A CASE OF WOOLLY HAIR NAEVUS

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This rare anomaly of the hair was first described by Wise (1927) and since then has received little attention in dermatological textbooks and literature. Wise reported on 'a peculiar form of birth mark of the hair of the scalp, hitherto undescribed, with a report of two cases'. He named the condition woolly hair naevus.

The disorder occurred in two unrelated 5-year-old girls, both of whom had light woolly hair on one portion of the scalp while the rest of the hair was straight and brown. One had a linear naevus of the skin of the back of the neck, right elbow and wrist, on the same side as the woolly hair on the scalp. In advancing his reason for the designation given to the condition, Wise stated that birthmarks of the hair are changes in the 'colour, consistency, structure and morphology of the hair shaft'.

Wise and Sulzberger (1932) described a similar scalp anomaly in two cases of 'acquired progressive kinking of the scalp hair accompanied by changes in its pigmentation'. The patients were adult males with kinky black hair growing over the frontal, temporal and parietal areas of the scalp which progressed rapidly when it developed. The condition appeared in adult life and the hair became darker in contrast to Wise's earlier cases which were children and in which the hair was of a lighter colour than that of the remainder of the scalp.

There was a lapse of 16 years before any further reports appeared in the literature. Anderson (1943) presented a case to the Los Angeles Dermatological Society. The patient was a 5-year-old girl and the lesion had been present since birth and had been gradually increasing in size. The naevus was on the left side of the scalp and consisted of an irregular, oval area in which the hair was lighter and closely kinked. The scalp in the affected area was of a café au lait colour which extended down in a streak on the left side of the neck. In the discussion on the case it was stated that the condition was apparently a naevus of an unusual type, showing woolly hair quite different from the hair on the other parts of the scalp.

Sweitzer (1948) presented a case to the Minnesota Dermatological Society which he called woolly hair naevus. The case was that of a 4-year-old girl who had a scanty growth of hair with a fine kinky texture. In this case the hair had been shed at the age of 5½ months and replaced by a scanty down; there had been no further growth since the child was 2 years old. There was no evidence of any inflammation of the scalp nor was there any evidence of naevi elsewhere. In the discussion of this case it was stated that in the original cases the condition was characterized by localized areas of woolly hair among normal hair, while in this case it was a generalized condition. There was a suggestion of an ectodermal defect in the quality of hair and, therefore, it did not seem that this was a genuine example of woolly hair naevus.

Since these reports four other cases have been described. Hoffmann (1953) described a case in which there was a considerable amount of seborrhoea of the woolly hair portion of the scalp. This case, first seen at the age of 4 years in 1917, had areas of bright, thin, spiral crinkled hair. The microscopic appearance of the affected hair showed it to be thinner than normal, more flat and less pigmented. The medullary part was more or less missing. The skin of the affected areas was browner than the skin of the remainder of the scalp and its surface was somewhat rough. Nine years after first being seen the condition was unchanged but it appeared that the affected hairs grew more slowly than the normal hairs. Hoffmann (1953) called the condition crinkly hair naevus (*naevus ulotrichicus capillitii*) and stated that differentiation must be made between crinkly hair naevus and negro hair and familial curly hair which involves the whole scalp.

Knierer (1955) described a case which resembled the condition reported by Wise and Sulzberger (1932); the patient was an adult and the hair in the affected areas was darker than elsewhere. Born (1957) and Post (1958) also reported cases which resembled those originally described by Wise.

In Post's patient, a 7-year-old boy, the woolly areas were accompanied by linear verrucous naevi on the face, neck and arm. In none of the reported cases has there been a family history of a similar abnormality.

Until now it appears that this condition has not been described in British literature and the present case is the first to be reported upon in Britain so far as is known.

Case Report

A boy, aged 6 years, was seen at the out-patient department; for about four years his hair had shown a patchy fuzziness, which was now thought to be becoming more obvious. The boy's mother stated that he had been born with no hair on the scalp and it was not until he was 2 years old that he began to develop scalp hair, which was noticed to have a peculiar patchy and fuzzy appearance, the fuzzy areas being of a lighter colour than the rest of the scalp hair. The first area to be noticed was over the back of the head and, thereafter, further areas had appeared.

During the past four years the areas had extended slightly, had remained fuzzy, and had been persistently lighter in colour than the unaffected areas. At no time was it noticed that the abnormal hairs were deficient in growth nor did they appear to be any weaker than those of the rest of the scalp.

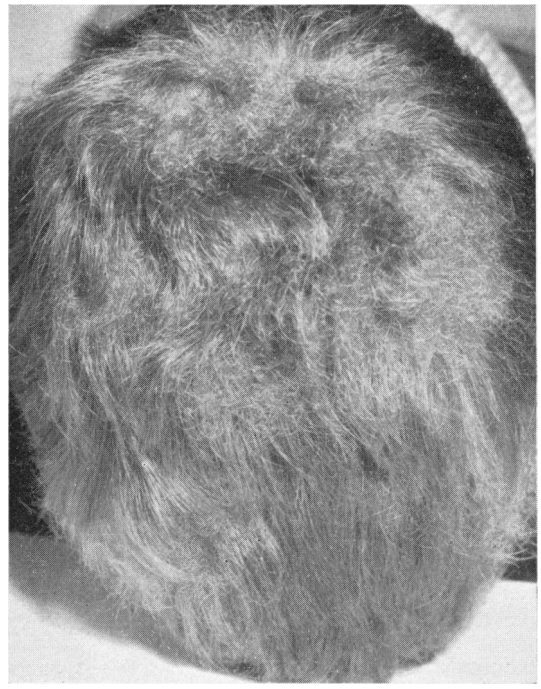

FIG. 1.—Woolly hair naevus.

Family History. The patient's parents were alive and well and had no hair defects or other abnormalities. The patient was the younger of two children, the elder brother being healthy with no congenital defects. The patient was a normal full-time spontaneous delivery. His maternal uncle was said to have had some defect of his hair but this was thought to have been a generalized condition. No examination could be carried out as the uncle had been in Canada for five years and had apparently now lost most of his hair. It was not considered that he had had a woolly hair naevus.

Past History. The patient's general health was excellent. He had measles at the age of 4 years and had been known to suffer from myopia for about one year, having worn glasses since that time.

Examination. The general appearance was that of a normal healthy boy, with no abnormal findings on general examination. Examination of the scalp showed normal distribution of hair with no obvious deficiency of growth. There was no seborrhoea or other scalp abnormality. The most striking feature was the presence of areas of fuzzy, lustreless hair which were noticeably lighter in colour than the rest of the scalp hair. The normal hair was dark brown and the affected hair was a much lighter brown. The involved areas were over the occipito-parietal regions on both sides, the temporo-parietal regions on the right and left and the frontal aspects of the scalp. These areas were separated by the much darker areas of straight normal hair (Fig. 1) and the affected hairs, despite their appearance, were of the same texture as the rest of the scalp and showed no increased fragility. The eyebrows were of normal appearance and there was no abnormality of the finger and toe nails or the teeth. Examination of the eyes revealed a web-like opacity over the pupil of the right eye (Fig. 2).

He was referred to the ophthalmologist whose report was as follows: 'The patient has a very extensive moderately dense membrane covering the pupillary area of the right eye. It arises from the lesser circle and, as can be readily seen, it allows free movement of the pupil. There are fine openings in the membrane and it is un-

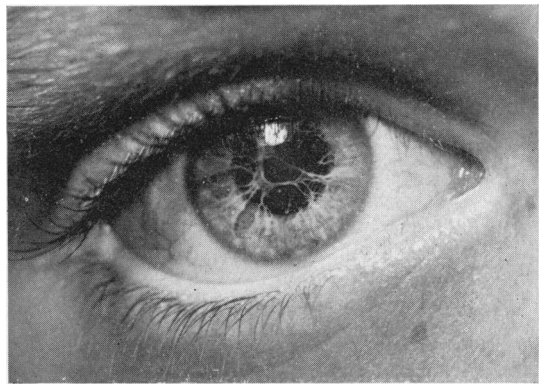

FIG. 2.—Persistent pupillary membrane.

likely that the vision in the eye is affected by it. No treatment is recommended.'

Microscopic examination of hairs from the affected and normal areas revealed no abnormality of the hairs. There was no evidence of monilethrix, trichorrhexis nodosa, pili torti, or other hair defects. The affected hairs were slightly curly and they were rather less pigmented than the normal straight ones.

Discussion

The case described is, in the writer's opinion, an example of the condition described by Wise as woolly hair naevus. There is, in common with all previously reported cases, no family history of similar naevoid changes in the scalp hair.

The diagnosis should be straightforward once the condition has been recognized. The differentiating points in distinguishing it from the condition described by Wise and Sulzberger (1932) are that it is not an acquired anomaly; it occurs in children; in all cases the affected areas are lighter in colour than the normal hair; it is not a generalized condition but a patchy one; the affected areas are interspersed by normal and darker hairs; it does not spread rapidly when it develops; and there is no microscopic abnormality of the affected hairs. Other hair abnormalities can be ruled out by the fact that in most of them there is a deficiency in hair growth and an increased fragility of the hairs which is not apparent in woolly hair naevus. Microscopic examination of the hairs will decide the diagnosis as other defects have a characteristic appearance.

Post (1958) suggested that there are three types of woolly hair naevus: (1) a type without any visible changes of the glabrous skin or scalp, (2) a type accompanied by linear naevi of the skin and (3) a type acquired in adult life in which the hair of the scalp assumes the characteristics of the pubic hair. While accepting the first two groups, the case under discussion being an example of the first, the writer feels that the third group should not be classified as a woolly hair naevus but should be classified as acquired progressive kinking of the hair as described by Wise and Sulzberger (1932).

The persistent pupillary membrane is not so rare a condition as it was once thought to be and has been reported as being found in 506 out of 1,600 eyes. It is most common in infants and children, and rare in old age as it tends to be absorbed with advancing years. Pupillary membrane remnants have the essential characteristic that if they retain contact with the iris they are always attached to the superficial mesodermal layer on its anterior surface usually at the lesser circle or occasionally more peripherally. In all cases they are extensile and do not impede movements of the pupil or interfere with vision unless they are very dense. As regards the present case, it is felt that the presence of a mesodermal defect associated with an ectodermal one is purely coincidental and of no special significance.

Summary

The literature on woolly hair naevus is briefly reviewed and a case is described which had no associated skin or scalp changes. The patient had a persistent pupillary membrane. As far as is known, no similar case has previously been reported in Britain and this is only the seventh example so far described in world literature.

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