Connective tissue nevi

Connective tissue nevi (CTN) are hamartomas in which components of the dermis (collagen, elastin, or glycosaminoglycans) are altered. Appear as firm, asymptomatic, skin-colored plaques composed of closely grouped papules with an orange peel-like surface texture.

Previous classifications made based on which dermal component was found in excess, genetic patterns of inheritance, and +/- organ involvement fail to explain all reported cases. oExample: Buschke-Ollendorff syndrome (BOS), an autosomal dominant form characterized by juvenile elastomas a/w osteopoikilosis (multiple osteosclerotic bone lesions), where phenotypic expression varies. Objective:

Characterize the clinical and histopathologic features of CTN and to highlight a spectrum of clinical disease. Methods: Retrospective study of cases selected based on strict clinical and histopathologic criteria. Patients identified by searching investigator's clinical and histopathology files using key words. Total of 33 patients with CTN included in study, 31 children and two adults.

One pathologist performed all histopathologic analyses. oSpecimens divided into 4 histopathologic groups: 1) " pure" collagenoma; 2) " pure elastoma"; 3) mixed type CTN with both collagen and elastic changes; 4) cellular CTN which an increased number of normal looking fibroblasts was observed + fiber changes. Results:

Average age of onset at which the first signs were observed was 2 years. Three clinical forms were distinguished: otype A characterized by localized papules confined to one site, with one case presenting as an ulcerated infiltrated plaque otype B with two or more sites of involvement most often

on thighs, lumbar region, thorax, abdomen, buttocks, arms, and legs otype C with unusually severe infiltration with functional impairment of a limb.

Histopathologic examination of lesional biopsy specimens showed 10 collagenomas, one elastoma, 18 mixed CTN, and an increased number of short, nonfasciculated spindle-shaped cells with small regular ovoid nuclei and scant cytoplasm resembling fibroblasts in 4 cases.

There was no correlation between clinical type and histopathologic findings. Discussion:

Type A: 36% of cases, histopathologic features corresponded to collagenomas, mixed forms, or cellular form with increased number of fibroblasts. Type B, 55% collagenomas, elastoma, and mixed forms were observed. Type C, of the 3 cases, two were related to mixed forms and one to cellular form. Histopathology did not provide information on prognosis. Of 5 patients with collagenomas, one had unexpected osteopoikilosis, which raises 2 hypotheses: either an elastic abnormality was present and undetected or there is heterogeneity in the type of CTN found in BOS. Physiotherapy was only treatment method of benefit in the monomelic forms that could cause functional impairment