Case 1

A 28-year-old woman

Chief Compliant: Beaded like papules eruption on both upper eye lid margin.

Since she was 10 years old, she suffered from hoarseness Present illness:

10 years later, she developed several asymptomatic shiny whitish papules on the margin of both upper lids. The lesions did not seem to progress.

She has normal growth and development Past History:

She has no history of seizure or other skin lesions

3 years ago, multiple cystic masses were palpated on her right breast and diagnosed as fibrocystic disease by surgeon.

She was reported traumatic subluxation and dislocation of her right hip joint since she was a young child developed permanent joint destruction.

Family History:

Physical examination: A healthy woman,

nil

HEENT Several small pale whitish translucent papules along the fine margin of the both upper eye lids, hoarseness of voice caused by hypertrophy of right vocal cord possible from substance deposition revealed by otolaryngologist.

no other skin lesions in the oral cavity

Skin multiple movable cysts in varying diameters palpable in right breast

Abdomen liver and spleen not palpable

CNS good consciousness, intelligence was within normal range

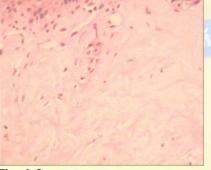
Investigation:

Breast sonogram

multiple cysts at 5 o'clock of right breast

X-ray hip joint Pelvic tilt due to scoliosis, No evidence of fracture or dislocation. Osteoporosis of the right femoral head, The LT.- hip is within normal limits





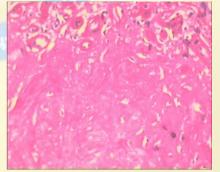


Fig. 1.1

Fig. 1.2

Fig. 1.3

- Histopathology Extensive deposit of homogeneous eosinophilic material with PAS-positive and diastaseresistance around capillaries in upper dermis.
- The bundles often oriented perpendicular to the skin surface.

(s99-9095)

Electron microscopy:

- Thickening of the basal laminae .
- Massive deposite of hyaline.
- Marked reduction of collagen fibrils.

Diagnosis: Lipoid proteinosis

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