

CALCIPHYLAXIS
CASE REPORT

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ABSTRACT

Calciphylaxis is a rare, often fatal, systemic disorder characterized by deposition (precipitation) of calcium phosphate salts (calcification) in the medial layer of the arteries and soft tissues. Calcification of the media is followed by fibrous hyperplasia of the intima with obliteration of the lumen and tissue ischemia, necrosis, and gangrene. The first description of calciphylaxis was published in 1962. Calciphylaxis occurs in patients with end stage renal disease. Almost all reported patients were on dialysis.

Keywords: Calciphylaxis; dialysis; hypercalciemia

CASE

A 36 year-old male presented with a chief complaint of bone pain and fever during his hemodialysis sessions. The patient is on renal replacement therapy since 29.12.1996 when he was urgently dialyzed due to high plasma creatinine and BUN. In year 2000 he suffered cerebral infarction. The patient has been complaining of severe bone pain for the past 12 months. On radiological examination we observed blood vessel calcinosis. Due to acral necrosis on 15.12.2004 the distal phalanga of fifth finger of left arm was amputated. On 14.01.2005 subtotal parathyroidectomy was performed by a thoracic surgeon.

Examination showed male looking significantly older than his actual age, in poor general condition. Head and neck examination revealed venous stagnation, pale skin and conjunctiva. CVS: heart proection dilated to the left on percussion; on auscultation - rhythmical with normal heart rate 85/min; RR140/80mmHg. Upper limbs: palmar psoriatic lesions, acronecroses on 3rd finger of right hand and 2nd, 3rd finger of left hand (Figure 1).

Lower limbs: It is well seen in Figure 2 the deformed left gluteal region due to a huge soft tissue calcification. Psoriatic lesions in the lumbar area are also evident.

Figure 1

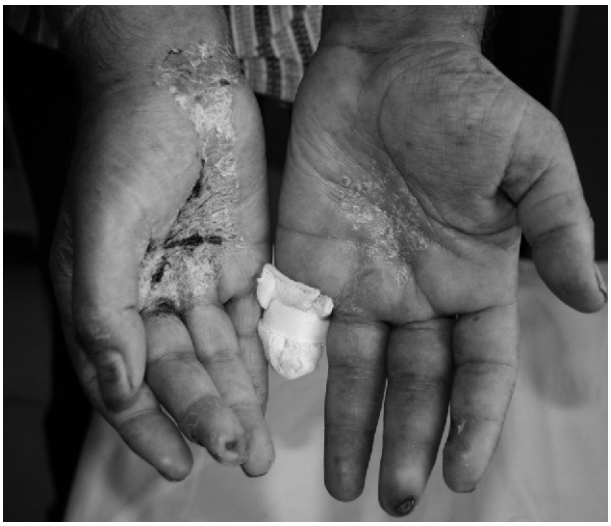
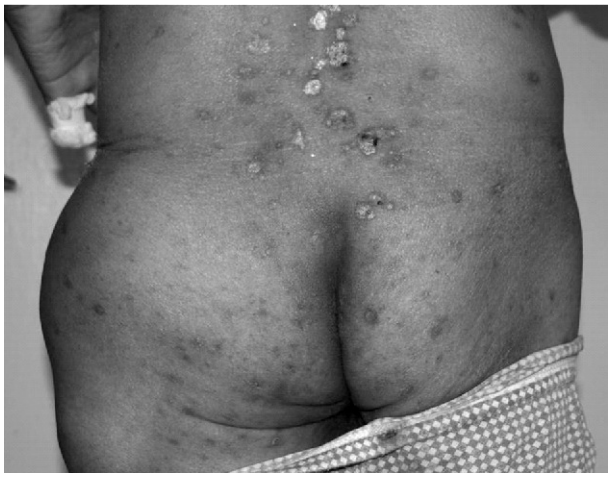


Figure 2



LABORATORY FINDINGS

Laboratory results show extremely high levels of parathormone (PTH) and elevated plasma phosphorus prior to parathyroidectomy. After the surgical intervention calcium and phosphorus levels went low and calcium supplementation and Rocaltrol were administered. It is seen from the table that a year later in the spring of 2006, the PTH is high again which is probably due to hyperplasia of the remaining parathyroid tissue.

	12.2004	02.2005	03.2005	04.2006
HGB g/l	77	71	91	115
Creatinine mol/l	636	505	645	
Calcium mmol/l	2,12	1,6	1,63	1,98
Phosphorus mmol/l	2,18	1,27	2,04	2,26
Parathormone pg/ml	2218			980
Ca x P	4,6	2,03	3,33	4,47
Plasma albumine g/l	32	30,2	33	
AP UI	289	356	145	

IMAGING

Ro graphia of chest: the abnormal findings were cranial pulmonary venous stagnation; calcinosis of the arcus of the aorta; high cardiothoracic index. More interesting was the Ro graphia of the hands and calves: shows calcinosis of the blood vessels - digital arteries, anterior and posterior tibial artery and the popliteal artery (Figure 3; Figure 4).

Figure 3

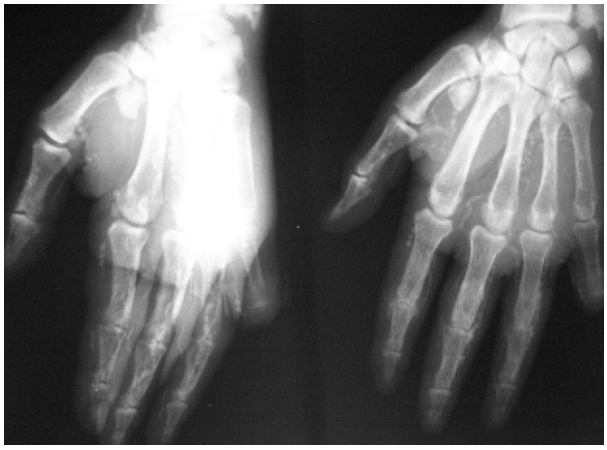
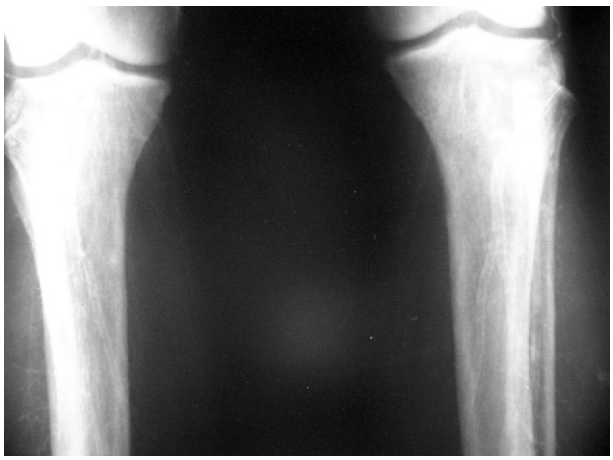


Figure 4



HISTOLOGY FINDINGS

Figure 5 shows a massive soft tissue calcificate in a dermal specimen from one of the amputated fingers. Punch biopsy was not performed because of high risk of gangrene following the manipulation. On Figure 6 we present a calcificate in the parathyroid gland.

Figure 5

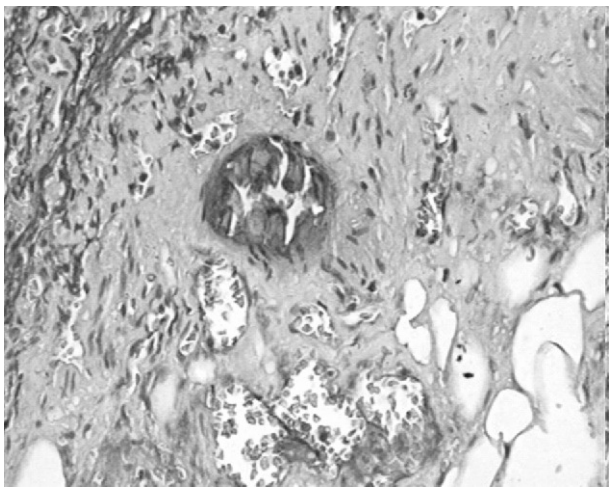
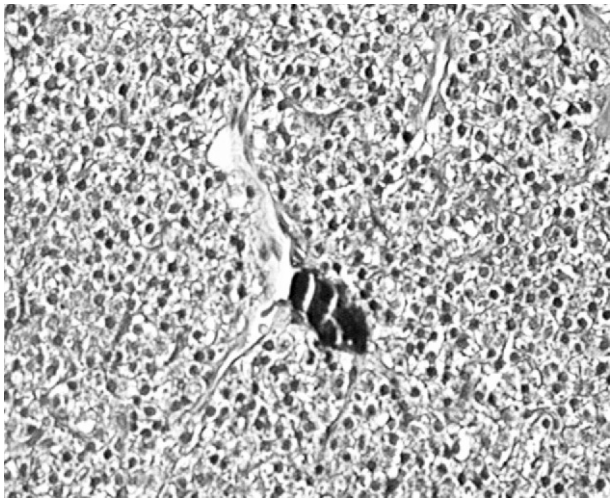


Figure 6



DIAGNOSIS

Our diagnosis of systemic calciphylaxis was based on the above described clinical features, laboratory findings (extremely high level of PTH, although the PxCa was not above the normal 5,5), radiological findings and histology. The conservative treatment we administered was aimed to control the plasma levels of calcium and phosphorus, relieve the severe pains, prevent septic conditions due to superinfections and control the arterial pressure.

DISCUSSION

Calciphylaxis is a rare, often fatal, systemic disorder characterized by deposition (precipitation) of calcium phosphate salts (calcification) in the medial layer of the arteries and soft tissues. Calcification of the media is followed by fibrous hyperplasia of the intima with obliteration of the lumen and tissue ischemia, necrosis, and gangrene (3,4). The first description of calciphylaxis was published in 1962 by Selye (1). Calciphylaxis occurs in patients with end stage renal disease. Almost all reported patients were on dialysis, or had recently received a renal transplant. The pathogenesis of calciphylaxis is poorly understood - several risk factors have been reported as follows: end-stage renal disease, hypercalcemic states, abnormalities in calcium-phosphorus concentration, hypercoagulable states, morbid obesity and recent weight loss, impaired protein-C activity, use of calcium carbonate, recent use of prednisone and hyperparathyroidism. All of the mentioned risk factors are frequently present in end stage renal disease patients and it is not clear why calciphylaxis is so rare (5,6). Although a conclusive effective therapy does not exist, parathyroidectomy can be safely performed and may benefit some patients with what is often an otherwise fatal disease (7). Unfortunately subtotal parathyroidectomy may be followed by hyperplasia of the remnants of parathyroid tissue as it was in our case.

REFERENCE

1. Selye, H., G. Gabbiani, R. Strebel. Sensitization to calciphylaxis by endogenous parathyroid hormone. - Endocrinology, 71, 1962, 554-8.
2. Jelic, T. M., A. M. Malas, S. S. Groves, et al. Nanobacteria-caused mitral valve calciphylaxis in a man with diabetic renal failure. - South Med. J., 97, 2004, 2, 194-8.
3. Budisavljevic, M. N., D. Cheek, D. W. Ploth. Calciphylaxis in chronic renal failure. - J Am Soc Nephrol., 7, 1996, 7, 978-982.
4. Adrogue, H. J., M. R. Frazier, B. Zeluff, et al. Systemic calciphylaxis revisited. - Am J Nephrol., 1, 1981, 3-4, 177-183.
5. Goodman, W. G. Vascular calcification in chronic renal failure. - The Lancet, 358, 2001, 1115-6.
6. Mehta, R. L., G. Scott, J.A. Sloand, et al. Skin necrosis associated with acquired protein C deficiency in patients with renal failure and calciphylaxis.- Am J Med.,88, 1990, 252-7.
7. Kriskovich, M. D., J. M. Holman, J. R. Haller. Calciphylaxis: Is there a role for parathyroidectomy? - Laryngoscope, 110, 2000, 4, 603-7.