

Isolated polycystic liver disease and aneurism: a case report

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Abstract: Isolated polycystic liver disease (PCLD) has not been associated with aneurysms and concomitant PLD has not been reported previously in association with bilateral popliteal aneurysms. A case of a middle-aged man with PLD, marfanoid habitus and bilateral popliteal aneurysms is presented.

Keywords: Isolated polycystic liver disease; aneurism; femoro-popliteal bypass

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Introduction

It is known that autosomal dominant polycystic kidney disease (ADPKD) and polycystic liver disease (PCLD) follow an autosomal dominant pattern of inheritance, and cerebral vascular manifestations have been described as occasional part of the disease spectrum (1). Furthermore, cerebral aneurysms are suggested to be part of the spectrum of PCLD (2). Based on the mutation load there are different appearances of the disease in different vessels. In patients with multiple liver and/or kidney cysts and a history of headache, subarachnoidal hemorrhage as well as carotid dissection must be considered. We present a case with aneurism due to PCLD. Previous publications support the screening of these patients for cerebral and intracranial aneurysms (2,3).

Case presentation

In January 2011 a 51-year-old man presented with a painful, pulsating right popliteal mass with no history of trauma. There was nothing else of significance in his

family or past medical history apart from pharmaceutically controlled hypertension. The patient was unusually tall for Greek circumstances but no high arched palate, no pectus excavatum and no joint laxity. Clinical examination showed bilateral pulsating popliteal swellings and a left sided inguinal hernia. A computed tomogram with contrast confirmed bilateral partially thrombosed popliteal aneurysms (right 5.2×6 cm² and left 2.6×2.5 cm²) (Figures 1,2) with PCLD, right common iliac and common femoral artery dilatation but no atherosclerotic disease or aortic aneurysmal changes. The patient underwent successful urgent femoro-popliteal bypass with vein graft and ligation of the aneurysm initially on the right followed by the same procedure on the left two months later. Histological evaluation of arterial sample did not reveal any signs of vasculitis or possible connective tissue disorder.

Discussion

PCLD is an assortment of rare human disorders and more particularly is an inherited Mendelian disorder (4,5). These

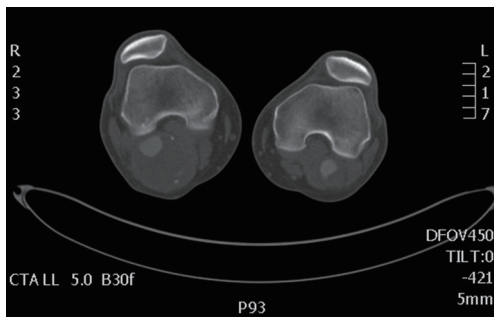


Figure 1 Initial findings with CT of the legs.

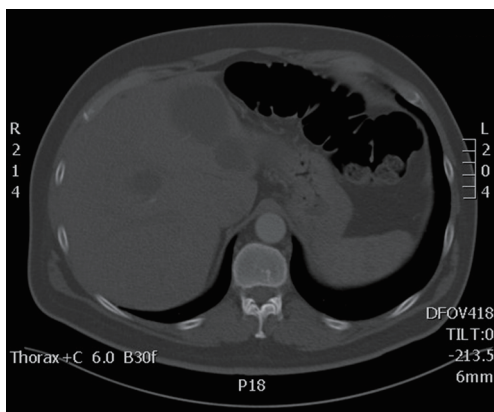


Figure 2 CT of the abdomen findings.

disorders are results from structural changes of the biliary tree development and (6) also consist a group of genetic disorders that initiated by mutations in many related genes of PLD and characterized by the development of multiple cholangiocyte derived hepatic cysts that with time replace liver tissue (7,8). Current medical therapies for PLD include symptomatic management (9) and surgical interventions (10).

In our case report the patient unusually diagnosed also with bilateral popliteal artery aneurysm (PAA). The PAA is the commonest peripheral aneurysm which presenting bilaterally in 50% of cases. It is noticeable that 80% of the patients are asymptomatic when diagnosed with PAP, and as follows from the literature, at a rate of 14% per year, these patients become symptomatic (11,12). Best results demonstrated and the complications minimized when the treatment is given before symptoms onset (13). The treatment of choice includes surgical techniques and endovascular interventions, as in our case, where the patient submitted successfully in femoro-popliteal bypass with vein graft and ligation of the aneurysm (14,15).

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None.

Footnote

Conflicts of Interest: The authors have no conflicts of interest to declare.

Informed Consent: Written informed consent was obtained from the patient for publication of this Case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

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