Nodular Cutaneous Amyloidosis Involving the Glan Penis: Report of a Case

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Abstract

Nodular cutaneous amyloidosis is a rare disease that predominantly affects women in their sixth and seventh decades. The genital organ is the rarest cutaneous location with only four reported cases of vulvar involvement. We report the first known case of this entity involving the glan penis. The clinical feature of nodular cutaneous amylodosis, the histopathology, the pathogenesis and the therapy are discussed.

Key word : Nodular Cutaneous Amylodosis, Glan Penis

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Nodular amyloidosis is the rarest cutaneous manifestation of amyloidosis[1]. It is characterized by single or multiple waxy, pink or brown papules, nodules[2-4], which vary in size from a few milimeters to several centimeters involving, in decreasing order of frequency, the legs, head, trunk, arms and genitalia. There are exceptional cases of plaques rather than papules or nodules. Genital nodular amyloidosis is extremely rare with only four reported cases of valvar involvement[5-6]. Herein, we report a case of nodular amyloidosis at the glan penis and review the literature pertinent to nodular cutaneous amyloidosis.

A CASE REPORT

A 50-year-old man had multiple asymptomatic, waxy, pink colored papules 2-4 mm in diameter around the dorsal surface of coronal sulcus of the glan penis for one year (Fig. 1). Physical examination of the nails was unremarkable.

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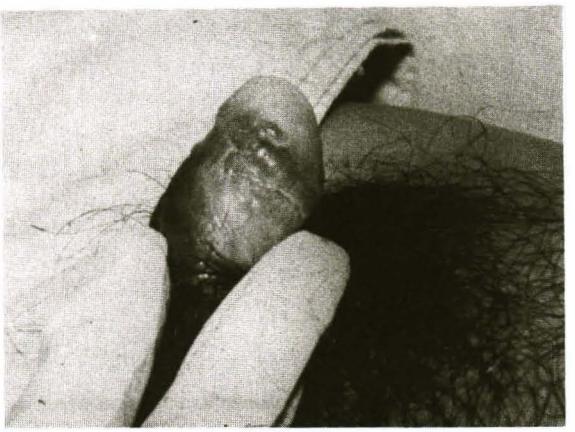

Fig. 1. Multiple asymptomatic, waxy, pink colored papules 2-4 mm in diameter around the dorsal surface of coronal sulcus of glan penis.

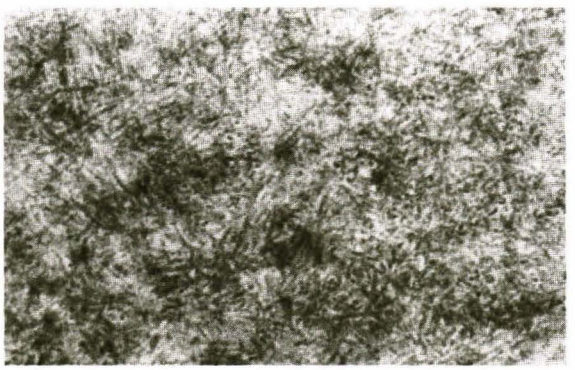

Fig. 3. Electronmicroscopic examination of the papules showed the extracellular hyaline deposit composed of straight, nonbranching filaments with diameter from 6 to 10 nm, characteristic of amyloid.

The histopathology showed atrophic epidermis with many dilated thin-walled blood vessels in the dermal papillae and upper reticular dermis. There was homogenized eosinophilic material deposit around these dilated vessels as well as along the collagen fibers. There was a mild interstitial lymphohistiocytic infilitrate and a few fibroblasts scattered in this material (Fig. 2). The Congo red stain for this

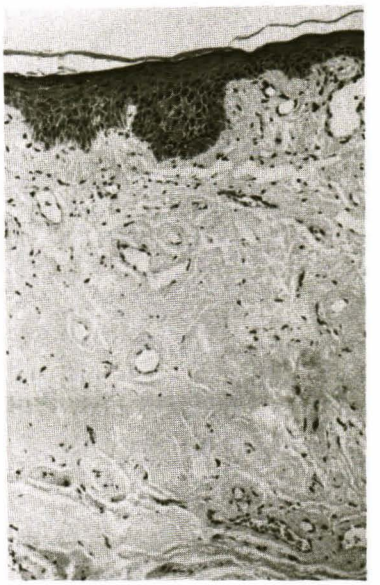

Fig. 2. The histopathology of the papules, showed atrophic epidermis with many dilated thin-walled blood vessels in the dermal papillae and upper reticular dermis. There is homogenized eosinophilic material deposit around these dilated vessels as well as along the collagen fibers.

material showed apple-green birefringence under polarizing microscope. In the electron microscopic examination, the extracellular hyaline deposits were composed of straight, nonbranching filaments with diameter from 6 to 10 nm, characteristic of amyloid (Fig. 3).

Complete blood count showed hemoglobin of 14.4 g per cent with white blood cell count of 5,300/mm^3, neutrophil 58 per cent, eosinophil 2 per cent, lymphocyte 38 per cent, monocyte 2 per cent, platelet count 187,000/mm^3. The erythrocyte sedimentation rate was 7mm/h, BUN was 14 mg/dl, Cr was 0.9 mg/dl. The liver function test, urine analysis and roentrogram of the chest were normal. The urine Bence Jone protein was negative and the serum immunoelectrophoresis showed normal pattern.

The skin lesions were surgically excised. There was no recurrent papule after 14 months.

DISCUSSION

Nodular cutaneous amyloidosis has been reported most commonly in patients in the sixth or

seventh decade with the mean age of 52 years (range from 20 to 83 years). Females are affected twice as frequently as males. The most common locations are the lower extremities, followed by the head, trunk, upper extremities and genital organs. There were only four reported cases of vulvar involvement (6). To our knowledge, this is the first reported case on the penis.

The clinical manefestation is usually multiple nodules (77%) ranging from 0.5 to 7 cm in diameter. They may be shiny, wax-like, tan-pink or yellow-brown, firm(7), rubbery(8) or anetodermic (7,9), may ulcerate(6) and may contain superficial telangiectasias(10). The clinical differential diagnosis includes inflammatory diseases such as scabietic nodule, sexually transmitted diseases such as condyloma lata and condyloma accuminata.

The histology of nodular cutaneous amyloidosis is the presence of hyaline eosinophilic material around the vessels of the entire dermis, whereas lichenoid and macular amyloidosis, in which deposits are limited to the papillary dermis(4,11,12). The deposits show apple-green birefringence with Congo red stain, crystal violet metachromasia, and thioflavine-fluorescence under a polarizing microscope. However, lipoid proteinosis and colloid milium may also show Congo red birefringence. Demonstration of typical straight, nonbranching filaments of 6 to 10 nm under electronmicroscopic study is necessary to confirm the diagnosis(11). The overling epidermis may be atrophic or anetodermic and plasma cells frequently surround the amyloid masses(8-10, 12).

The pathogenesis of amyloid deposits in nodular amyloidosis is hypothesized that a focal proliferation of plasma cells occurs, with secretion of light chain immunoglobulin. There are some studies of the nodular cutaneous amyloidosis to support this hypothesis(13-15). Of these, Breathnach et al(13) showed potassium permanganate-resistant Congophilia and lack of binding of antibodies against amyloid A protein in tissue sections. Kobayashi and Hashimoto(14) used immunofluorescence and immunoperoxidase techniques to show negative reactions with antiepidermal keratin antisera in amyloid deposits of nodular form. In contrast, positive staining was demonstrated in macular, lichenoid and basal cell epithelioma-associated amyloid. Husby et al(15, 16) demonstrated lambda-1 and kappa III immunoglobulin light chain amino acid sequences respectively, in purified tissue extracts of nodular cutaneous amyloid. Northcutt and Vanover demonstrated the presence of numerous intracellular and extracellular and extracullar Russell bodies which indicated active immunoglobulin synthesis in the histology from the biopsy of nodular cutaneous amyloidosis.

Many the therapeutic methods are used to treat cutaneous nodular amyloidosis. Intralesional corticosteroid therapy is not effective(17) and may accelerate amyloid deposition. Variable success has been noted with surgical excision, electrodessication and curettage, and cryotherapy(17,18). Truhan et al (18) reported exellent cosmetic results initially after treatment of nodular amyloidosis with carbon dioxide laser but a recurrent nodule was noted after 9 months. Recently dermabrasion was reported with effective results without recurrence after 26 months (1). The patient in this study was successfully treated with surgical excision and follow-up at 14 months showed no clinical evidence of recurrence.

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Nodular Cutaneous Amyloidosis ที่อวัยวะเพศชาย : รายงานผู้ป่วย 1 ราย

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Nodular Cutaneous Amyloidosis เป็นโรคผิวหนังที่พบได้น้อยมาก มักพบในหญิงอายุประมาณ 60–70 ปี รอยโรคที่อวัยวะเพศมีรายงานน้อยมาก พบเพียง 4 รายที่เป็นบริเวณอวัยวะเพศหญิง รายงานนี้ขอนำเสนอผู้ป่วยที่เป็นชาย ซึ่งเป็นรอยโรคบริเวณองคชาติ พร้อมได้อภิปรายเรื่องอาการแสดงของโรค พยาธิสภาพ พยาธิกำเนิด และการรักษา

คำสำคัญ : Nodular Cutaneous Amyloidosis, อวัยวะเพศชาย

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