STEATOCYSTOMA MULTIPLEX

(A case report)

By

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According to Mount (1937), Bosellini in 1898 gave the first description of this order under the title of 'Multiple Follicular Cutaneous Cysts.' Subsequently, it has been described under several other names such as sebocystomatosis, hereditary epidermal polycystic disease and steatocystoma multiplex (Rook *et al*, 1968). The most widely used terminology however, is steatocystoma multiplex (Mount, 1937; Sachs, 1938, Noojin and Reynold, 1948; Schiff *et al*, 1958; Kligman and Kirschbaum, 1964).

The disease is characterised by the development of multiple, asymptomatic, yellowish tumours on the trunk, extremities and the face. In some cases, scrotum may be the only site involved (Ronchese, 1944). The tumours vary in size and are attached to the overlying skin. At times comedones may also be observed. The disease usually begins in adolescence or early adult life (Mount, 1937), but cases have been recorded where the disease started during infancy or was present even at birth (Sachs, 1938).

Familial occurrence has been recorded by Klausner (1917); Ingram and Oldfield (1937), Sachs (1938) and Noojin and Reynold (1948). It is believed to be transmitted as a dominant trait.

Kligman and Kirschbaum (1964) studied the histopathology of this condition in detail and observed that the cyst is usually situated in the mid dermis. It is lined by a thin keratinizing epithelium and characteristically contains hair follicles, sebaceous glands and eccrine sweat glands. On the basis of these findings these tumors were considered to be a variety of dermoid cyst.

The present report concerns 4 cases of steatocystoma multiplex in a family, with their clinical and histopathological characteristics.

Case Reports

Case 1 : A 32 years old female presented with multiple, yellowish, firm, non tender nodules of varying sizes, located mainly on the face, neck and presternal areas (Fig. 1). They started appearing at the age of 5 or 6 years and were gradually increasing in size and number.

Case 2 : Sixteen years old eldest son of Case No. 1 came with similar lesions which started at the age of 7 or 8 years. The lesions were smaller in size and less in number than those of Case No. 1 and they were distributed on the face, neck, trunk and scrotum. On some lesions a black punctum was also seen.

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Case 3: Twelve years old second son of Case No. 1 presented with a similar picture although the nodules were still smaller in size and less in number (Fig. 2).

Case 4: Six years old fifth daughter of Case No. 1 had only a few lesions distributed on the nasolabial folds and forehead.

Biopsies taken from Case Nos. 1, 2 and 3 revealed in each case an intricately folded cyst whose wall was lined by keratinizing epithelium (Fig. 3) In some areas, there were large cells possessing abundant eosinophilic cytoplasm that projected into the lumen in a fashion similar to that of apocrine secretion. At places sebaceous lobules were seen in the wall of the cyst (Fig. 4) and in one section a hair follicle was also seen lying in the lumen of the cyst.

Comments

The clinical and histological features of these cases are quite characteristic as described by Schiff *et al* (1958); Kligman and Kirschbaum (1964) and Lever (1967). The presence of keratinizing epithelium, apocrine glands, sebaceous glands and hair follicles could be demonstrated in the sections studied, conforming to the views of Kligman and Kirschbaum (1964) that steatocystomas are true dermoid tumours representing a wayward development of the primary epithelial germ. Pedigree analysis (Fig. 5) although traced only to three generations shows that the disease manifested in both sexes in subsequent generations. There was no history of consanguinity of marriage. These findings corroborate the views of Klausner (1917); Ingram and Oldfield (1937); Sachs (1938) and Noojin and Reynold (1948) that the disease is transmitted as an autosomal dominant.

Summary

Four cases of steatocystoma multiplex occuring in a family are presented. Histologically, the tumours were lined by keratinizing epithelium containing apocrine glands, sebaceous glands and hair follicles. The view that these tumours are dermoid cysts is corroborated.

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