Acquired cutis laxa is an uncommon disorder sometimes associated with monoclonal gammopathy and multiple myeloma, although the mechanism of this link is unclear. We report here a case of a 34-year-old man with generalized acquired cutis laxa and monoclonal light chain disease with renal and neurological involvement. Electron microscopy examination of a skin sample revealed shortened and fragmented elastic fibres in the reticular dermis and normal collagen bundles. Immunogold labelling revealed anti-lambda antibodies closely bound to the microfibrillar component of elastic fibres, thus supporting a causal relationship between monoclonal gammopathy and the changes in skin elasticity. 

**Key words:** monoclonal gammopathy; cutis laxa; myeloma; elastolysis; electron microscopy.

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Acquired cutis laxa and monoclonal gammopathy

Histopathology of a skin biopsy showed no abnormalities on haematoxylin-and-eosin-safran staining. However, there were marked abnormalities of the elastic network, including reduction in elastic fibres, which were fragmented, shortened and clumped on orcein staining (Fig. 2). Electron microscopy of a skin sample revealed that the elastic fibres were shortened and fragmented, with an abundant microfibrillar component, contrasting with normal collagen bundles in the reticular dermis. Immunogold labelling revealed that anti-lambda antibodies were bound to the microfibrillar component of elastic fibres (Fig. 3). The same technique was applied to a skin biopsy from a control patient with no cutis laxa but with monoclonal gammopathy.

DISCUSSION

This is the first report demonstrating binding of monoclonal gammopathy to elastic fibres using electron microscopy, thus providing further evidence of a causal relationship between monoclonal paraprotein and the development of acquired cutis laxa.

A few cases of acquired cutis laxa have been reported to be associated with monoclonal gammopathy and multiple myeloma (4–8). In these cases, as in our own, no correlation was observed between the level of monoclonal paraprotein in the serum and the severity of the acquired cutis laxa. In cases of myeloma associated with amyloidosis, elastolysis was thought to be due to
direct destruction of elastic fibres by amyloid deposits (9). Other mechanisms have also been discussed, including allergic hypersensitivity reaction and inflammatory cell effects (macrophages and neutrophils) (2). When performed, electron microscopy showed degenerative changes in elastic fibres and electron-dense amorphous or granular aggregates in the vicinity of the elastic fibres (10). A causal link between monoclonal paraprotein and acquired cutis laxa has also been suggested, as direct immunofluorescence revealed slight deposition of IgG around elastic fibres of the dermis in 4 cases (6–8). However, no antibodies were highlighted by immunofluorescence skin studies in most cases of cutis laxa with myeloma (3, 11). The demonstration of monoclonal antibodies bound to elastic fibres in the present case does not definitely prove causality in the occurrence of elastolysis. Monoclonal gammopathy is frequent in the general population, and acquired cutis laxa is very rare. The association may therefore occur by chance and the passive fixation of the monoclonal antibody would thus not be responsible for the development of elastolysis, which could alternatively be explained by a genetic polymorphism in one of the component of elastic fibres (fibulin-5 or elastin) (12). However, the fixation seemed to be very specific to elastic fibres in our case. We believe that the monoclonal paraprotein binding to elastic fibres was the primary event and that this may have triggered complement activation, leading to elastolysis. An alternative hypothesis is impairment of the maturation and assembly of elastic fibres consecutively to the binding of monoclonal immunoglobulin to fibulin-5 (12).

The authors declare no conflict of interest.

REFERENCES