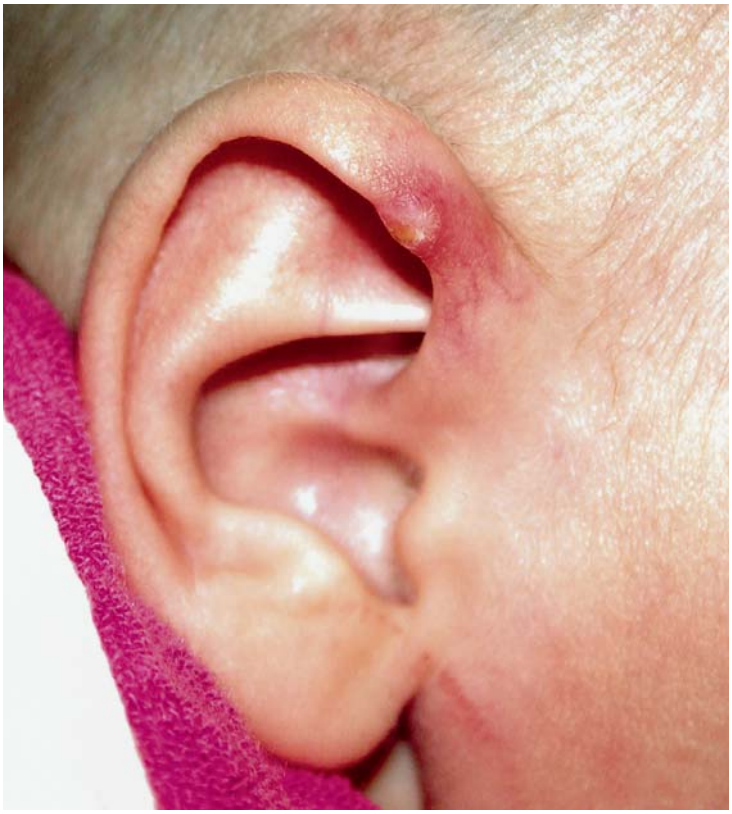
What Is Your Diagnosis?



A 3-month-old infant presented with an asymptomatic nodule on the helix of her right ear that had been present since birth.

PLEASE TURN TO PAGE 90 FOR DISCUSSION

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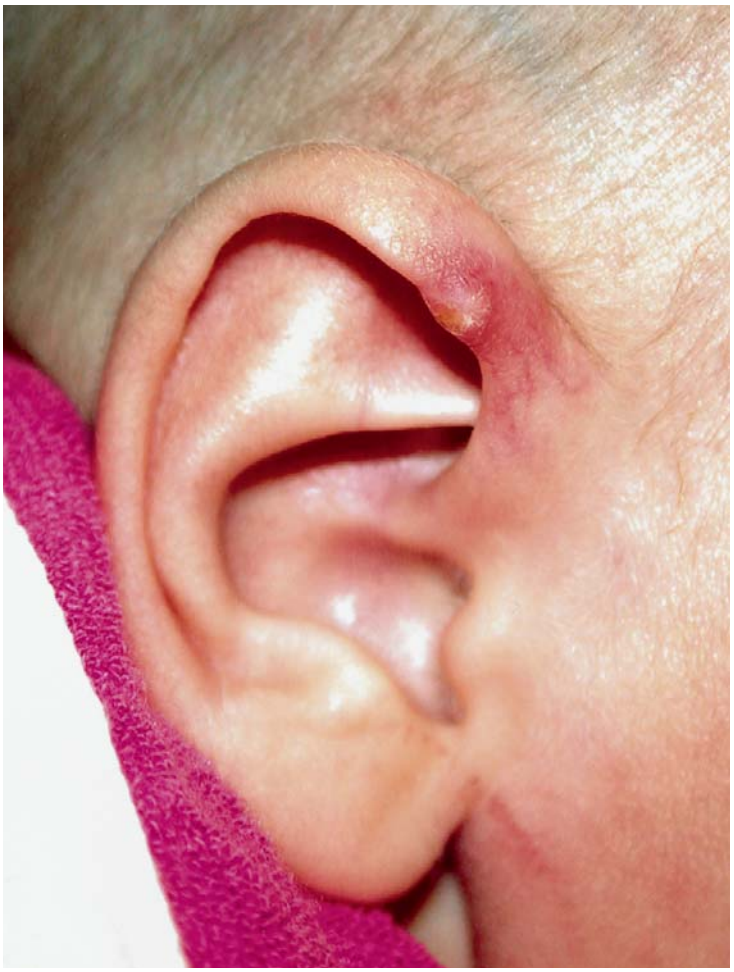
The authors report no conflict of interest.

The Diagnosis: Solitary Congenital Nodular Calcification



Solitary congenital nodular calcification (also known as Winer nodular calcinosis and subepidermal calcified nodule) was first described by Winer[1] in 1952 in a report of 3 children with calcific nodules on the knee, index finger, and plantar region. Since then, 19 cases of congenital calcification of the ear have been reported in the literature.[2-7] A congenital nodular calcification is noted at birth or in early childhood and presents as an asymptomatic, firm, yellow or white nodule on the ear, face, scalp, or extremities.[3] The surface may be smooth or verrucous, and with gentle pressure, the nodules may occasionally discharge a chalky substance. Case studies have reported a male-female ratio of 2:1.[3,5]

There are 4 types of calcinosis cutis: dystrophic, metastatic, iatrogenic/traumatic, and idiopathic.[8] In dystrophic calcinosis cutis, deposition of calcium occurs secondary to damage to connective tissue, and there are normal serum levels of calcium and phosphorus. This is seen in conditions such as CREST syndrome, systemic lupus erythematosus, and dermatomyositis. Metastatic calcinosis cutis is seen in conditions such as hyperparathyroidism, hypervitaminosis D, the milk-alkali syndrome, sarcoidosis, and renal failure and is due to aberrant metabolism of calcium and phosphorus with subsequent deposition of calcium salts in normal tissues. Extravasation of calcium chloride infusion is one example of iatrogenic/traumatic

calcinosis cutis in which calcium-containing materials are introduced to tissues following minor trauma. Idiopathic calcinosis cutis occurs in otherwise normal tissue with normal levels of calcium and phosphate metabolism.

Solitary congenital nodular calcification represents a form of idiopathic calcinosis cutis, though the precise etiology remains unclear. Mehregan[6] described the appearance of a congenital calcified nodule of the ear in a neonate whose helix was folded at birth, which suggests that trauma may play a role.[6] Winer[1] postulated that the nodules are formed following calcification of a sweat duct hamartoma. Other theories include calcification of a pre-existing pilomatricoma, nevus, or wart; degranulation of mast cells with subsequent calcification; and calcification of subcuataneous fat necrosis.[2] In a review of 21 cases, there was no evidence of pilomatricoma or nevus nor was there a correlation between mast cell distribution and histologic appearance of the lesion.[5] Won et al[7] described a subepidermal calcified nodule that occurred within a hair follicle nevus. Although the precise etiology remains unclear, it is thought that a subepidermal calcified nodule of the ear represents a specific entity.

Shave or surgical excision is the treatment of choice for a subepidermal calcified nodule. The diagnosis can then be confirmed histologically. These lesions exhibit hyperkeratosis, acanthosis, and amorphous calcium deposition in the papillary dermis. Occasionally there is ulceration with transepidermal elimination of calcium.

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