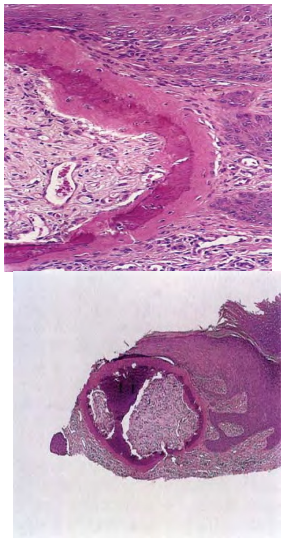

Osteoma Cutis

Strictly defined, osteoma cutis refers to the presence of bone within the skin in the absence of a preexisting or associated lesion, as opposed to secondary types of cutaneous ossification that occur by metaplastic reaction to inflammatory, traumatic, and neoplastic processes.

Bone arises in skin and soft tissues through mesenchymal (membranous) ossification without cartilage precursors or models (enchondral ossification, as in the skeletal system).

The lesions of osteoma cutis differ from tumoral calcinosis in that they represent bone formation, and calcinosis refers to calcium salt deposits.

In addition to colonic polyposis , epidermal cysts of the face and the scalp, and multiple fibromas, osteomatosis in the form of intraosseous (not cutaneous) osteomas may develop within the membranous bones of the head in Gardner syndrome .

Although said to be rare, with no well-defined data on the incidence, a plethora of conditions and syndromes may be found in association with osteoma cutis, and the frequency of its occurrence varies accordingly. Primary lesions with no underlying cause are even rarer, but they account for approximately 20% of all cases.

Osteoma cutis is not life threatening, although local discomfort and/or disfigurement may lead the patient to seek consultation.

No particular race is predisposed to developing osteoma cutis.

Generally, no distinct sexual predominance exists. However, one cause of osteoma cutis, Albright hereditary osteodystrophy, occurs with a female-to-male ratio of 2:1.

Osteoma cutis may occur at any age.

- Patients may report having hard areas in the skin.
- A familial occurrence of Albright hereditary osteodystrophy may be present.

- The presentations of osteoma cutis can be highly variable, with clinical entities that are defined by the number, the form, and the location of the lesions.
- Osteomas may present as single or multiple, extremely hard nodules, plaques, or miliary tumors.
- The face, the extremities, the scalp, the digits, and the subungual regions are the most commonly affected sites.

Causes

Osteoma cutis is a feature in several groups of patients.

- Albright hereditary osteodystrophy, which includes most patients with pseudohypoparathyroidism and pseudopseudohypoparathyroidism, is due to an autosomal dominant defect in the alpha subunit of intracellular guanyl nucleotide-binding protein (G protein).
- The characteristic phenotype includes short stature, a round face, defective teeth, mental retardation, brachydactyly, and osteomas of the soft tissue and the skin.
- Tetany is often the presenting sign of pseudohypoparathyroidism, formerly called Albright hereditary osteodystrophy. In addition to skeletal system abnormalities, lesions of osteoma cutis are frequently observed.

- Single, small osteomas, arising later in life, sometimes with transepidermal elimination of bony fragments may be a cause.
- Multiple miliary osteomas of the face following acne, neurotic excoriation, or dermabrasion are possible causes.
- Congenital plaquelike osteomatosis or limited dermal ossification is generally present from birth; the skin of the scalp or the extremities is often affected.
- Fibrodysplasia ossificans heteroplasia and fibrodysplasia ossificans progressiva are possible causes.
- Miscellaneous rare disorders with or without cartilaginous elements include osteomas of the distal extremities and multiple osteomas of childhood unrelated to Albright hereditary osteodystrophy.