Genetical Studies on Skin Diseases

IX. Multiple Follicularcyste (Bosselini)

By

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"Multiple Follicularcyste" in Japan was reported first by Komaya1) (1922), and then the detailed reports by Hashimoto,2) Mukai,3) Minami4) and other authors followed. Siemens,5) Ito,6) Mukai and Noojin-Reynolds7) called attention to the inheritance of this disorder, but the lack of sufficient details of the family history makes this problem remain still unsolved.

I examined a case of this disease and studied statistically from genetical point the cases collected from Japanese literature.

Case Report

Mr. N. F. aged 25 came to our clinic in Aug. 1951. Healthy appearance and no visible anomaly of development excepting color-weakness. His father died of lymphosarcoma, and 6 members in his family had the same skin disease (Fig. 1). Color-weakness was seen in both one of his brothers and his cousin. He could not notice the beginning of his disorder. Since he had noticed the presence of these nodules, they became gradually enlarged and remained in various sizes. The nodules were distributed on the front, hair-covered head surface, neck, back, groin, thighs and arms. No nodules on the scrotum, ears and periauricular regions. The number of these nodules was 500 and more in total. They adhered to the overlying skin but not to the subcutis. They were uncolored, except a few ones with a slight-bluish tone, tumorlike nodules varying in size from pin-head to sparrow-egg. The nodules on the arms and thighs were so small that they could be noticed only by palpation, but they were larger in size on the neck, back, chest and groin. Most of them were palpable in elastic firm, but on larger nodules was pseudo-fluctuation noticed and cream- or butterlike fluid escaped along the needle by puncture.

Cholesterol in blood was 272 mg/dl, other blood examinations pre-
sented no abnormal results.

Histological Findings; Epidermis normal. Large cyst which contains sudanophilic homogene substance is distributed from cuits to subcutis. The wall of cyst consists of 1–3 layers of epithelial cells and connective tissue. On the innermost of cyst wall is seen keratin lamellae. Atrophic sebaceous gland cells are found in incorporation to a part of the cyst wall. Follicle mouth is usually dilated and keratotic plug seen there. There are cellular infiltrations around cyst wall which consist of epitheloid cells, round cells and giantcell-like cells. Outside of these cells, perivascular round cell infiltration is slightly found. The figure of lipophagy in fatty tissue also can be found.

COMMENT

This disorder was variously named by different authors from their points of view: multiple Follicularcyste (Bosselini,⁸ 1898), steatocystoma multiplex (Pringle,⁹ 1899), Sebocystomatosis (Günther,¹⁰ 1917), multiple sebaceous cysts (McPhedrom,¹¹ 1905), maladie polikystique épidermique héréditaires (Sézary et Levy-Coblenz,¹² 1931), Nevo-cystico-tricosebceo diffuso (Lisi, F.,¹³ 1932), etc.

Thirty-three cases including the present case were collected from Japanese literature and studied statistically from the genetical view point. Sex: 4 females and 28 males. This shows that this disease is seen more frequently in males as Mount,¹⁴ Lisi, etc. had observed. Most of the patients become first aware of their disease in their age of 15–25 years (Table I). The cases with no statement of family history are 11, and in 7 out of 22 cases with family history, is the family occurrence of this disorder given: in 1 case it occurred through 3 generations, in 5 cases
through 2 generations and in 1 case in two brothers.

About pathogenesis, Bossellini, Pringle, Komaya and many other authors gave a role to hyperkeratosis in follicle, and Schaumann\(^{15}\) added to this the anatomic malformation of sebaceous gland mouth. Moreover, Noodjin-Reynolds suggested the role of transmitted lipid metabolic abnormality.

In my case as in Noodjin-Reynolds’ case, cholesterol in blood showed abnormally high contents. It shows that the patients had lipid metabolic abnormality. From this it is suggested that this disorder is connected with lipid metabolic abnormality. The fact that lipid metabolism is most active in puberty, when the appearance of this disease is most frequently observed, is favorable to this assumption. It seems to be logical to assume that the cyst formation is due to the transmitted malformation of sebaceous gland mouth, accelerated by both lipid metabolic abnormality and hyperkeratosis. It is very natural that the hyperkeratosis and malformations of the sebaceous gland mouth are found in various degrees. Therefore the lesions can present different appearances varying from apparent nodules to the ones which are so slightly affected that it is impossible to diagnose them as this disease. In family history, the latter ones cannot be recognized as this disease. From the above, it is considered that this disease is inherited in dominant type containing so-called latent case in the abovementioned sense, though Siemens describes that it presents “indefinite” dominante type and Noodjin-Reynolds writes “responsible single” dominant.

In histological findings, cell infiltration around cyst-wall was usually reported, and Schaumann, Lisi, Yamazaki\(^{16}\) and Mukai found giant cells. Yamazaki reported remarked cell infiltration around atrophic hair-root attended to the cyst. Comparatively remarkable cell infiltration was seen also in my case. But similar histological finding with giant cells is seen in epithelioma adenoides cysticum, which is the disease with similar cyst based on the congenital disposition (Ito\(^{17}\), Yamazaki\(^{18}\), Maschkilljsson\(^{19}\), etc.). Therefore, this finding seems to be based on the effect of the cyst and not to work as etiological factor.

No complication was reported in 7 cases out of 10, and in 23 cases is given no statement of complication. Only 3 cases presented some complications: lipoma and nevus molis (Yamazaki), nail anomaly (Tani-guchi-Sozu\(^{20}\)) and color-weakness by the present author.

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<th>Age years</th>
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CONCLUSION

A case of "multiple Follicularcyste" was reported. Six patients were found to have had this disease through 3 generations. A genetical study of the 33 cases collected from Japanese literature was made, and the author concluded that this disease is inherited in dominant type.

References

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