Xanthogranulomatous pyelonephritis presenting as palmoplantar keratoderma

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DESCRIPTION

A 60-year-old woman with type 1 diabetes mellitus and a history of repeated urinary tract infections due to *Proteus mirabilis* presented with diffuse palmoplantar hyperkeratosis of 4 months duration (figure 1) and acute retrosternal pain. An ECG showed a diffuse concave ST segmental elevation and the echocardiogram showed a mild pericardial effusion, suggestive of pericarditis. Laboratory analysis showed negative troponin, leucocytosis, elevated erythrocyte sedimentation rate (110 mm/h) and C reactive protein (10 mg/dL). A CT scan

revealed a hydronephrotic, non-functioning left kidney with an inflammatory infiltrate in the perinephric fat and a bilateral coraliform calculus. A left radical nephrectomy was performed and the histological diagnosis of xanthogranulomatous pyelonephritis was confirmed (figure 2A, B). Complete clinical remission of the cutaneous lesions and pericarditis was observed 1 month after surgery.

Palmoplantar keratoderma is characterised by abnormal thickening of the skin on the palms and soles. It can be hereditary or acquired; when acquired it is associated with malignancy, drugs, infections and chronic systemic diseases, but it can also be idiopathic.¹

Xanthogranulomatous pyelonephritis is an uncommon chronic destructive inflammatory disease of the renal parenchyma. Most cases occur in the setting of obstruction due to infected renal stones, mostly by *Escherichia coli* and *P. mirabilis*.²

To the best of our knowledge, this is the first case reported of acquired palmoplantar keratoderma associated with xanthogranulomatous pyelonephritis.

Figure 1 Symmetric, diffuse and yellowish hyperkeratosis of the plantar surfaces, compatible with diffuse plantar keratoderma.

Learning points

- ▶ Acquired palmoplantar keratoderma is a multiaetiological disorder and may be the first manifestation of a systemic disease.
- ▶ Recognition of the underlying cause is essential for successful treatment.
- ▶ Although very rare, xanthogranulomatous pyelonephritis should be considered in the differential diagnosis of palmoplantar keratoderma.

Figure 2 (A) The renal cortex shows some globally sclerotic glomeruli interspaced with others with periglomerular fibrosis, but otherwise relatively preserved. The interstitium is severely compromised by an intense inflammatory infiltrate, with abscess foci and areas of necrosis. There is severe tubular atrophy with peritubular fibrosis (H&E, ×100); (B) Renal medulla with a diffuse inflammatory infiltrate, which includes large numbers of foamy histiocytes (lipid-laden macrophages). Some areas of necrosis can also be seen (H&E, ×100).

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