



AUTOIMMUNE CONNECTIVE TISSUE DISEASES

PLATE-LIKE OSTEOMA CUTIS IN THE SETTING OF NEPHROGENIC SYSTEMIC FIBROSIS

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Background: Osteoma cutis is the aberrant formation of bone within the skin associated with various neoplastic, traumatic, and inflammatory processes, including connective tissue disease. Nephrogenic systemic fibrosis (NSF) is the development of cutaneous sclerosis following gadolinium contrast administration, most often in the setting of renal failure. Cutaneous osseous metaplasia has been described in five cases of NSF, often occurring later in the disease course with clinical features atypical for classic NSF, including palpable bony masses and white nodules. We present a case in which extensive plate-like osteoma cutis developed on the lower extremities in longstanding plaques of NSF.

Observation: A woman in her 40s with longstanding, severe systemic lupus erythematosus complicated by renal insufficiency presented with indurated plaques of the elbows, thighs, and lower legs. Biopsy showed typical findings of NSF. The patient had multiple MRIs with gadolinium in the past, and gadolinium staining of the tissue was positive (138.5 ppm). Over the subsequent years, she developed progressive, painful rock-hard subcutaneous nodules coalescing into plate-like plaques within the existing sclerotic NSF lesions. On pathology, the papillary dermis was largely replaced by a sheet of metaplastic bone, consistent with osteoma cutis. After failing to respond to diltiazem and colchicine, progression of the patient's osteoma cutis halted with IVIG, and softening of plaques and was noted with decreased pain. Although there are five reports of NSF associated with osseous metaplasia in the literature, none presented with extensive, plate-like disease similar to our patient. Furthermore, this is the first report of improvement following IVIG.

Key message:

1. Osteoma cutis is a rare, but well-described complication of NSF that presents with palpable bony nodules within sclerotic NSF plaques.
2. No uniformly effective treatments exist, but IVIG may be an option to halt progression and/or soften existing plaques.

