

Pheochromocytoma of the organ of Zuckerkandl

Unusually small lesion detected with magnetic resonance imaging

Khaled M. Elsayer, MD, MS, John R. Leyendecker, MD, BS, Vamsidhar R. Narra, MD, FRCP, Jeffrey J. Brown, MD, FACP.

ABSTRACT

A case report of a 40-year-old female with an unusually small pheochromocytoma of the organ of Zuckerkandl is discussed. The tumor is diagnosed by magnetic resonance imaging (MRI) examination, which was requested to evaluate her as a potential renal donor. There is a family history of Carney's triad (gastric leiomyosarcoma, extra-adrenal pheochromocytoma and pulmonary chondroma). The MRI technique and findings are discussed.

Saudi Med J 2005; Vol. 26 (1): 107-110

Pheochromocytomas are rare tumors, which classically present with hypertension, although symptoms may be nonspecific or absent. Early diagnosis of pheochromocytoma is important, as these lesions are amenable to surgical excision but are potentially harmful if missed.

The diagnosis of pheochromocytoma is primarily based on clinical presentation and the presence of urinary or serum catecholamines. Imaging is performed to detect and localize the tumor in patients with clinical signs and symptoms suggestive of pheochromocytoma. Magnetic resonance imaging (MRI), computed tomography (CT) scanning, and meta-iodobenzylguanidine (MIBG) radionuclide imaging have all been used to evaluate these lesions. The sensitivity for lesion

detection is high for adrenal pheochromocytomas. However, lesion detection is more challenging for small extra-adrenal tumors.

We report a case in which an asymptomatic pheochromocytoma involving the organ of Zuckerkandl was detected with MRI. The pheochromocytomas in this location are typically large at the time of diagnosis. This case is unusual due to the small size of the lesion at the time of diagnosis and its precise localization to the organ of Zuckerkandl.

Case Report. A 40-year-old woman who was referred to MRI as part of her evaluation as a potential renal donor for her brother with diabetic

From the Mallinckrodt Institute of Radiology (Elsayer, Narra, Brown), Washington University School of Medicine at Saint Louis, and the Department of Radiology (Leyendecker), Wake Forest University School of Medicine, St. Louis, Missouri, United States of America.

Received 25th July 2004. Accepted for publication in final form 27th September 2004.

Address correspondence and reprint request to: Dr. Khaled M. Elsayer, Mallinckrodt Institute of Radiology, 510 South Kingshighway Blvd., St. Louis, Missouri 63110, United States of America. Tel. +1 (314) 3622967. Fax. +1 (314) 3624550. E-mail: elsayerk@mir.wustl.edu

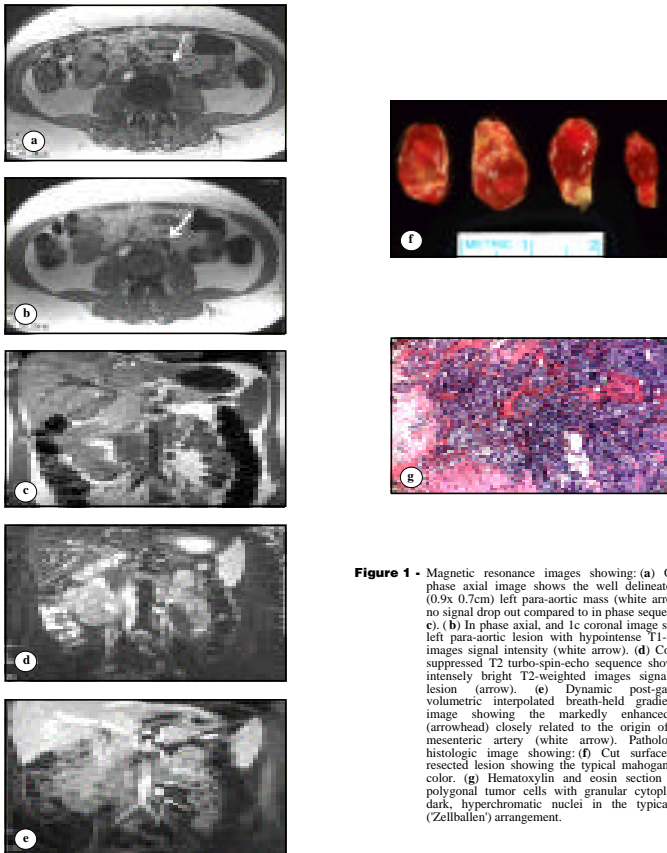


Figure 1 - Magnetic resonance images showing: (a) Opposed-phase axial image shows the well delineated small (0.9x 0.7cm) left para-aortic mass (white arrow) with no signal drop out compared to in phase sequence (b & c). (b) In phase axial, and (c) coronal image shows the left para-aortic lesion with hypointense T1-weighted images signal intensity (white arrow). (d) Coronal fat suppressed T2 turbo-spin-echo sequence showing the intensely bright T2-weighted images signal of the lesion (arrow). (e) Dynamic post-gadolinium volumetric interpolated breath-held gradient echo image showing the markedly enhanced lesion (arrowhead) closely related to the origin of inferior mesenteric artery (white arrow). Pathologic and histologic image showing: (f) Cut surface of the resected lesion showing the typical mahogany brown color. (g) Hematoxylin and eosin section showing polygonal tumor cells with granular cytoplasm and dark, hyperchromatic nuclei in the typical nested ('Zellballen') arrangement.

nephropathy. She had 2 sisters who had been diagnosed with pheochromocytomas, a pattern that suggests familial pheochromocytoma. Genetic testing for von Hippel Lindau disease and multiple endocrine neoplasia type 2 was negative. She did not have a history of hypertension. Urinary catecholamine levels were within normal limits. Magnetic resonance imaging was performed at 1.5 T (Siemens Medical Systems, Erlangen, Germany). The imaging protocol included coronal T2-weighted half-Fourier single shot turbo-spin-echo (HASTE) images, transaxial in-phase and opposed-phase gradient echo images, transaxial turbo-spin-echo (TSE) T2-weighted images, and 3D volumetric interpolated breath-held gradient echo (VIBE) images. Volumetric interpolated breath-held gradient echo images were acquired before, and at 25 seconds, 60 seconds and 120 seconds after intravenous contrast material administration at a dose of 0.1 mmol/kg (Omniscan, Amersham Health, Princeton, New Jersey).

Half-Fourier single shot turbo-spin-echo images were acquired with the following parameters a pulse repetition time (TR) of 4450 msec, a echo delay time (TE) of 59 msec, and a slice thickness of 6 mm with no interslice gap. T2-weighted fast spin echo (FSE) images were acquired with a TR of 2485 msec, a TE of 95 msec, and frequency selective fat saturation. In-phase and opposed-phase images were acquired with a single double echo gradient sequence with a TR of 205 msec and TE values of 2.4 msec for the opposed-phase images and 4.2 msec for the in-phase images. The slice thickness was 4 mm with no gap. Volumetric interpolated breath-held gradient echo images were acquired with a TR of 7.6 msec and a TE of 3 msec. Magnetic resonance images showed a small 0.9 x 0.7 cm left para-aortic mass (Figure 1). The mass showed no evidence of signal loss on opposed phase images (Figure 1a) compared to in-phase images (Figures 1b & 1c). The mass was hypo-intense on T1-weighted images and very bright on T2-weighted images (Figure 1d). On dynamic post-contrast images, the mass showed marked contrast enhancement. The arterial phase VIBE sequence confirmed the left para-aortic location of the mass immediately adjacent to the origin of the inferior mesenteric artery (Figure 1e). Gross pathology and histologic characteristics were typical for an extra-adrenal paraganglioma (Figures 1f & 1g). The adrenal glands were normal in appearance. On the basis of the MRI findings and the clinical history, the patient underwent surgical resection of the left para-aortic mass, which was found at pathology to represent a pheochromocytoma.

Discussion. Pheochromocytomas are a subclass of paragangliomas. Paragangliomas arise

from specialized neural crest cells usually within the adrenal gland or sympathetic chain. Paragangliomas have been classified according to location as branchiomic, intravagal (upper mediastinal), aortic sympathetic (retroperitoneal), and visceral (pelvic, vagal, mesenteric).¹ In adults, sympathetic ganglia are widely distributed and are particularly numerous along the fibers of the hypogastric plexuses involving the urogenital organs and within the sacral plexus. These are not generally known by individual names, and their precise locations vary. Two exceptions are the adrenal medulla and the organ of Zuckerkandl.² Some investigators have loosely used the plural (organs of Zuckerkandl) to include all para-aortic ganglia between the origin of the inferior mesenteric artery and the aortic bifurcation. However, in its narrowest sense, the organ of Zuckerkandl is located adjacent to the origin of the inferior mesenteric artery.^{2,4} Although, sensitivities for detecting unilateral adrenal pheochromocytomas with CT and MRI have been reported to be as high as 100%, extra-adrenal paragangliomas present a greater diagnostic challenge.³ The sensitivities of CT for detecting extra-adrenal paragangliomas was 64%, 88% for MRI and 64% for MIBG, in a large series.⁵ In another report, 75% of extraadrenal paragangliomas were detected prospectively with MRI.⁶ The missed lesions in this series were all less than 4 cm in size. We expect that the lesion detection sensitivity could be greatly improved using state-of-the-art MRI performed with high spatial resolution sequences. However, further data are needed to support this view.

There are relatively few reports of the pheochromocytoma in MRI involving the organ of Zuckerkandl. Terk et al⁷ described a lesion that may have arisen from this location, however, precise localization was difficult due to the large size of the tumor. The typical features of pheochromocytomas on MRI include bright signal intensity on T2-weighted images and marked contrast enhancement.⁶ However, these imaging features are not specific for pheochromocytoma, and should be considered in the context of the clinical and laboratory data. In the present case, both the T2-weighted, fat suppressed images and the contrast-enhanced dynamic images clearly demonstrated the lesion. In addition, TSE and HASTE images were equally effective for lesion detection and localization. The lesion appears very bright on the fat suppressed T2-weighted images and shows marked contrast enhancement. The use of VIBE images in this case, permitted high resolution dynamic contrast enhanced imaging during several phases of enhancement. The high spatial resolution of the VIBE technique was useful for delineating this small tumor. The dynamic post contrast images helped characterize the

enhancement pattern of the lesion, and the arterial phase VIBE sequence allowed precise localization of the tumor adjacent to the inferior mesenteric artery origin.

Acknowledgment. We thank Dr. James S. Lewis Jr, Department of Surgical Pathology, Washington University at Saint Louis and Dr. Jeffrey Moley, Department of Surgery, Washington University at Saint Louis, United States of America, for providing surgical and pathological information.

References

1. Sessions RB, Harrison LB, Forastiere AA. Paragangliomas. In: Devita VT Jr, Hellman S, Rosenberg SA, editors. *Principles and Practice of Oncology*, 5th ed. Philadelphia (PA): Lippincott-Raven; 2001. p. 900.
2. Tiscler AS. Paraganglia. In: Sternberg SS, editor. *Histology for pathologists*. New York (NY): Raven; 1997. p. 1154.
3. Amparo EG. Where is the organ of Zuckerkandl? *Am J Roentgenol* 1993; 160: 662.
4. Zuckerkandl E. Ueber Nebenorgane des sympathicus im retroperitonealraum des menschen. *Verh Anat Ges* 1901; 15: 85-107.
5. Jalil ND, Pattou FN, Combemale F, Chapuis Y, Henry JF, Peix JL, et al. Effectiveness and limits of preoperative imaging studies for the localization of pheochromocytomas and paragangliomas: A review of 282 cases. *Eur J Surg* 1998; 164: 23-28.
6. Van Gils ABG, Falke THM, Van Erkel AR, Arndt JW, Sandler MP, van der Mey AG, et al. MR imaging and MIBG scintigraphy of pheochromocytomas and extra-adrenal functioning paraganglioma. *Radiographics* 1991; 11: 37.
7. Terk MR, de Verdier D, Colletti PM. Giant extra-adrenal pheochromocytoma: Magnetic resonance imaging with gadolinium-DTPA enhancement. *Magn Reson Imaging* 1993; 11: 47-50.