Periorbital purpura (raccoon's eyes)

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DESCRIPTION

We report a case of a 62-year-old female patient with a 3-year history of non-traumatic palpebral and periorbital purpura (figure 1) and episodes of syncope. She had no other alteration on clinical examination.

The laboratory study showed a complete blood count, serum electrolytes, renal function, coagulation and bilirubin within normal limits, alkaline phosphatase 171 IU/L (normal range 45–116), γ-glutamyltransferase 192 IU/L (12–43), alanine aminotransferase 50 IU/L (9–52) and aspartate aminotransferase 41 IU/L (14–36).

Antinuclear antibody, antimitochondrial antibodies, antismooth muscle antibody, antiliverkidney microsome antibodies and infectious screening were all negative. It was identified as λ light-chain monoclonal IgG gammopathy by immunofixation, confirmed in a free light-chain assay: free λ light chains 130 mg/dL (5.71–26.30) and κ/λ ratio 0.10.

Serum β-2-microglobulin was 3.55 mg/L (1.09–2.53) and bone marrow plasma cell percentage was 5.2%, most of them with an aberrant immunophenotype on flow cytometry.

A CT showed hepatomegaly and bilateral pleural effusion.

Echocardiographic findings were consistent with restrictive cardiomyopathy.

The cardiac electrophysiological study demonstrated an atrioventricular nodal re-entrant tachycardia and the event recording monitoring system showed periods of asystole greater than 3 s. A pacemaker was implanted. Cardiovascular MR revealed global and subendocardial late gadolinium enhancement of the myocardium.

The liver biopsy showed κ light-chain deposition, these deposits were granular and do not stained with Congo Red (figure 2).

These biochemical characteristics make differential diagnose between light-chain deposition disease (LCDD) and immunoglobulin light-chain (AL) amyloidosis.[1] The patient was diagnosed with LCDD with liver and cardiac involvement and started treatment with dexamethasone and bortezomib. The disease progressed and the patient died of liver and cardiac failure within 8 months of diagnosis.

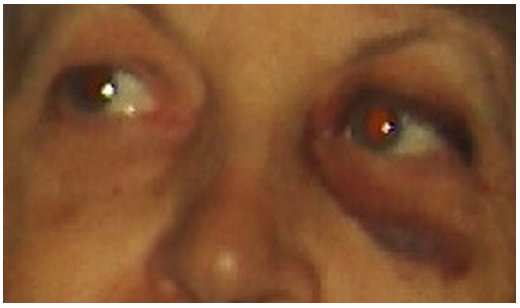

Figure 1 Palpebral and periorbital purpura.

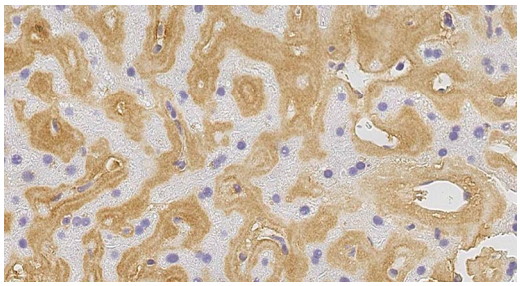

Figure 2 An immunoperoxidase stain of a liver biopsy (×40 magnification) showing κ light-chain deposition. A stain for λ light chains was negative.

Learning points

- Some of the possible causes of raccoon's eyes are traumatic (basal skull fracture and rhinoplasty); neuroblastoma; lymphoma; kaposi's sarcoma and clonal plasma cell proliferative disorders (AL amyloidosis, light-chain deposition disease (LCDD) and multiple myeloma).[2]
- In LCDD, raccoon's eyes is a consequence of vascular fragility induced by the deposition of light-chain fragments.[3]
- LCDD is a non-amyloid monoclonal immunoglobulin, in comparison to AL amyloidosis, the tissue deposits are almost always composed by a constant region of the κ light chains, they are granular not fibrillar and do not bind Congo Red or thioflavin-T.[1]

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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