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Massive renal and adrenal calcifications in a young dialysis patient with familial Mediterranean fever

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A 34-year-old patient was hospitalized for progressive development of asthenia, muscle weakness and weight loss. This Greek man had familial Mediterranean fever (FMF) which led to end-stage renal disease. After 4 years of haemodialysis, a thoracic computed tomography scan revealed massive homogeneous calcification of both kidneys and adrenal glands (Figure 1) confirmed by abdominal scan (Figure 2). Endocrine tests showed adrenal insufficiency.

FMF is an autosomal recessive disease, characterized by recurrent attacks of fever with serositis, which affects populations around the Mediterranean Sea. The mutated gene is localized on the short arm of chromosome 16 and codes for a protein of 781 amino acids called pyrin or marenostrin. In FMF, inflammation attacks are accompanied by extremely high concentration of serum amyloid protein A (SAA) which causes typical secondary amyloidosis [1]. Soft tissue and vascular calcifications are classical complications in the dialysis population. The pathogenesis of such calcifications is complex and multifactorial. In this case, it is related to the fact that amyloid fibrils (SAA, AL, β_2-microglobulin and amyloid P component) have a high affinity for calcium, as suggested by other authors [2].

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Conflict of interest statement. None declared.

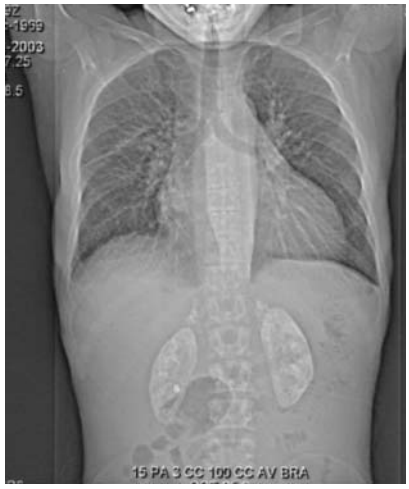

Fig. 1. This abdominal scout view shows massive, homogeneous calcification of both kidneys and adrenal glands.

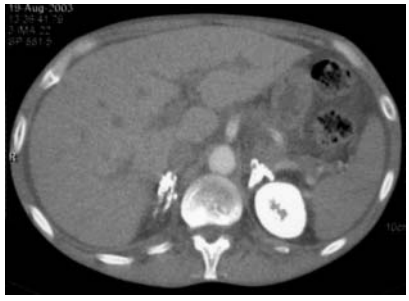

Fig. 2. An abdominal CT scan showing calcification of both adrenal glands and left kidney.

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