



SAR·DFP

Society of Abdominal Radiology
Disease-Focused Panels



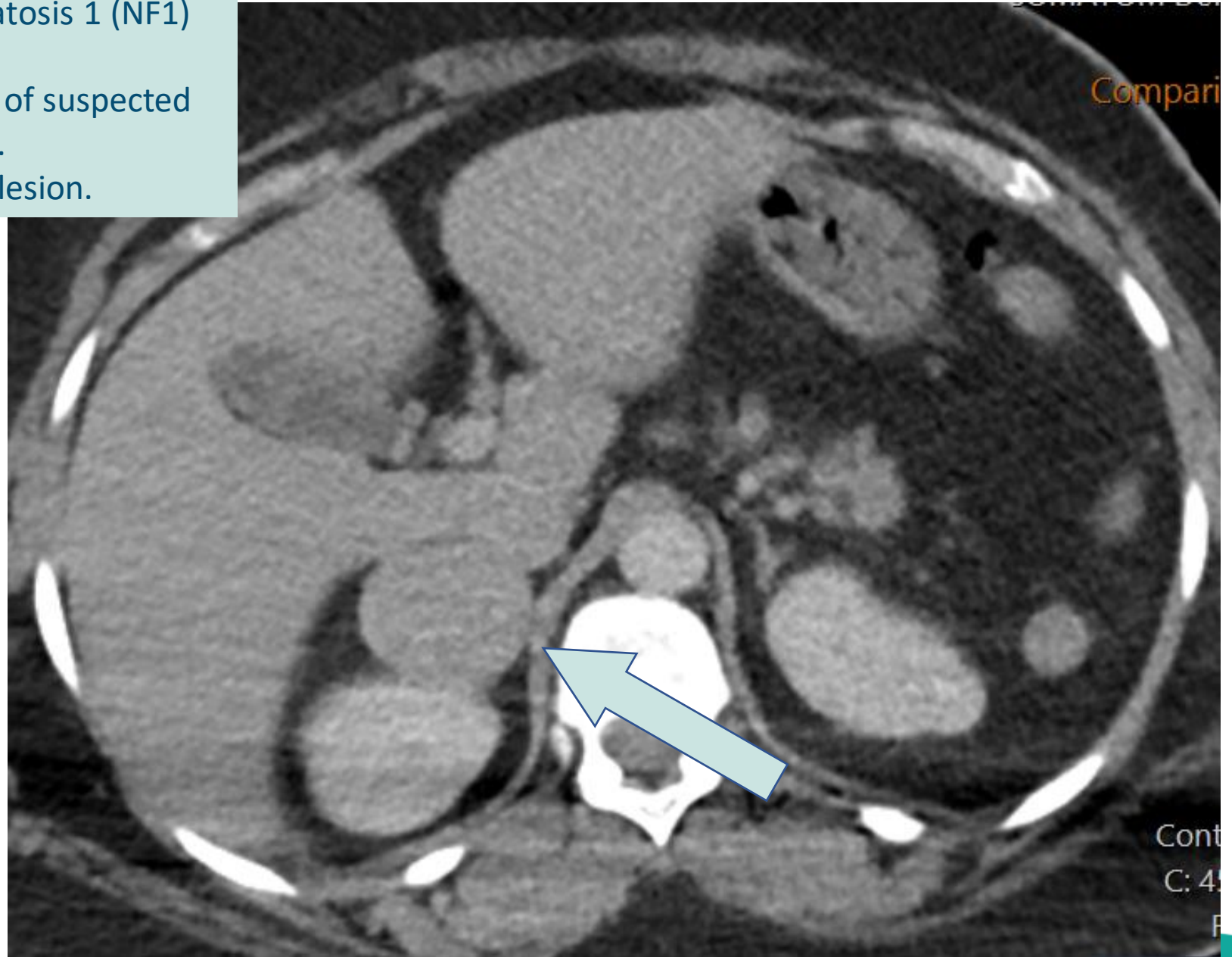
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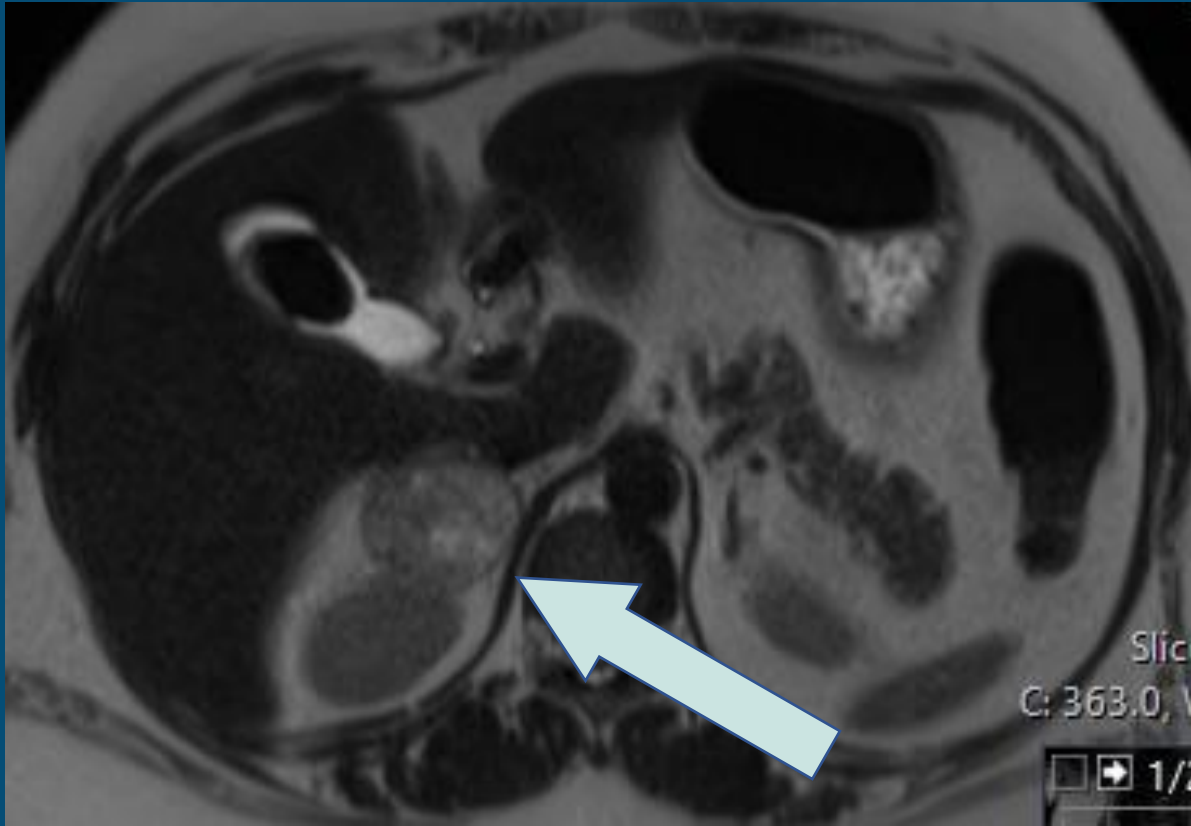
Pheochromocytoma in Neurofibromatosis¹

Presented By: Malak Itani MD

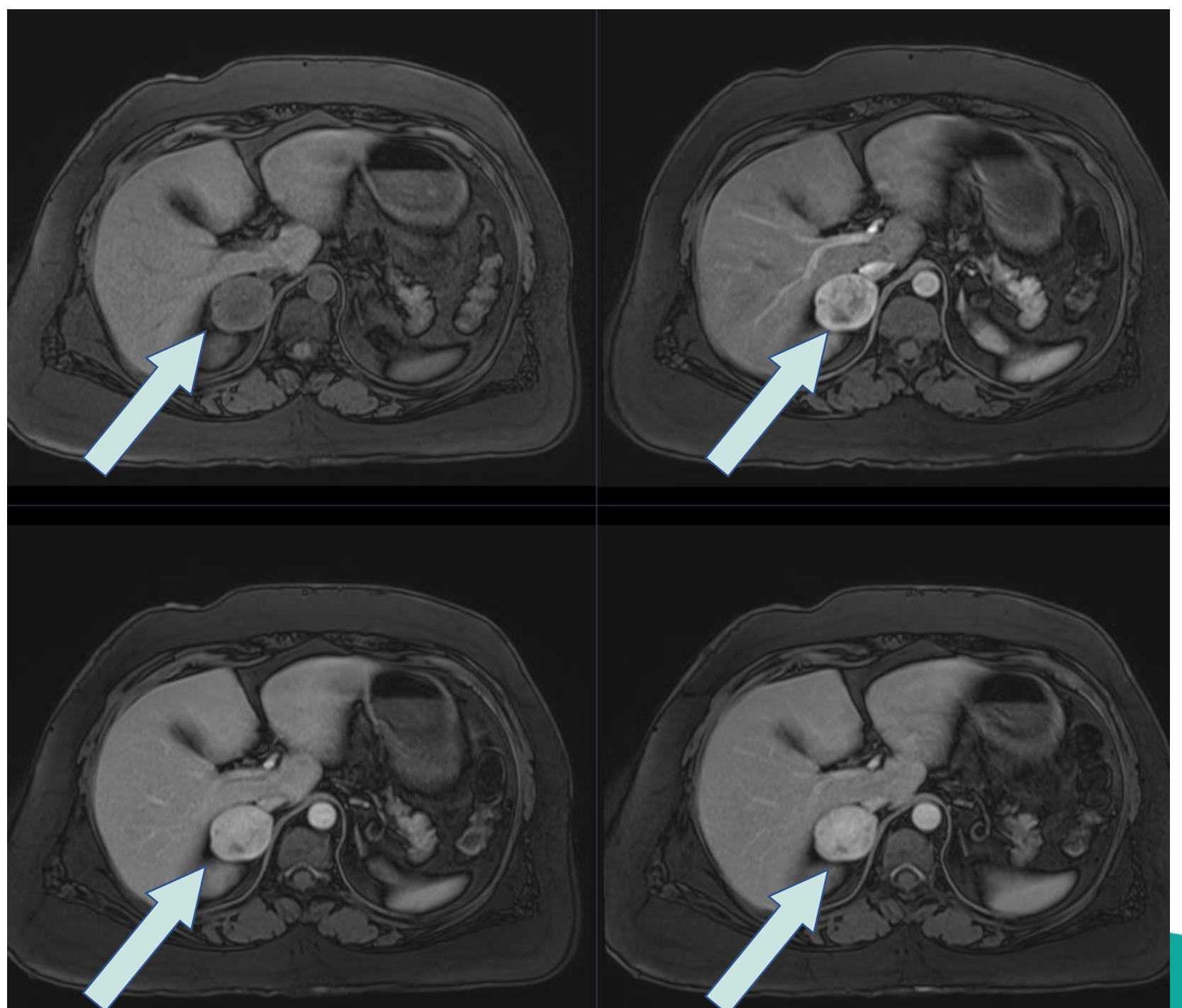
46-year-old female with neurofibromatosis 1 (NF1) presented with right facial drooping. CT scan of the chest was done as part of suspected stroke imaging protocol during COVID. There was an incidental right adrenal lesion.



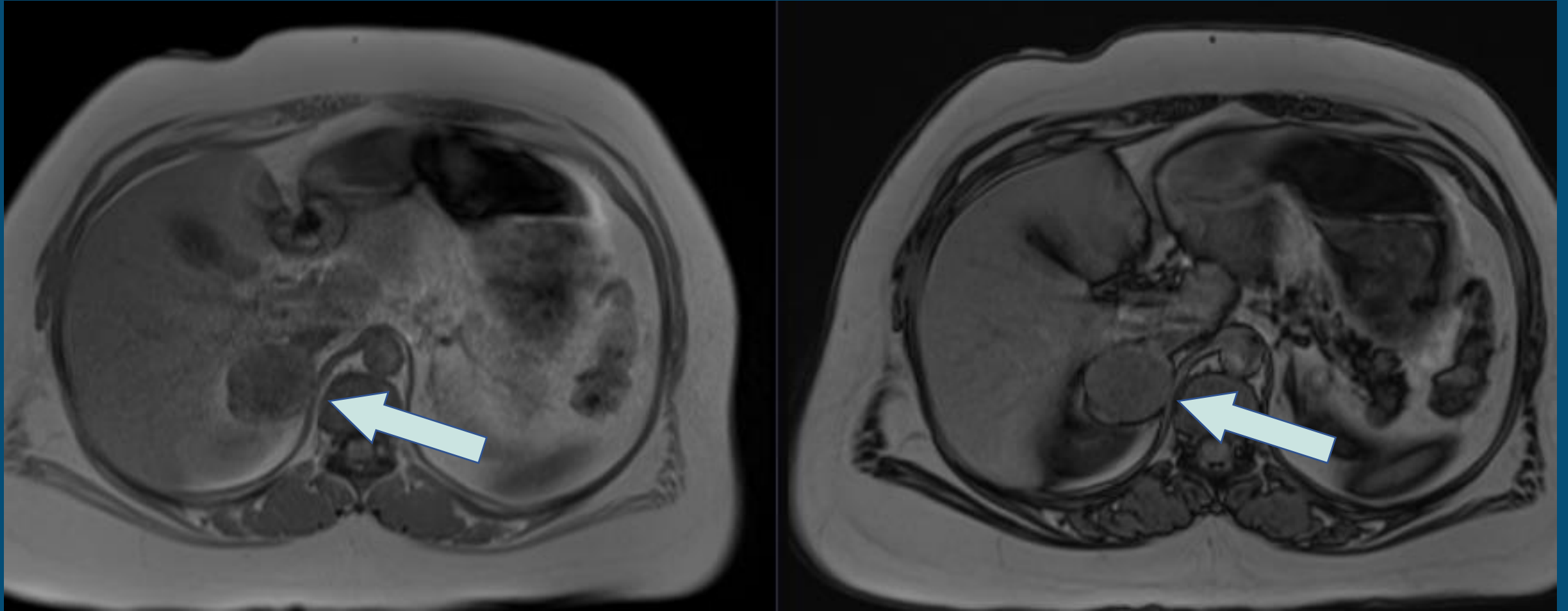
MRI of the abdomen was performed for further characterization. Axial and coronal T2-HASTE images demonstrate a heterogeneous lesion with intermediate to high-signal intensity on T2 weighted images.



Multiphase fat-suppressed T1 weighted images (VIBE) demonstrate intense, early, heterogeneous enhancement.

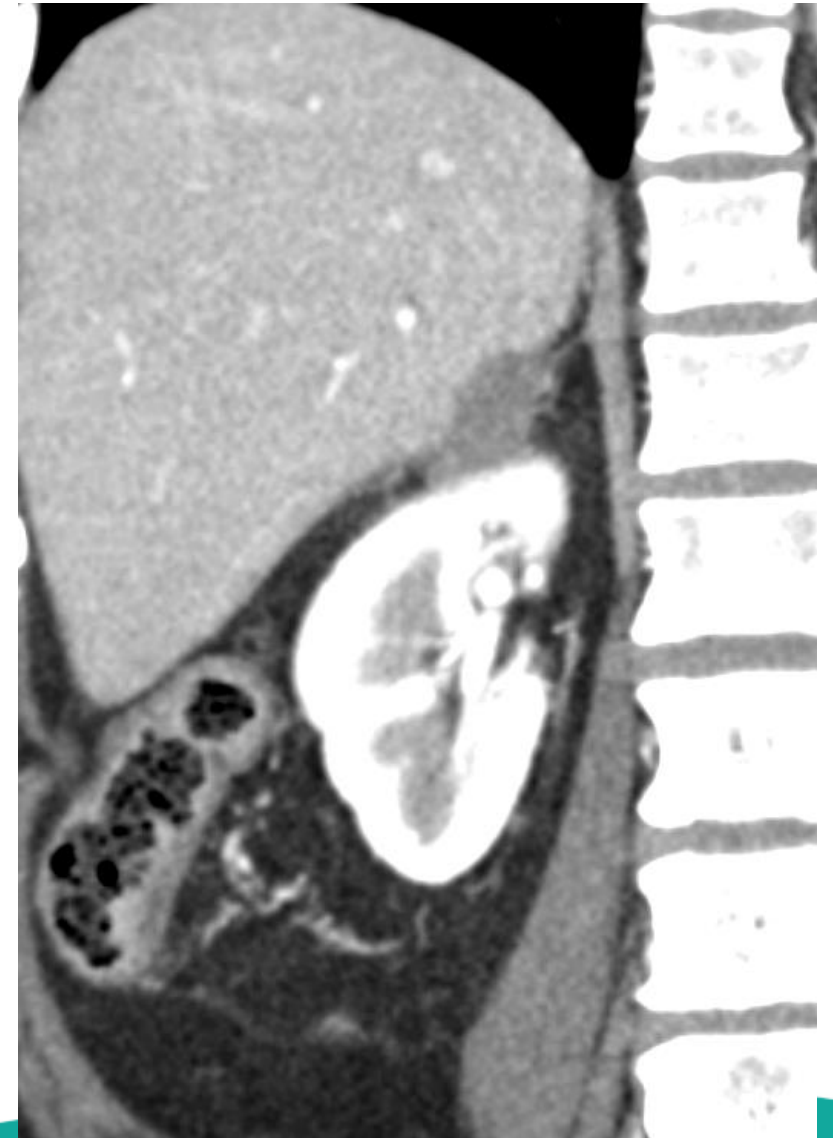


Axial in-phase and opposed-phase images show no evidence of intravoxel fat or iron.



Imaging Interpretation and Surgical Pathology

- Given MRI findings, and the patients history of NF1, the radiologic diagnosis was pheochromocytoma.
- The right adrenal lesion was resected and surgical pathology was consistent with pheochromocytoma.



Discussion

- Neuroendocrine tumors are associated with some of the phakomatoses, including neurofibromatosis type 1 (NF1).
- NF1 affects 1 in 4,000 individuals*.
- Patients with NF1 are at increased risk of tumors, including neuroendocrine tumors such as pheochromocytomas, paragangliomas, and pancreatic neuroendocrine tumors*.
- The life-time risk for developing pheochromocytoma is 1-5% in patients with NF1*, compared to the overall prevalence of 2 per 100,000**.



* Lodish MB, Stratakis CA. Endocrine tumours in neurofibromatosis type 1, tuberous sclerosis and related syndromes. *Best Pract Res Clin Endocrinol Metab.* 2010;24(3):439-449. doi:10.1016/j.beem.2010.02.002

** Kim JH, Moon H, Noh J, Lee J, Kim SG. Epidemiology and Prognosis of Pheochromocytoma/Paraganglioma in Korea: A Nationwide Study Based on the National Health Insurance Service. *Endocrinol Metab (Seoul).* 2020;35(1):157-164. doi:10.3803/EnM.2020.35.1.157

Discussion

- In cases of suspected pheochromocytoma, biochemical testing is usually performed by evaluating serum and urine metanephrines.
- On CT, pheochromocytomas enhance avidly, which causes them to have a high wash-out, mimicking adenomas on adrenal protocol studies, although they tend to be more heterogeneous.
- On MRI, the classic descriptor is the “light-bulb” appearance due to their high signal intensity on T2 weighted images.
- Functional imaging, e.g. with DOTATATE PET/CT, can help localize pheochromocytomas if anatomic imaging is non-revealing in the setting of elevated metanephrines.

THANK YOU

