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Eruptive vellus hair cysts and steatocystoma multiplex. Variants of one entity?

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Summary Eruptive vellus hair cysts and steatocystoma multiplex are two clinically similar conditions which show multiple papules and nodules, mainly located over the anterior chest wall. Most cases can be differentiated on histological examination, but in some patients overlapping histological features have been described. We present a patient who showed features of both entities and interpret this as suggesting that eruptive vellus hair cysts and steatocystoma multiplex are variants of one disorder which originates in the pilosebaceous duct.

The condition of eruptive vellus hair cysts was first described in 1977.[1] Esterly and colleagues described a 6-year-old ghrland a 7-year-old boy who had asymptomatic distinctive hyperpigmented and monomorphic papules over the anterior chest wall.[1] Histological examination revealed that the papules were composed of small keratin-filled cysts located in the mid-dermis, and containing numerous transversely or obliquely cut vellus hair shafts.[1] Subsequently, this condition was shown to have an autosomal dominant inheritance.[2] Over recent years, several studies[3–6] have revealed histopathological similarities between eruptive vellus hair cysts and steatocystoma multiplex, a condition which was first described in 1899.[7] Although typically these disorders show different histological characteristics, they have several clinical features in common. These include their age of onset, localization, the appearance of the lesions, and the mode of inheritance. Cases in which the cysts histologically show features of both steatocystoma multiplex and eruptive vellus hair cysts have been described as hybrid cysts.[3,6]

Case report

A 22-year-old woman presented with 40–50 asysmptomatic nodules and papules over the anterior chest wall. The lesions had first appeared at the age of 15 years and had increased in size, number and extent. There was no family history of similar lesions. Examination showed smooth, skin-coloured or light-brown nodules and papules, which varied in size between 2 and 5 mm (Fig. 1). She had been on no medication and was otherwise well. Skin biopsies had been taken in 1991 and 1994.

Histological examination of both biopsy specimens revealed folded cystic tumours in the mid-dermis. The first biopsy showed an associated rudimentary follicular structure attached to the cyst. The cyst was lined by between two and six layers of stratified squamous epithelium which showed a discrete granular layer, and contained fragmented vellus hairs in addition to keratinous material. The second biopsy (Figs 2 and 3) showed similar features but, in addition, step sections showed sebaceous elements in a single and grouped fashion within the cyst wall, and protruding into its lumen (Figs 4 and 5). Both specimens showed a stratified epithelial layer with a sometimes small but detectable granular layer (Figs 3 and 4).

Discussion

Since the original description of eruptive vellus hair cysts, there have been a number of case reports but few

Figure 1. Multiple papules and nodules.

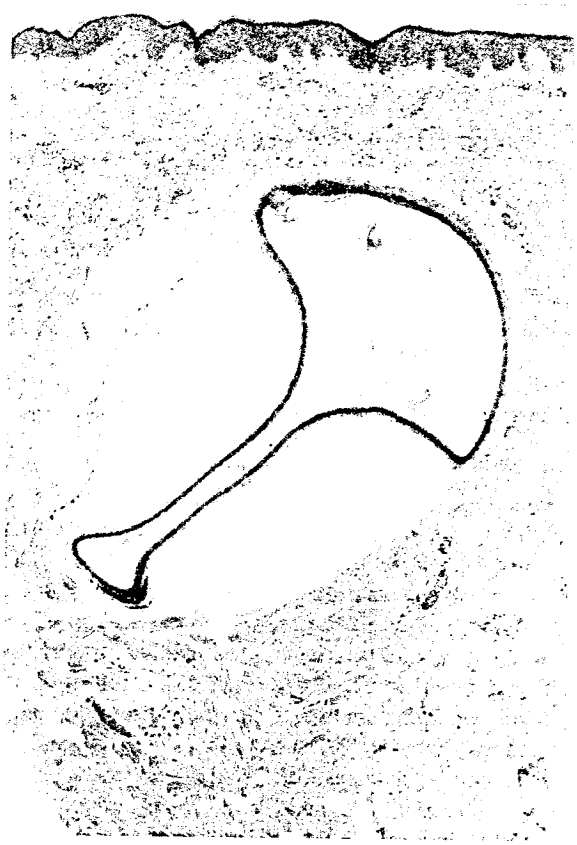

Figure 2. Folded cystic proliferation in the mid-dermis (haematoxylin and eosin × 40).

cohort studies. In most reported patients, the eruption of several papules over the chest wall starts in adolescence or young adult life.[8–10] Occasionally, the lesions may be congenital or have a late onset,[11–15] or develop during

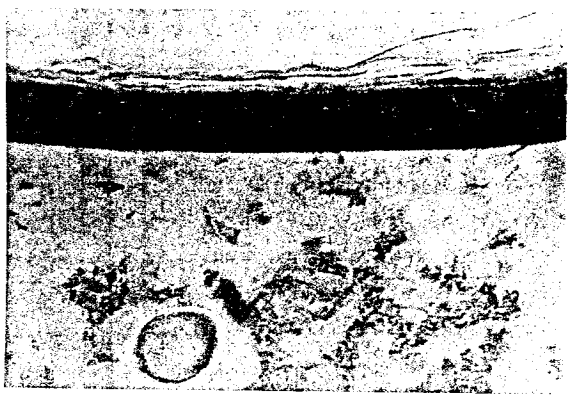

Figure 3. Histologically, the cyst wall is composed of between four and six stratified epithelial layers including a small granular layer. The cyst lumen contains keratinous material and a vellus hair shaft (haematoxylin and eosin × 600).

Figure 4. Within the cyst wall a single sebocyte protrudes into the lumen. In addition, the granular layer of the cystic epithelium is demonstrated (haematoxylin and eosin × 600).

childhood.[8,10,12] The sex distribution is equal.[2,8] Eruptive vellus hair cysts, as well as affecting the anterior chest, can occasionally affect the face or extremities.[16] Histologically, eruptive vellus hair cysts are identical to

Figure 5. Aggregates of sebocysts within the cyst wall (haematoxylin and eosin × 1000).

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singular vellus hair cysts, which represent a hamartoma of the follicular duct. Singular vellus hair cysts develop spontaneously and do not seem to be genetically determined.

In steatocystoma multiplex, multiple cutaneous cysts develop on the trunk.[7] Histologically these are identical to those of eruptive vellus hair cysts, and the mode in inheritance is similarly an autosomal dominant one.[2] Likewise, the onset is mainly in adolescence or early adulthood and, sometimes, in addition to the chest wall, the face or extremities can be involved. Sometimes steatocystomas can appear singly (steatocystoma simplex).

Despite the similarities there are distinct histological changes which allow differentiation of the two disorders. The histological diagnosis of eruptive vellus hair cysts requires the demonstration of cystic proliferation with a stratified epithelium of two to six layers. The granular layer leads to the accumulation of keratinous material beside the multiple and fragmented vellus hair shafts within the cyst lumen. The pattern of differentiation resembles that of the infundibular duct. Steatocystomas are characterized histologically by a stratified squamous epithelium without a granular layer. Instead, there is a constant hyaline lining similar to that of the sebaceous duct. Large sebaceous glands or single sebocytes, located within the cyst wall, are diagnostic for steatocystoma. Occasionally fragmented vellus hair shafts are seen within the cyst lumen.[17,18]

Some authors have described the appearance of steatocystoma multiplex and eruptive vellus hair cysts in one patient,[4–6,19] but there are few reports of overlapping histological features within one specimen.[3,6,20] One report describes cysts with the histological features of eruptive vellus hair cysts but with additional sebaceous glands within the cyst wall.[3] The findings were interpreted as a hybrid cyst of eruptive vellus hair cyst and steatocystoma multiplex.[3] In contrast, a patient with eruptive vellus hair cysts has been reported in whom an eosinophilic hyaline lining membrane was present instead of a granular layer, and in whom sebaceous elements were absent.[6] Our patient shows overlapping histological features of both eruptive vellus hair cysts and steatocystoma multiplex (Figs 5 and 6),[3] and we feel that this suggests that the two conditions are variants of one entity. These hamartomas seem to originate from the isthmus and pilosebaceous duct junction of the hair follicle. The resulting cystic tumours may develop with a predominantly follicular differentiation, as eruptive vellus hair cysts, or with differentiation and imitation of the sebaceous duct, as in steatocystoma multiplex.[17] The similarities of eruptive vellus hair cysts and steatocystoma multiplex have led to the two conditions being considered as multiple pilosebaceous cysts.[5] Our patient demonstrated the classical histological features of eruptive vellus hair cysts in the first biopsy, with overlap features in the second biopsy. We suspect that, if step sections are taken on multiple biopsies, more overlapping histological changes will be seen in patients with eruptive vellus hair cysts and steatocystoma multiplex.

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