Case for diagnosis

Caso para diagnóstico

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DISEASE HISTORY

Twenty-three year-old man had noticed the emergence of lumps in the neck that had started 5 years before with progressive increase in the number of such lesions in the anterosuperior region of the chest and also in the inguinal region ,some evolving into erythematous lesions with drainage of a yellowish fluid with strong odor. The patient denied systemic symptoms. Background: he also denied similar lesions in relatives. When clinically examined the patient presented papulonodular lesions varying from 0,5 to 2,0 cm, of a yellowish color mainly in the anterosuperior chest, lateral cervical region, (Figure 1) and bilateral inguinal region. Histopathological examination of skin fragment taken from the anterior region of the chest showed presence of cystic formation with sebaceous gland on its wall in the dermis (Figure2) and typical stratified squamous epithelium that showed its stratum corneum wavy shaped, refractile and strongly eosinophilic located in the cystic wall (Figure 3). Laboratory tests without alterations.

FIGURE 1: Papulonodular lesions varying from skin to a yellowish color, and from 0,5 to 2,0 cm, located in the neck and anterosuperior region of the chest

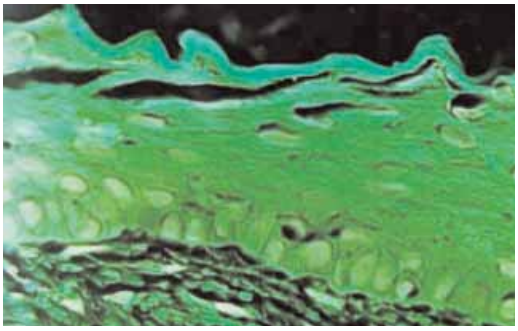

FIGURE 3: Cystic wall covered by typical stratified squamous epithelium that showed its stratum corneum wavy shaped, refractile and strongly eosinophilic. These characteristics are observed in the sebaceous gland duct. (HE, 1000X)

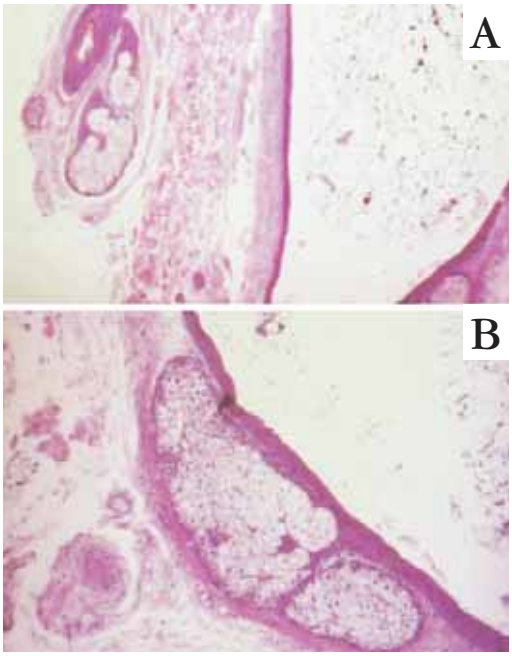

FIGURE 2: A. Cystic formation located in the dermis.(HE, 40X); **B.** Cystic formation with sebaceous gland in its wall (HE,100X)

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COMMENTS

Steatocystoma multiplex is a rare genetic disorder that it is characterized by multiple dermal cysts, asymptomatic and of variable size.[1,2,3] Its etiopathogenesis remains obscure but there are many theories to explain its origin such as: it results from sebaceous retention cysts, nevoid nature or hamartomas; or they are a variety of dermoid cyst. [4]

Lesions are most commonly found on the chest and proximal extremities although other sites are also described such as: axillae, vulva, central area of the chest, and inguinal region (common in women). Other regions like face and scalp are rare to be found.[3] Lesions grow slowly and they have a content that can be liquid or creamy. [1] Although the majority of them are asymptomatic, some lesions can inflame and fester.[3]

Differential diagnosis is made in many diseases: acne conglobata or cystic nodule, hidradenitis suppurativa, dermoid cyst, milia, follicle cyst, myxoid cyst and beard pseudofolliculitis.[4]

The disease begins in adolescence and early adulthood and equally incurs in both sexes. It is an autosomal dominant disease although various sporadic cases have been described. [1,2] It has been associated with paronychia congenita type 2, hypertrophic lichen planus, hidradenitis suppurativa, acrokeratosis verruciformis and hypohidrosis.[1,5]

Mutation of keratin 17, a protein found in several epithelial structures such as ungual lay, hair follicles and sebaceous glands has been associated with the genesis of familiar steatocystoma, as well as in patients with paronychia congenita type 2. As for the sporadic cases, this mutation was not found suggesting that the disease has a multifactorial feature.[2,6]

Histologically, steatocystomas are dermal cysts covered by an eosinophilic and wavy layer of epithelial tissue. Sebaceous glands in general are present in the cyst wall and hairs can occur in its antrum.[1,4]

Treatment options are scarce and a few present a satisfactory result.[1,2] Although this condition is not a threat to individual health it frequently is a cosmetic problem which justifies its treatment. Needle aspiration decreases the size of the lesions but the result remains only for some months.[7] Surgical excision through various techniques is also described.[1] Intralesional injection of corticosteroids and incision and drainage are good options for inflamed lesions.[2] Isotretinoin is known for its anti-inflammatory property and for this reason it is advised for ulcerating lesions but with recurrences in some cases[8,9]. The association of isotretinoin and cryotherapy in non ulcerating lesions showed good clinical and cosmetic response. Cryotherapy and dermabrasion can also be used but they showed limited results besides leaving a residual scar.[7] The use of CO2 laser is an ideal technique for the treatment of multiple lesions and/or lesions located in areas aesthetically important such as the face. It does not require anesthesia, the lesions are treated in a single session, it is a minimally invasive procedure presenting quick healing, good aesthetic results and low percentage of recurrence.[10] ❑

Resumo: Esteatocistoma múltiplo é um raro transtorno genético autossômico dominante que se caracteriza por múltiplos cistos dérmicos de tamanho variável e assintomáticos. Descreve-se o caso de um paciente do sexo masculino, de 23 anos, com quadro clínico e evolutivo típicos dessa desordem.
Palavras-chave: Cistos; Mutação; Queratina-17

Abstract: Steatocystoma multiplex is a rare genetic disorder, autosomal dominant, that is characterized by multiple asymptomatic dermal cysts which vary in size. It is described here the case of a 23 year-old male patient with a typical clinical and evolutional progression of this disease.
Keywords: Cysts; Mutation; Keratin-17

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