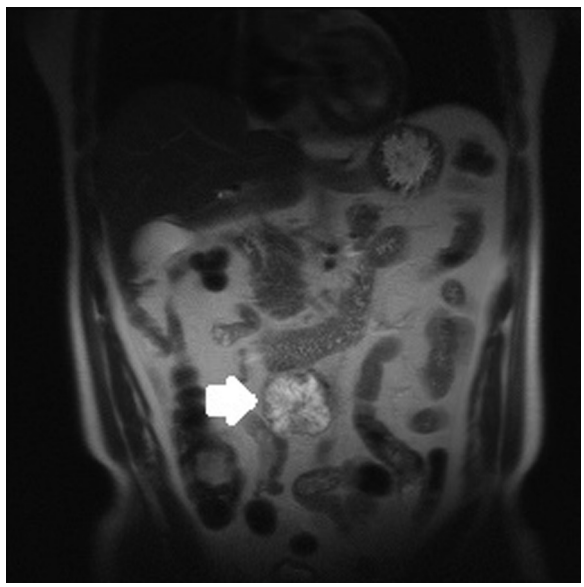

Fig. 3 – Coronal T2-weighted half-Fourier acquisition single-shot turbo spin-echo (HASTE) image demonstrates a predominately hyperintense retroperitoneal mass (arrow).