

Giant Bilateral Angiolipomas Presenting As Syncope: Case Report And Review Of The Literature

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Citation

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Abstract

We present the case of a 41 year old woman found to have giant bilateral angiomyolipomas diagnosed by CT scan during evaluation for syncope. The patient was later found to have decreased mental acuity and evidence of adenoma sebaceum. She did well with transfusion and observation. Our report reviews the literature and common management options for this condition.

CASE REPORT

A 41 year old female presented to the emergency department after a syncopal episode on a shopping mall. Her vitals were stable and physical examination revealed decreased mental acuity, adenoma sebaceum, and a palpable abdominal masses bilaterally in the upper quadrants. Hematocrit decreased to 25% from 40% approximately 6 months prior at a well patient office visit. Her serum creatinine was 1.5 mg/dL (baseline). CT scan of the abdomen and pelvis was obtained (figure 1) revealed no evidence of active retroperitoneal bleed but bilateral massive 30 cm angiomyolipomas which practically replaced the functioning renal parenchyma were noted. She was transfused with 5 units of packed red blood cells and ultimately managed with observation and has done well.

Figure 1

Figure 1: CT scan shows massively enlarged kidneys with bilateral angiomyolipomas replacing nearly all functional renal parenchyma.



DISCUSSION

Tuberous Sclerosis Complex (TSC) affects approximately 1:10,000 births. The classic triad consists of mental retardation, seizures, and adenoma sebaceum is only present in a minority of patients with TSC. Angiomyolipomas (AMLs) are found in 60-80% of patients with TSC, and when present are more often larger, bilateral, and symptomatic [1]. Our patient had evidence of the TSC complex by history and examination. Treatment and

surveillance options should be adjusted from the original recommendations by Oesterling et. al. on symptomatic AMLs that are larger than 4 cm. They stated that tumors 4 cm or greater, if symptomatic, should be studied angiographically and considered candidates for selective arterial embolization or partial nephrectomy. If the lesions are smaller than 4 cm or asymptomatic, they should be followed regularly with CT scan or US [2]. In this report, no separation was made between sporadic cases and those associated with TSC in the treatment guidelines.

Dickinson and colleagues studied optimal treatments based on size and symptoms on AMLs and suggested that medium sized lesions (4-8 cm) had variable presentations and were best managed with serial imaging and intervention should only be undertaken if changes in size or symptoms (i.e. flank pain) occurred. All lesions > 8 cm should require prophylactic treatment prior to the development of symptoms [3]. Hadley and colleagues published on AMLs in patients with TSC and deferred intervention on large asymptomatic lesions (average size being 13.9 cm) until signs (flank pain, need for blood transfusion, concern for malignancy) developed. They cited an earlier report Nelson and Sandra that prophylactic treatment for AMLs in TSC has yet to be defined to deter TSC patients from undergoing multiple treatments based on size alone [5]. This is in contradiction to a paper by Simmons et al. whose guidelines consider prophylactic embolization or renal sparing surgery for asymptomatic lesions greater than 4 cm [6].

There have been recent case reports of conservative treatment in patients with giant symptomatic angiomyolipomas. Danforth et al. reported on two patients who were managed conservatively for greater than 20 years. One patient, a 32 year old male, with lesions 7-7.5 cm in size, was admitted three times with flank pain (as well as fever and hematuria) over 20 years. He required no blood transfusions while maintaining stable renal function during this time and only underwent minor procedures (percutaneous cyst drainage). The other patient, a 23 year old female (diagnosed at age 10 with TSC), presented with flank pain with a palpable mass, nausea, vomiting, and microhematuria. She underwent left nephrectomy after a radionuclide scan showed no flow to this kidney. Her right kidney, with lesions as large as 24 x 14 cm, was managed over 24 years with 44 blood transfusions and 110 days of inpatient care but avoided dialysis. The patient subsequently underwent nephrectomy, dialysis, and placed

on the renal transplant list [7].

Partial nephrectomy for TSC patients with AMLs may not be feasible, as stated by Harabayashi and colleagues [8]. However, there have been instances where nephron sparing surgery preserved renal function in patients with giant bilateral AMLs associated with TSC. Ciancio et al. reported a 39 year old female that underwent simultaneous right nephrectomy (35 x 14 x 10 cm) and left partial nephrectomy (4350 grams for the right kidney and 1150 grams for the left specimen) when she presented with a 4-day history of diffuse abdominal pain, nausea, and vomiting. The patient had a nadir of her creatinine at 1.3 mg/dL at 2 weeks post-operatively after peaking at 6.3 mg/dL [9].

CONCLUSIONS

Diagnosis of the TSC requires careful physical examination to determine the presence of adenoma sebaceum and the degree of decreased mental acuity. Imaging studies of the abdomen and pelvis can then determine the presence of angiomyolipomas. Treatment and surveillance options are based on size of the lesions and the presence or absence of symptoms. Treatment options can include active surveillance, selective embolization, partial and radical nephrectomy.

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