CASE REPORT

A case of spontaneous perirenal hemorrhage secondary to polyarteritis nodosa

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A case report of a patient who develops a spontaneous perinephric hemorrhage secondary to polyarteritis nodosa (PAN) is described. The diagnosis of PAN was delayed in this patient somewhat due to a previously unrecognized relation to a recent perihepatic hemorrhage. Unenhanced CT findings in this case were nonspecific, but follow up angiography demonstrated characteristic subsegmental and interlobular renal artery aneurysms. Spontaneous perinephric hemorrhage is an uncommon complication of PAN however an elevated level of suspicion regarding the diagnosis of a systemic vasculitis should be considered in any case of spontaneous renal, hepatic, or gastrointestinal hemorrhage.

Key Words: polyarteritis nodosa, spontaneous perirenal hemorrhage, renal aneurysm

Case report

A 55 year old man with multiple sclerosis and autoimmune hemolytic anemia presented to the emergency room via ambulance with a complaint of right upper quadrant/right flank pain following a minor fall in the shower. The patient was assessed and found to be stable without need for transfusion. Two days following discharge, the patient was readmitted with shock and required urgent transfusion and exploratory laparotomy for 5.5 L of hematoma and blood in the abdomen associated with a linear laceration of the liver.

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One year following this presentation, the patient was again seen in the emergency room with a 12 hour history of malaise, fatigue and diffuse mild abdominal pain noted while lying quietly at home. The patient was hemodynamically stable, however a decreasing trend in serum hemoglobin from 102 to 82 over 2 hours prompted ongoing packed red cell transfusions. Abdominal ultrasound revealed the presence of a large right retroperitoneal hematoma of uncertain origin. With a serum creatinine of 132, this was followed by a noncontrast CT scan of the abdomen confirming a large right retroperitoneal hematoma associated with a right subcapsular renal hemorrhage Figure 1. The patient's hemoglobin stabilized over 6 hours with a total of five units of PRBC and the patient convalesced well over the next 3 days. In light of this patient's history, the patient consented to a selective

arteriogram of the right kidney and hepatic artery Figures 2, 3. This study reported multiple old renal infarcts, and multiple subsegmental renal artery aneurysms. The hepatic artery demonstrated numerous aneurysms the largest of which measured 1.4 cm x 0.9 cm. Further bloodwork demonstrated

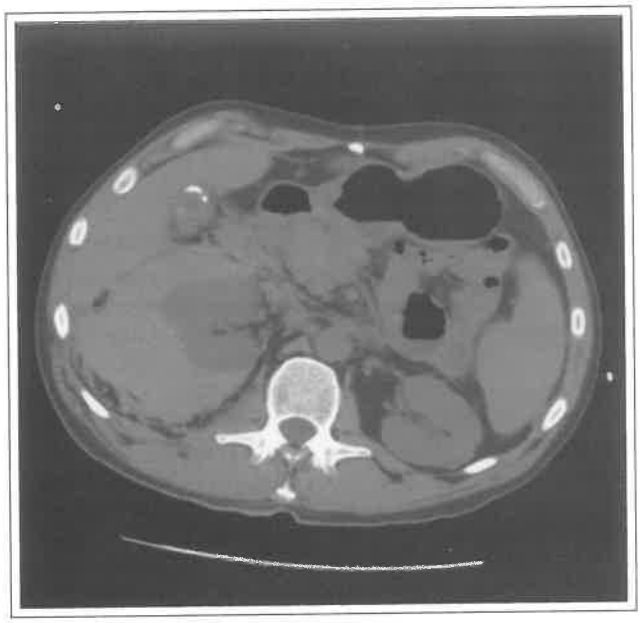

Figure 1. Noncontrast CT scan of the abdomen demonstrating large right retroperitoneal hematoma.

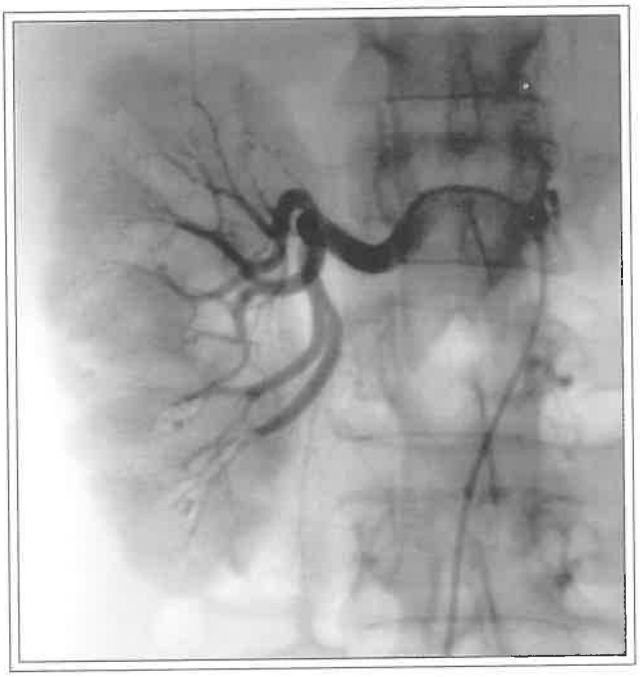

Figure 2. Selective angiogram demonstrates subsegmental and interlobular aneurysms of varying size.

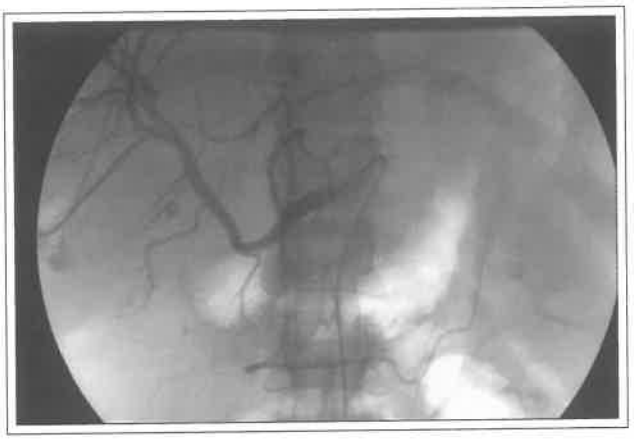

Figure 3. Selective hepatic artery angiogram reveals several aneurysms consistent with the diagnosis of PAN.

ANCA negative, C3 negative, C4 negative and hepatitis screen negative results. The patient was diagnosed with polyarteritis nodosa and started therapy with cyclophosphamide and prednisone.

Discussion

Polyarteritis nodosa (PAN) is one of the earliest described of the systemic vasculitides originally documented by Kussmaul and Maier in 1866. A multisystem disease characterized by a panarteritis of small to medium sized arteries. Histology generally reflects periarterial inflammation and necrosis of the elastic lamina which is the causative pathway to the formation of aneurysms, thrombosis and rupture in this disease.[1,2]

Previous studies have identified the most common sites of PAN affliction as: renal (80%-100%), cardiac (70%), liver (40%-60%), and gastrointestinal (30%-50%). The incidence of PAN in the general population is quoted as two to three cases per million with 30% of patients hepatitis B surface antigen positive. It is a disease most likely affecting patients in the 3rd to 4th decade of life. Arkin[3] in his 1930 study of PAN described a tetrad of frequently presenting symptoms: renal disease, polyneuritis, polymyosistis, and abdominal pain.[4,5] Untreated, the fully differentiated form carries a survival rate of 12% with timely immunosuppressive treatment increasing survival to 75%-80%.[7]

The radiographic findings of PAN on CT include enhancing aneurysms of the interlobular and segmental arteries, hypoperfusion of the parenchyma distal to these lesions, and areas of parenchymal atrophy. Angiography usually demonstrates a

decrease in number of vessels secondary to multiple thrombi, and a patchy nephrogram phase from multiple subsegmental infarcts.[1,6] As common as renal aneurysms are with PAN, other disease processes may present with a similar radiographic picture such as SLE, Wegner's Granulomatosis, rheumatoid arthritis, gram negative sepsis, necrotizing angiitis, and intravenous drug abuse.[7,8]

Wunderlich in 1856 described the first case of spontaneous perirenal hemorrhage and ascribed its etiology to an "apoplexy of the renal capsule".[6] Schmidt in 1908 described the first case of spontaneous perirenal hemorrhage secondary to PAN.[10] Since then, over 250 cases of spontaneous perinephric hemorrhage have been reported in the literature over the past 50 years.[5,11] Several investigators have attempted to determine the risk of perinephric hemorrhage with respect to various renal pathologies.[5,11] Of 194 patients with this diagnosis, Novicki et al found 16% with benign or malignant renal tumors, 15% had nephritis, and 4% were accounted for by inflammatory vascular disease. Brkovic et al demonstrated somewhat dissimilar correlations in their study of 19 patients with spontaneous perinephric hemorrhage with angiomyolipoma, polycystic kidney disease, and autoimmune vasculitides accounting for 22% of the total. Renal cell carcinoma as a cause of hemorrhage made up 11% of the total number in this study. Risk factors that may increase the rate of spontaneous polyarteritis nodosa aneurysm rupture include severe uncontrolled hypertension, thrombocytopenia and pregnancy.

In summary, spontaneous perirenal hemorrhage secondary to polyarteritis nodosa is an uncommon event. Early recognition and treatment of PAN provides significant survival benefit in a disease with untreated mortality approaching 90%. Despite improvements in technology, computed tomography may be best supported with selective angiography and biopsy of affected organs for patients in which there is high index of suspicion of polyarteritis nodosa. □

References

1. Hershman A, Blum R, Lee YC. Angiographic findings in polyarteritis nodosa. *Radiology* 1970;94:147-148.
2. Guillevin L, Lhote, F. Classification and management of necrotizing vasculitides. *Drugs* 1970;53:805-816.
3. Arkin A. A clinical and pathological study of periarteritis nodosa. A report of 5 cases, one histologically headed. *QJM* 1930;6:401-427.
4. Albert DA, Rimon D, Silverstein MC. The diagnosis of polyarteritis nodosa: A literature-based decision analysis approach. *Arthritis Rheum* 1988;31:1117-1119
5. Bonsib SM. Polyarteritis Nodosa. *Sem Diag Path* 2001;18:14-23.
6. Peterson C, Willerson JT, Doppman JL, Decker JL. Polyarteritis nodosa with bilateral renal artery aneurysms and perirenal hematomas: angiographic and nephrotomographic features. *Br J Radiol* 1970;43:62-71.
7. Brkovic D, Moehring K, Doersam J et al. Aetiology, Diagnosis and Management of spontaneous perirenal hematomas. *Eur Urol* 1996;29:302-307.
8. Allen AW, Waybill P, Singh H, Brown DB. Polyarteritis nodosa presenting as spontaneous perirenal hemorrhage: angiographic diagnosis and treatment with microcoil embolization. *JVIR* 1999;10:1361-1363.
9. Wunderlich CRA. Handbuch der Pathologie und Therapie, ed 2. Stuttgart, Ebner und Seubert, 1856.
10. Schmidt JE. Uber Periarteritis nodosa. *Beitr Pathol Anat* 1908;43:455-456.
11. Novicki DE, Turlington JT, Ball TP Jr. The evaluation and management of spontaneous perirenal hemorrhage. *J Urol* 1980;123:764-765.