ANSWER TO PHOTO QUIZ (PAGE 90)

RACCOON SIGN

DIAGNOSIS

The patient was diagnosed with amyloid light-chain (AL) amyloidosis.[1,2] Originally, he presented with a smouldering multiple myeloma. During follow up care, he developed purpura on his hands. A rectal biopsy was performed suspecting amyloidosis, but this was negative for amyloid depositions. The purpura where only present when the patient used gardening tools and resolved spontaneously.

Over the following years, the purpura worsened. A lip biopsy was again negative for amyloid. After this, a skin biopsy of the affected area on his hand was performed. This was positive for Congo red staining and demonstrated a typical apple-green birefringence in polarised light. Furthermore, it was positive for amyloid P and lambda staining and negative for amyloid A and kappa staining. The diagnosis AL amyloidosis was made based on these biopsy results in combination with his known monoclonal plasma cell disorder in the bone marrow.[3] The bone marrow biopsy was not revised for amyloid deposition. The serum level of lambda measured was 722 mg/l, with a kappa/lambda ratio of 0.02.

Our patient was treated with various lines of therapy. During treatments, the purpura seemed to remain steady, but worsened as the disease became refractory. Eventually our patient also developed periorbital distribution of purpura, the highly characteristic raccoon sign.

AL amyloidosis is caused by deposition of protein derived from immunoglobulin light chain fragments associated with monoclonal plasma cell disorders. The rare systemic disease can present with various symptoms including proteinuria, hepatosplenomegaly, and cardiomyopathy. The diagnosis can be made based on serum and urine protein electrophoresis with immunofixation, serum free light chain ratio analysis, in combination with an abdominal fat pad biopsy and bone marrow biopsy.[4] In our case, a rectal biopsy was initially performed, which has a slightly lower sensitivity (50-70%) compared to an abdominal fat pad biopsy (60-80%), but a higher sensitivity than a bone marrow biopsy (50-55%) and skin biopsy (50%). Skin involvement of amyloidosis is seen in approximately 40% of patients with AL amyloidosis. It is caused by amyloid depositions and clinical presentation depends on the site of deposition. Presentations include waxy thickening, easy bruising, and subcutaneous nodules or plaques. The purpura in our patient are probably caused by small vessel amyloid deposition in combination with minor trauma. Amyloidosis is also associated with bleeding diathesis such as factor X deficiency.[5] Factor X and other coagulation tests were normal in our patient. Our patient never showed signs of other organ involvement.

With this case, we hope to establish more knowledge about the skin manifestations of amyloidosis, its manifestation in the course of an apparently normal multiple myeloma disease, and to demonstrate the difficulty in diagnosing this rare disease.

REFERENCES

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