

Wilm tumor and Vasculitis

Heather Borders, MD

06/19/2012

History

Eleven year old female was being followed for a left renal lesion. After approximately four years, the patient developed a large left renal mass.

Diagnosis

Wilm tumor and Vasculitis

Discussion

Wilms tumor accounts for 87% of pediatric renal masses. Its peak incidence is at 3–4 years of age. Patient age in this case is outside of the typical range. Renal cell carcinoma is also a consideration in this age group.

Malignancy is found in around 5% of patients with vasculitis. The reason for this association is unclear, although postulated mechanisms include immune complex formation with tumour antigens, shared tumour and vessel wall antigens, and impaired clearance of normally produced immune complexes. Additionally, some research suggests a viral etiology.

In this patient the vasculitis was present prior to the development of Wilm tumor. The vasculitis has features of polyarteritis nodosa, such as multiple aneurysms and stenoses of medium size vessels but lack of renal artery involvement is unusual.

Polyarteritis nodosa (PAN) is a systemic illness characterized by necrotizing inflammation of small- and medium sized arteries, leading to aneurysm formation. It is associated with hepatitis B virus (HBV) in about 7% of cases. The most common clinical symptoms are persistent fever, weight loss, and polyarthralgia (the patient in this case did not report these symptoms).

PAN is rare in childhood and represents one of the rarest vasculitides in children. Ozen et al. defined musculoskeletal and renal involvement as the major diagnostic criteria for PAN in childhood. Ten additional minor criteria were also defined: (1) cutaneous findings, (2) gastrointestinal involvement, (3) peripheral neuropathy, (4) central nervous system involvement, (5) hypertension, (6) cardiac involvement, (7) lung involvement, (8) constitutional symptoms, (9) presence of acute-phase reactants, and (10) presence of hepatitis B surface antigen. They proposed that the presence of five of these criteria, including at least one major criterion, is highly suggestive of PAN in childhood. The patient in this case does not meet all of these criteria, but the appearance and involvement of the mesenteric vessels still suggests PAN as the most likely diagnosis.

Findings

Large hypodense mildly heterogeneous and well circumscribed left renal mass without metastatic disease.

Abnormal mesenteric vasculature on CT and angiogram; multiple aneurysms and stenoses involving medium size vessels. Celiac axis, SMA were involved and IMA was occluded. Renal arteries were spared.

Reference

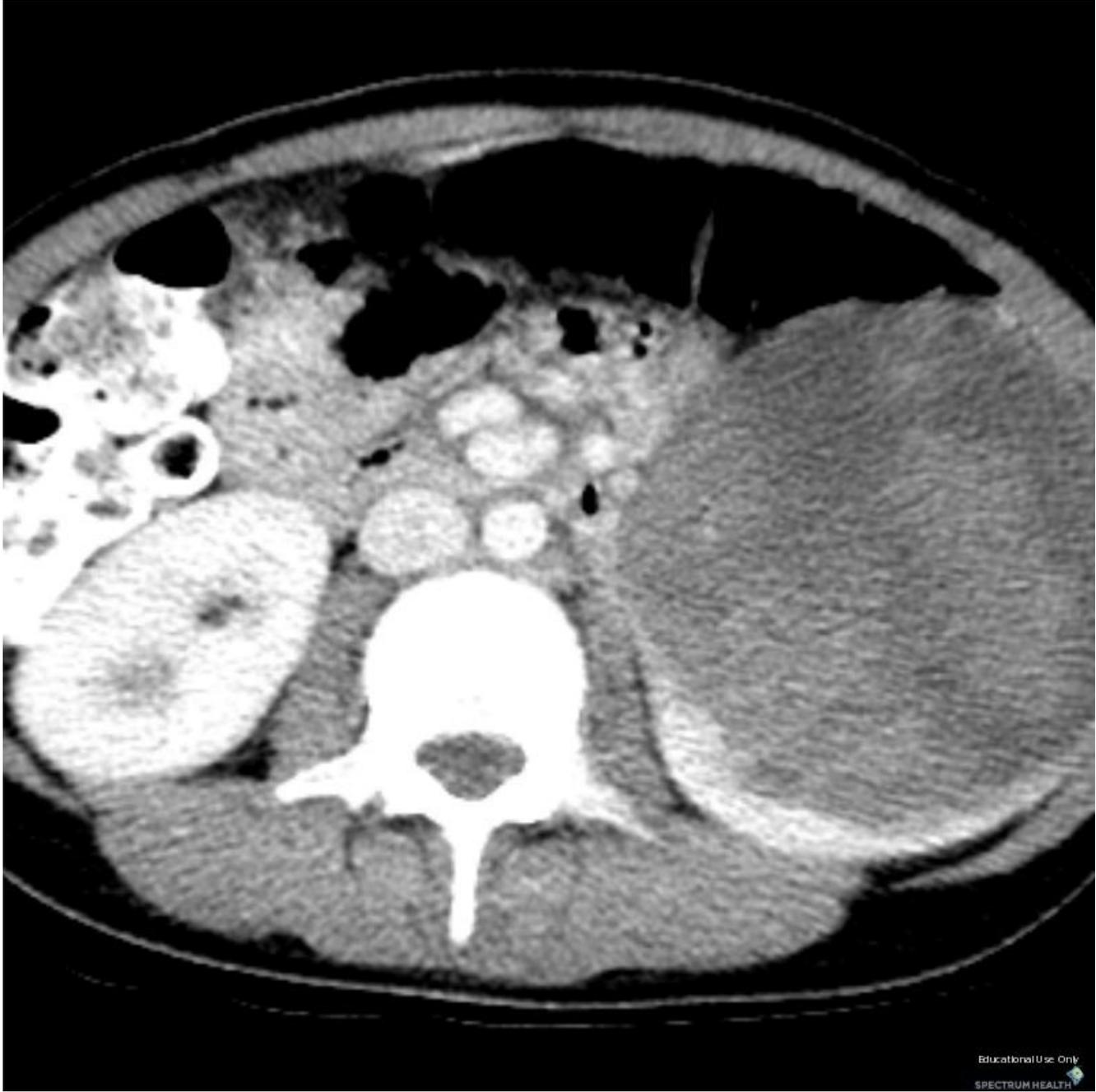
Pediatric Renal Masses: Wilms Tumor and Beyond Lisa H. Lowe, MD. November 2000

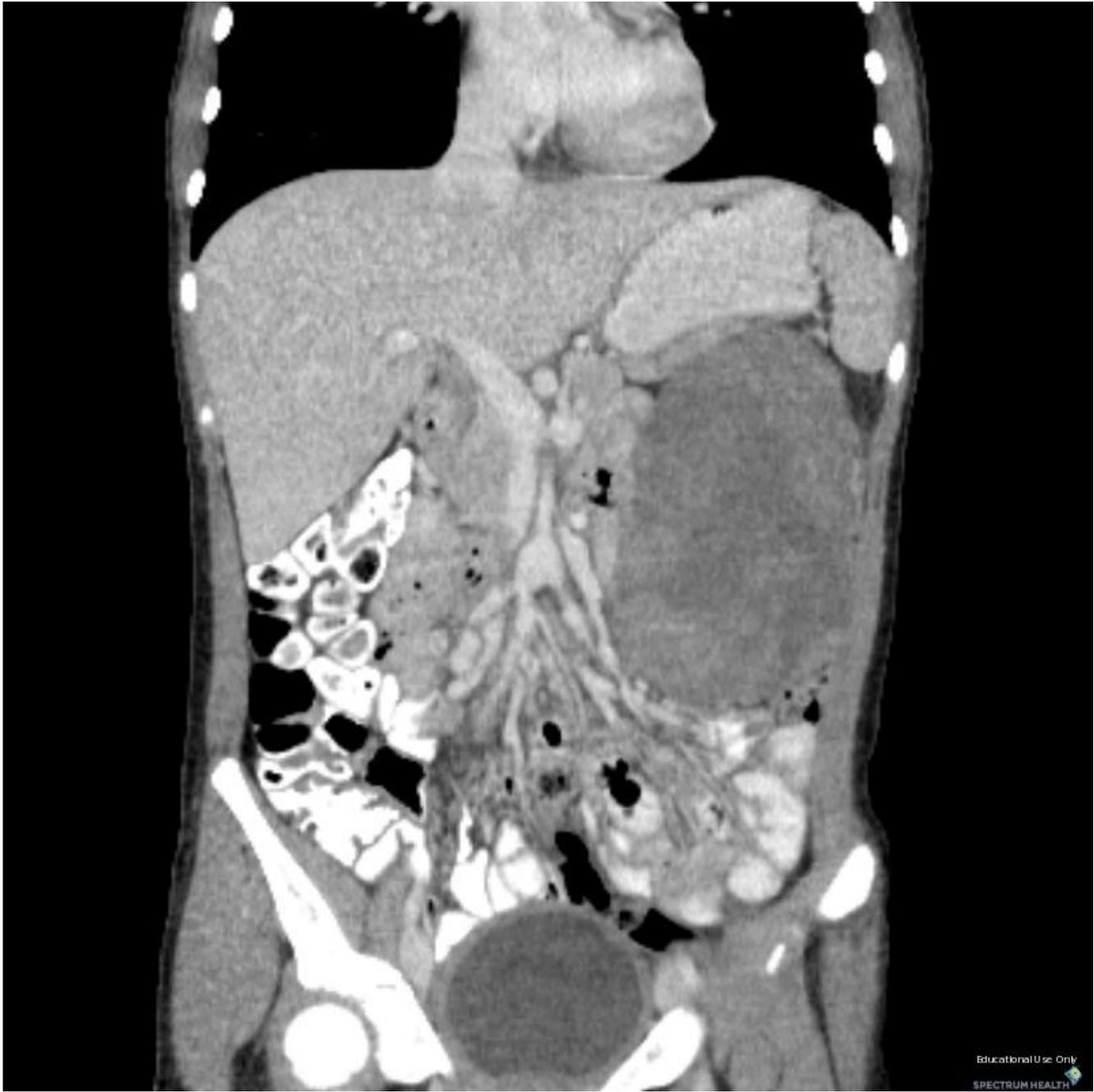
RadioGraphics, 20, 1585-1603.

Renal vasculitis associated with renal cell carcinoma. Mark Lloyd, MRCP. J R Soc Med June 2002 vol. 95 no. 6 305-306

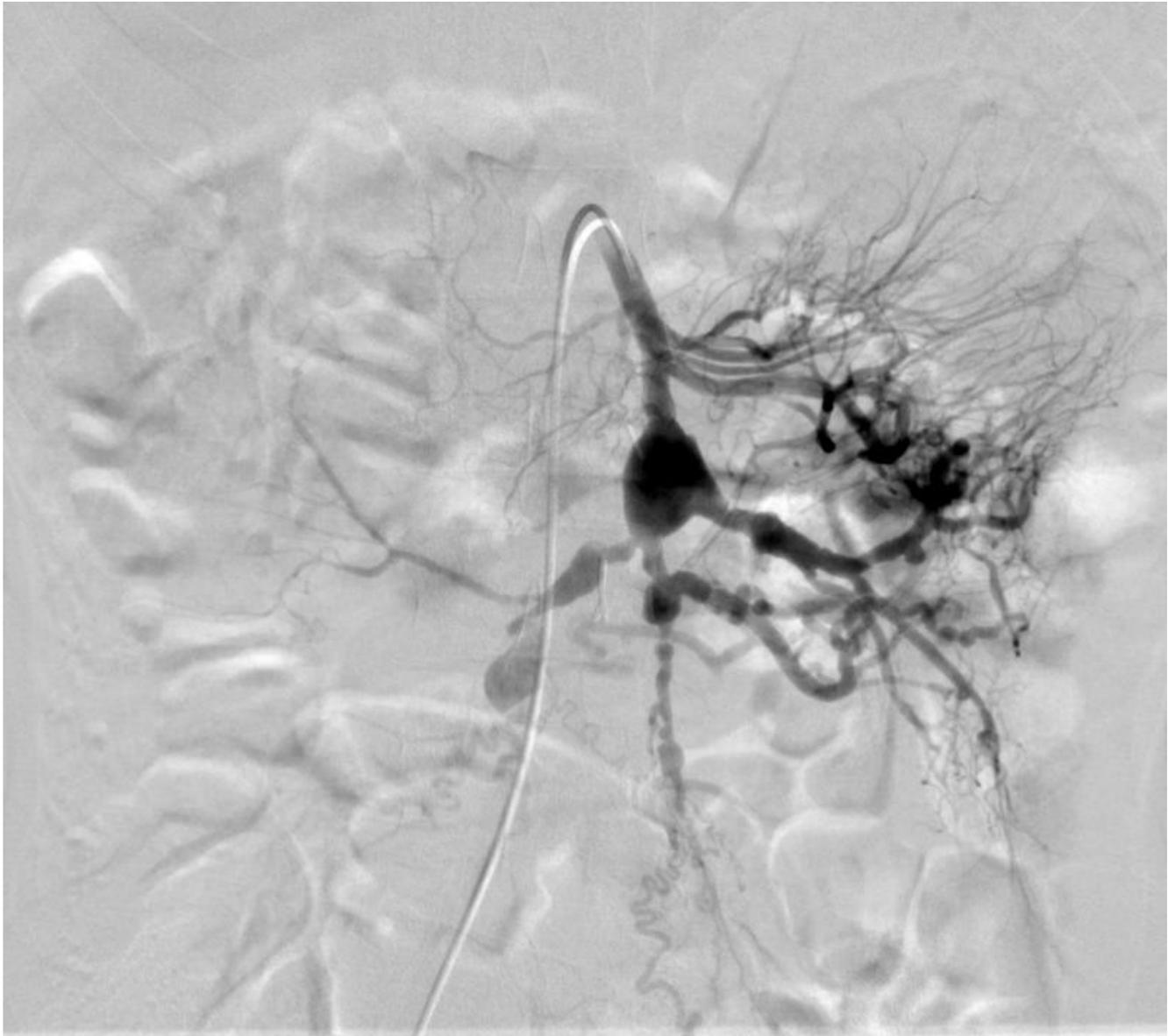
Vasculitides associated with malignancy. Paul R. Fortin.

Pediatr Radiol (2010) 40:766–769. Abdominal manifestations of polyarteritis nodosa demonstrated with CT Ibrahim Adaletli &





Educational Use Only
SPECTRUM HEALTH





Sponsored By



Disclaimer

This teaching site is partially funded by an educational grant from GE Healthcare and Advanced Radiology Services, PC. The material on this site is independently controlled by Advanced Radiology Services, PC, and GE Healthcare and Spectrum Health have no influence over the content of this site
Content Download Agreement

The cases and images on this website are owned by Spectrum Health. Permission is granted (for nonprofit educational purposes) to download and print materials to distribute for the purpose of facilitating the education of health professionals. The authors retain all rights to the material and users are requested to acknowledge the source of the material.

Site Disclaimer

This site is developed to reach healthcare professionals and medical students. Nothing this site should be considered medical advice.

Only your own doctor can help you make decisions about your medical care. If you have a specific medical question or are seeking medical care, please contact your physician.

The information in this website is provided for general medical education purposes only and is not meant to substitute for the independent medical judgment of a physician relative to diagnostic and treatment options of a specific medical condition.

The viewpoints expressed in these cases are those of the authors. They do not represent an endorsement. In no event will Advanced Radiology Associates, PC, Spectrum Health Hospitals (Helen DeVos Children's Hospital) or GE Healthcare be liable for any decision made or action taken in reliance upon the information provided through this website.