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TEACHING CASE

A case report of dystrophic localized amyloidosis that developed in the left thigh

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Abstract

We report on a 50-year-old man with dystrophic localized amyloidosis who noticed a soft tumor in his left thigh about 20 years ago, after which the tumor has gradually enlarged. The multicystic tumor showed hemorrhage, hematoma, necrosis, fibrosis, and tiny nodules and various polymorphous granulomas were observed. One was rich in eosinophilic amorphous materials and cholesterol crystals, and was poor in cell reaction. Another was formed by granuloma consisting of multinucleated giant cells, foamy cells, and macrophages. Transitional granulomas between the two were also observed. The materials showed eosinophilia and red staining and apple-green birefringence in polarized light by alkaline Congo-red stain, and they were also resistant to potassium permanganate pretreatment. They were also positive for amyloid P component and consistently negative for amyloid A, κ- and λ-light chains, β_2-microglobulin, and transthyretin. Therefore, it was suggested that this might be an amyloid derived from the hematoma, which has not been reported to date.

Keywords: Soft tissue cystic tumor; Dystrophic localized amyloidosis; Alkaline Congo-red stain; Giant cell reaction; Thigh

Introduction

Amyloidosis is not a single disease, but is rather composed of a group of diseases. Amyloid fibril proteins and their precursors in humans are divided into 24 types by the International Society of Amyloidosis with respect to their chemical nature: (1) amyloid light (AL) chain, which is derived from the immunoglobulin light chain (Ig LC) as primary amyloidosis, (2) amyloid-A (AA), which is derived from serum AA by the liver as secondary amyloidosis, (3) β-amyloid protein as Alzheimer disease and cerebral amyloid angiopathy, (4) transthyretin as senile systemic amyloidosis and familial amyloid polyneuropathy, (5) β_2-microglobulin with hemodialysis, and others [13,18].

Localized amyloidosis deposits limited to a single organ and formed by amyloid proteins usually produced in the local organ, as well as soft tissue amyloidosis are rare [13]. In localized amyloidosis, tumor-forming

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deposits of amyloid are most often encountered in the lung, larynx, skin, urinary bladder, tongue, and eye. In most cases, their amyloid consists of Ig LC protein [15], and they represent a localized form of AL amyloid.

We report on a 50-year-old man with dystrophic localized amyloidosis presenting as a multicystic tumor with hematoma in the left thigh.

Clinical summary

A 50-year-old man was referred to our clinic with a huge painless tumor in his left lateral thigh measuring 16×14 cm in size, which appeared over 20 years ago and has gradually enlarged (Fig. 1). It was a firm and non-tender mass; systemic symptoms were absent, and his general condition was good. He had no history of trauma or hemodialysis. Hip and knee functions were normal, and no regional lymphadenopathy was found. Plain radiographs showed a soft tissue swelling with no involvement of skeletal structures. Total protein was 7.5 g/dl, and his albumin globulin ratio was 1.9. Monoclonal gammopathy in serum and Bence–Jones protein in his urine were not detected. Magnetic resonance imaging (MRI) showed a multicystic mass measuring $16 \times 14 \times 9$ cm with well-circumscribed irregular margins. The tumor was located between the left tensor fasciae latae muscle and the sartorius muscle, and the lesion covered the rectus femoris muscle. The irregular wall was well-enhanced with Gadrinium, but the cyst was not. His clinical signs and MRI suggested a benign soft tissue tumor. Aspiration cytology was performed 1 month before surgery and showed blood coagula material and no malignancy. The mass was subsequently resected. The patient's postoperative course was uneventful, and he was discharged without undergoing additional therapy. The patient is alive and well 10 months after surgery.

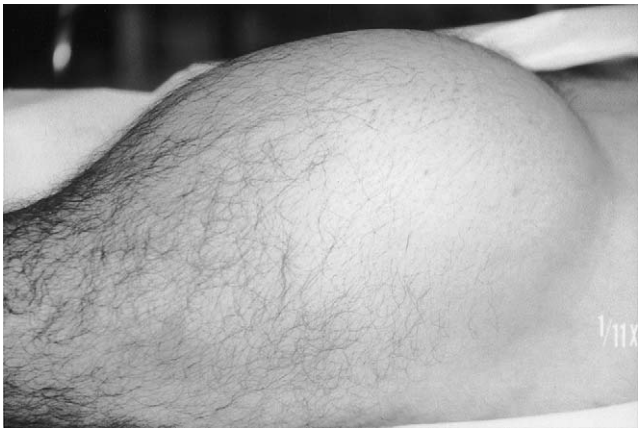

Fig. 1. (External finding) The large tumor was located in the swollen left thigh.

Pathological results

The tumor was located in muscles and adipose tissue, weighted 780 g after resection, was $19 \times 14.5 \times 7$ cm in size, and had multiple cysts that contained blood. The cyst wall was 1.7 cm in depth and showed fibrous thickening. The tumor showed hemorrhaging, necrosis, fibrosis, and multiple nodules ranging from 1 cm in diameter to $5 \times 3.5 \times 3$ cm (Fig. 2). The large nodules were formed by multiple tiny nodules ranging from 0.5 to 1.5 cm, in which various polymorphous granulomas were observed. One was rich in eosinophilic amorphous material and cholesterol crystals and was poor for cell reaction (Fig. 3a). Another was made up of small pieces of amorphous material and cholesterol clusters, followed by a granuloma consisting of foreign body-type multinucleated giant cells, foamy cells, and macrophages (Fig. 3b). Transitional granulomas between the two were also observed. The giant cells had one or two asteroid bodies. The amorphous material measured up to 2 mm in diameter, showed eosinophilia on Hematoxylin–eosin staining, and orange-red staining and green birefringence in polarized light by alkaline Congo-red staining (Figs. 4a and b). These materials were resistant to potassium permanganate ($KMnO_4$) pretreatment. They were surrounded by macrophages, foamy cells, and multinucleated giant cells, which appeared as if they were engulfing and adhering to the amyloid granules (Figs. 4c and d). Foamy cells and macrophages phagocytized Berlin blue-positive hemosiderin. Also, the amyloid was positive for amyloid P (Calbiochem, La Jolla, CA, USA), showing a diffuse pattern (Fig. 5), and this was consistently negative for AA (prepared by Imada et al. [8]), Ig κ(116-133)-, and λ (118-134)-LC constant regions (prepared by Hoshii et al. [7]). Only a few amyloid depositions were faintly or ambiguously positive for β_2-microglobulin (Nordic Immunological

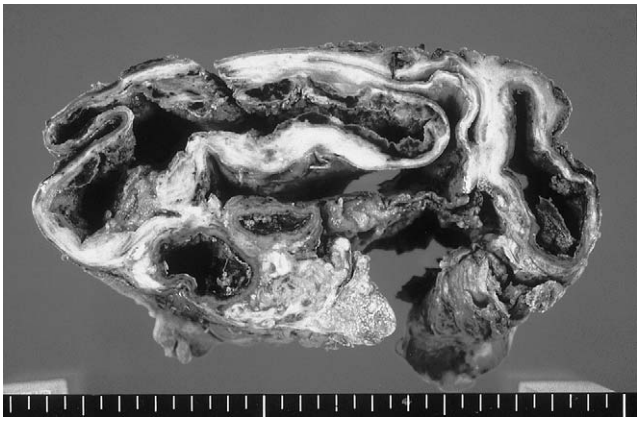

Fig. 2. (Macroscopical finding) The large cystic tumor had thickened fibrous cyst wall, hematoma, hemorrhage, necrosis, and multiple nodules in the cut surface.