PSEUDOXANTHOMA ELASTICUM (GRONBLAD-STRANDBERG SYNDROME)

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Summary

A case of Gronblad-Strandberg syndrome with characteristic skin changes and angioid streaks in both eyes without any cardio-vascular lesions occurring in a 35 years old female is described. Histological examination of the skin from the affected site revealed extensive degeneration of elastic tissue with deposition of calcium. There was no history of consanguinity in parents. No other member of the family was affected. A brief account of relevant literature is given.

Pseudoxanthoma elasticum

(PXE) is an uncommon hereditary systemic elastorrhexis involving the cutaneous, ocular and cardio-vascular systems. The characteristic lesions are small, multiple, waxy yellow, lax redundant papules over skin folds like neck, axilla etc., and angioid streaks in the elastic lamina of Bruch with cardio-vascular abnormalities like hypertension, haemorrhages, cardiac ischaemia etc. Degeneration of elastic fibers with deposition of calcium is the basic pathology.

Balzer[1] in 1884 described the skin lesions of PXE while reporting an autopsy of a patient who died of Tuberculosis. He believed it to be one of the true xanthomatoses. Darier[2] in 1896 described this as a separate entity and conferred the present name PXE based on the histopathological features of the disease.

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The association of skin lesions and angioid streaks in a patient of PXE is known as Gronblad-Strandberg syndrome. This association was first reported by an ophthalmologist Gronblad[3] and dermatologist Strandberg in Sweden.

Nearly 400 cases of PXE have been reported in literature since its original description but cases reported from India are not many[4-12]. We are presenting here a case of Gronblad-Strandberg Syndrome.

Case Report

A female patient aged 35 years reported to the out-patient clinic of skin and V. D. of S. S. Hospital, Varanasi with the complaints of multiple, small yellow coloured papules and laxity of skin over the folds of neck and axillae for 7 years. The lesions were asymptomatic except for the cosmetic defect. She also complained of diminution of vision in both eyes for 7 months. There was no history of consanguinity in parents and no other member in the family was affected.

Examination revealed the presence of multiple, variously sized (3 to 6 mm),

waxy papules situated over the folds of neck, axillae, anti-cubital fossae and periumbielical regions (Fig. 1 Page No. 92). The skin over these sites and eyelids was hyperextensible with loss of elasticity. Shiny, discrete whitish-yellow papular lesions were present on the mucous membranes of soft palate and cheeks. No calcification was observed clinically. Examination of other systems did not reveal any abnormality. Peripheral pulses were normal.

Investigations

BP 110/80 mm of Hg: Haemogramme, blood sugar and serum cholesterol were within normal limits. X-ray chest and barium meal series were normal.

Ophthalmoscopy

Fundus examination revealed the presence of greyish brown streaks radiating from the optic disc (angioid streaks) in both eyes.

Skin biopsy

Histological examination of the skin revealed extensive degeneration of elastic tissue in the dermis indicated by pale staining with H & E stain. Special staining of the section with Verhoeff and counter-staining with Van Gieson demonstrated the darkly stained, swollen, fragmented and curled elastic fibers in the dermis. Collagen fibers stained pink and appeared normal (Fig. 2 Page No. 92). Staining with Von Kossa revealed blackish discolouration in the mid-dermis indicating the deposition of calcium.

Discussion

Gronblad-Strandberg syndrome is a rare autosomal recessive disorder with partial sex limitation to females. It has been stated that the skin lesions are common in females and angioid streaks are more frequent in males[13]. Both these lesions were present in out-patient who was a female. Angioid streaks are also seen in paget's disease, sickle cell disease, familial hyperphosphataemia, idiopathic thrombocytopenic purpura, lead poisoning[10] and Ehlers-Danlos syndrome. The other lesions reported in the fundus of Gronblad-Strandberg syndrome, which were not present are mottling by pigmentary deposits, choroidal atrophy and sclerosis, atrophy of the pigmentary epithelium and retinal detachment[14]. Absence of consanguinity in parents and absent family history of the disease have also been reported by others[12]. Cardiovascular abnormalities have been reported in upto 85% patients of PXE[13]. In the present case no such lesions were observed.

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TRUE

Treatment of chromoblastomycosis has been unsatisfactory in the past. Recurrence is frequent after excision or curettage and cautery. A number of favourable reports on the use of 5-FC in this condition have appeared. Susceptible fungi deaminate 5-FC to 5-Fluorouracil intracellularly, which is then toxic since 5-Fluorouracil is a known metabolic antagonist.

Human tissues lack the deaminase and so do not metabolize 5-FC. Even topical use of 10% ointment of 5-FC alone has been claimed to be curative. At this time 5-FC may well be the drug of choice for Chromoblastomycosis.

Ref: Year Book of Dermatology 1975. p 97.