Case Report

Amyloidosis Presented with Whitening and loss of Hair which Improved after Dimethylsulfoxide (DMSO) Treatment

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A 67-year-old male patient presented with rapid progression of whitening and loss of hair in past 2 months was consulted due to the suspicion of hypothyroidism. He had been told to have cardiomegaly for 3 years. Thyroid function was within normal limit. Prostate biopsy was performed because of prostatic hypertrophy and mild elevation of serum acid phosphatase. Amyloid accumulation was observed in the biopsy specimen. Subsequent skin biopsies revealed the same result. The scalp hair and beard grew and turned to black gradually several months after dimethyl sulfoxide (DMSO) treatment. These findings suggest that some of the manifestation of amyloidosis may respond to DMSO treatment.

Key Words: Amyloidosis, Whitening and loss of hair, Dimethyl sulfoxide (DMSO)

Amyloidosis was considered rare, and the incidence was reported to be 0.5% in the total autopsy cases, less than one half of these cases had been diagnosed clinically.¹⁾ Thus amyloidosis may be often unrecognized antemortem. Although loss of hair was reported in some cases of amyloidosis by dermatologist,²⁾ the incidence may be rare or unnoticed because there was no description about loss of hair in other studies containing 236 cases,³⁾ and 42 cases.⁴⁾ Furthermore, whitening of hair has not been described in the reported cases of amyloidosis. On the other hand, cardiomegaly is common in amyloidosis.^{3,4)} Here we report a case of primary amyloidosis presented with rapid progression of whitening and loss of hair 3 years after the finding of cardiomegaly. The condition of hair improved several months after dimethyl sulfoxide (DMSO) treatment.

CASE REPORT

In November 1982, a 64-year-old male patient was told to have cardiomegaly after a physical check up. He had no history of hypertension or rheumatic disease. Three months later he visited a local medical practitioner because of the pitting

edema of lower legs. Cardiomegaly, atrial fibrillation and hepatomegaly were diagnosed. Edema subsided after diuretic treatment. However, he felt occasional left chest pain since September, 1984. The condition of hair had been normal at that time (Figure 1) until May 1985 when rapid progression of whitening and loss of hair appeared. Two months later, the beard, axillary and genital hair lost completely. The scalp hair and eyebrow turned white and reduced in amount (Figure 2). He visited a dermatologist in August 1985 and was transferred to the outpatient clinic of Endocrinology division for the suspicion of hypothyroidism.

Physical examination revealed mild macroglossia and hepatomegaly (2 fingers breadth below right costal margin with elastic consistency and smooth surface). Chest X-ray showed cardiomegaly (cardio-thoracic ratio: 60.7%). ECG revealed atrial fibrillation. Cardiac echogram showed dilatation of atrium and ventricles and mild insufficiency of mitral and tricuspid valves. Serum T₃, T₄, BUN and creatinine values were within normal limit. Serum GOT and GPT values were slightly elevated (GOT 39 mU/ml, normal 11–30 mU/

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Fig. 1. The picture of the patient taken at 1984.

Fig. 2. The picture of the patient taken at November 1985 before DMSO treatment.

ml; GPT 39 mU/ml, normal 4–30 mU/ml). Urine protein was negative. Since serum acid phosphatase was slightly elevated (4.01 U, normal <4 U) and the prostatic hypertrophy (walnut size) was observed, he was transferred to the urologist, and prostate biopsy was performed. Amyloid was identified by congo red stain in the specimen.

After diagnosis, he had been admitted to the Jichi Medical School Hospital between November

Fig. 3. Scalp biopsy showed amyloid infiltration (Congo red stain) $\times 40$: Amyloid substance located near the atrophied hair follicle (arrow).

Fig. 4. The picture of the patient taken at October 1986, 10 months after DMSO treatment.

5 and December 18, 1985 for further examination. Further studies showed that serum protein value of 5.7 g/dl, albumin 3.7 g/dl, globulin 2.0 g/dl and normal value of prostatic acid phosphatase. Serum immunoglobulin electrophoresis revealed that IgG value of 1026 mg/dl, IgA value of 97 mg/dl, IgM value of 27 mg/dl, IgD value of 4 mg/dl, IgE value of 1350 IU/ml (normal <500 IU/ml). M protein was not found. Bone marrow showed mild increase of mature plasmocyte. Abdominal echogram showed normal pattern of liver, spleen, pancreas and kidney.

Repeated skin biopsies (scalp, leg, back) showed amyloid substance around the blood vessels and papilla (Figure 3, scalp). The substance was resistant to the treatment of potassium permanganate. He was diagnosed to have primary amyloidosis.

DMSO treatment was started on December 5, 1985 (2.5 ml, orally two times a day, 1 ml = 78%). About 8 months after treatment, the scalp hair and beard began to grow and turn to black gradually (Figure 4). Cardiomegaly and hepatomegaly still exist after therapy however the severity seems not aggravate by symptoms, X-ray and physical examination.

COMMENTS

The diagnosis of amyloidosis depends on tissue biopsy. Although it was accepted that rectum biopsy was a safe and highly diagnostic method,³⁾ there was a report showing that prostate biopsy was also a simple and good diagnostic method.⁵⁾ In our study, prostate biopsy gave the clue to diagnosis and it was confirmed by subsequent skin biopsies. Unless the patient had enlarged prostate with slightly elevated serum acid phosphatase, the diagnosis would have been delayed, because this relatively rare disease presented itself with such uncommon symptoms of rapid onset on whitening and loss of hair.

Since the patient had no obvious chronic inflammatory disorder and the specimen of biopsy resisted to potassium permanganate,⁶⁾ he was considered to have primary amyloidosis. Heart disease is very common in primary amyloidosis, and is reported to involve 90% of the patients with primary amyloidosis.⁴⁾ Amyloid can invade myocardium, endocardium, valves, conduction system, pericardi-

um, coronary vessels, causing congestive heart failure, valvular disease, arrhythmia and angina pectoris clinically.^{7,8)} Hepatomegaly is also common in amyloidosis.³⁾ The patient had been found to have cardiomegaly and hepatomegaly about 3 years before the symptoms of whitening and loss of hair. Thus, it is likely that cardiomegaly and hepatomegaly appeared as manifestations of amyloidosis.

The loss and whitening of hair also probably resulted from deposition of amyloid substance as shown by the skin biopsies of the scalp. The course of primary amyloidosis is usually progressive. There were only a few controversial reports about the effect of treatment of primary amyloidosis with DMSO.^{9,10)} In this patient, the regrowth and blackening of the hair seems to be brought about by the DMSO treatment. The result suggests that DMSO may be effective in some of the symptoms of amyloidosis. No adverse effect was noted during the treatment except for the unpleasant odor of the drug.

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