Cutaneous Nodular Amyloidosis: An Unusual Penis Localization: Case Report

Kutanöz Nodüler Amiloidoz: Nadir Penis Tutulumu

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ABSTRACT A 42-year-old man applied to our clinic with a complaint of nodular lesions on penile skin for 5 years. He had chronic renal insufficiency and he had been a hepatitis C carrier. Dermatological examination revealed many skin colored nodular lesions both on dorsal and ventral side of corpus penis. The histopathological examination of the nodular skin lesions revealed amyloidosis. Detailed investigations for systemic involvement were found to be negative. To our knowledge, it is the first reported case of localized cutaneous amyloidosis of of penis skin, reported in Turkish literature.

Key Words: Primary amyloidosis; penile diseases


Anahtar Kelimeler: Primer amiloidoz; penis hastalıkları

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A myloidosis is characterized by extracellular deposition of a substance mainly composed of fibrous protein called amyloid. It may be deposited in any organ of the body and may produce compression and dysfunction of these organs. Primary localized cutaneous nodular amyloidosis is a rare disease in which amyloid deposition is limited to the skin and manifested as tumefactive nodules.

We present a case of primary cutaneous nodular amyloidosis localized on the skin of penis corpus in which deep tissues and glans of penis were spared. To our knowledge, it is the first reported case of localized cutaneous amyloidosis of the proximal part of penis skin.

CASE REPORT

A 42-year-old man was admitted to our clinic with a complaint of nodular lesions on penile skin. His lesions had started 5 years ago. By time, the num-
umber of the lesions had increased. He informed that he had applied to a physician previously and had had cryotherapy. However; lesions reoccurred at the same area 1 year later after cryotherapy and had increased not only in size but also in number in a 4-months duration. Medical history revealed that he had chronic renal insufficiency and he had been on hemodialysis for 20 years. The etiology of the kidney disease was unknown; however, from the history of the patient, we learnt that the illness had started after an urinary infection in childhood. He had been also a hepatitis C carrier for 17 years.

On dermatological examination, there were many skin coloured nodular lesions both on dorsal and ventral sides of corpus penis. They were smooth-surfaced and dome-shaped; the biggest nodule was approximately 1x1 cm in diameter. There was no subjective symptoms. The nodules were localized especially on the root of the penis. Glans penis was spared (Figure 1, 2). An excisional biopsy was performed. The histopathologic examination revealed massive deposition of amorphous eosinophilic material occupying whole dermis (Figure 3). This material was consistent with amyloid protein in crystal violet and congo red staining. The immunohistochemical stainings which were performed with streptavidin-biotin peroxidase technique revealed positive results for immunoglobulin kappa and lambda chain and amyloid P component (Figure 4 a,b,c). Amyloid A component was negative. No pathology was detected in the investigations of serum protein electrophoresis and serum immunofixation electrophoresis. Urinalysis could not be performed because of anuric chronic renal insufficiency. Chest radiogram, bone survey, abdominal ultrasonography and echocardiogram did not show systemic involvement. The patient was diagnosed as primary cutaneous nodular amyloidosis and he is in our follow-up for a possible development of systemic amyloidosis.

DISCUSSION

Amyloidosis is classified into two groups as systemic and localized disease. Systemic amyloidosis is more common than localized form. Primary cutaneous amyloidosis has three subgroups called...
macular, papular (lichen) and nodular amyloidosis. Nodular amyloidosis is the rarest cutaneous amyloidosis form. In nodular type, amyloid is composed of immunoglobulin light chains (kappa and lambda) and rarely β2-microglobulin. Some authors consider nodular amyloidosis as an extramedullary plasmacytoma in which amyloid fibrils are produced locally by plasma cells.

Localized amyloidosis of penis is a rare entity. There are less than 50 cases related to urethra and nine cases of glans penis in the literature. To our knowledge, this is the first case of localized cutaneous amyloidosis of penis skin without association of glans penis.

In cases associated with glans penis amyloidosis; single or multiple painless firm nodules have been reported. Kawsar et al., presented a case with plaque-like confluent lesions on glans penis. In our case, the lesions were firm, painless nodules like other cases but they were localized on the skin of corpus penis. Hematuria, hemospermia, voiding difficulties and painful erection may be the signs of urethral involvement. In our case, there was not any symptom of urethral involvement.

Dialysis-related amyloidosis is a severe complication of hemodialysis and is composed of β2-microglobulin. As our patient had been on hemodialysis for 20 years, we primarily suspected that our patient’s amyloidosis might be due to dialysis. However the immunohistochemical staining of the biopsy specimen revealed immunoglobulin kappa and lambda chain and amyloid P component. Amyloid A and β2-microglobulin were negative. Also bone and joint radiographies were normal.

Our patient’s hepatitis-C virus (HCV) serology was also positive. We investigated the possibility of amyloidosis secondary to HCV infection. However, in secondary systemic amyloidosis deposition of AA protein is seen. Erbagci et al. reported a diffuse biphasic amyloidosis (coexistence of macular and lichen amyloidosis) in an HCV seropositive patient. They thought that HCV either directly or by immunological mechanisms may cause dermoeidermal junction damage, and degenerated keratin filaments are converted into amyloid. We could
not explain the association of HCV infection in our case. We think, the seropositivity of HCV may be secondary to hemodialysis in our case.

Primary localized nodular amyloidosis may progress to systemic amyloidosis in 7-50% of patients. Therefore, these patients should be followed up for systemic evaluation. Our patient did not have any symptoms and signs of systemic amyloidosis. He is still on our follow up for a possible development of systemic amyloidosis and plasma cell dyscrasia.

As a conclusion, we believe that our case has interesting features. One of them is that he had been on dialysis for 20 years and had been a carrier of HCV for 17 years accompanying primary localized nodular amyloidosis. On the other hand no systemic involvement was detected that would point to a primary or secondary systemic amyloidosis. Besides, the penile localization of nodular amyloidosis makes this case different.

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REFERENCES