

BLACK PIEDRA IN A CHILD WITH PILI TORTI, BAMBOO HAIR AND CONGENITAL ICHTHYOSIFORM ERYTHRODERMA

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Piedra (Spanish: stone) is a chronic fungous infection of the hair characterized by the presence of tiny, adherent, gritty, brown or black nodules dotted along the supra-follicular part of the hair shaft. The nodules may be microscopic in size or may be visible and palpable, and a hair may bear one or several dispersed irregularly along its length. Piedra has no effect on hair growth and does not, in humans, cause breakage or fraying; it causes no symptoms apart from a sandy feeling when the hair is drawn through the fingers. The disease has also been described under such terms as trichosporosis, trichomycosis nodularis, tinea nodosa and many others.

Two varieties of piedra occur, white and black, the colours being those of the colonies of fungi in culture, not of the nodules on the hair. Although many cultural variants of the causative fungi have been described in the past, it is now generally accepted that only two are, in fact, involved, viz. *Trichosporon beigeli* in white piedra and *Piedraia hortai* in black piedra.

White piedra was first described in 1865 in London by Beigel, according to the *Nouvelle Pratique Dermatologique*.¹ Beigel found the nodules not only on growing hair but also on hair used in false chignons. The light-brown nodules of white piedra are found on beard and moustache hair and sometimes on scalp hair, and infection of the perineal hair has been described. White piedra occurs in South America, England, Europe and Japan, and odd cases have been found in North America, India, Algeria and Nigeria.

Black piedra was probably first isolated in 1876 in Colombia by Ozorio and Arango, and the fungus was cultured by Desenne in 1878, but the final sorting out of the black and white varieties was the work of Horta in 1911. Only scalp hair is affected by black piedra, and the nodules are dark brown or black. Black piedra is endemic in South America, Indonesia and Cochin-China—all tropical areas with a very high rainfall. In the lush climate of some parts of South America even overhead telephone wires get a kind of piedra caused by *Tillandsia*. Moisture other than humidity may predispose to the infection which is commoner in swimmers than in non-swimmers.

The literature does not mention the possibility of spontaneous cure in piedra, and black piedra may apparently persist for years after the host has returned from a tropical to a temperate climate. Both types of piedra are susceptible to simple local treatment with shampooing and the application of fungicides such as a 1:2000 solution of bichloride of mercury or ammoniated mercury ointment.

Piedra is unrelated to trichomycosis axillaris (leptothrix), a fungous infection of the axillary and, sometimes, the pubic hairs, caused by *Nocardia tenuis*. This common condition is found all over the world and is characterized by soft, yellow, red or black concretions around the hair shafts. Other conditions that might be confused with piedra are monilethrix, trichorrhexis nodosa, bamboo hair, and nits, but all are easily excluded by microscopy.

In none of the standard works on mycology (e.g. Brumpt² Conant *et al.*,³ Langeron and Vanbreuseghem,⁴ and Simons⁵), is there any mention of piedra occurring in man in Africa. We have, however, found two articles referring to white piedra in Nigeria⁶ and Algeria,⁷ and Loewenthal⁸ informs us that he has seen 1 case of what seemed to be black piedra in Uganda. Neither variety of piedra has hitherto been described as occurring in Southern Africa.

Piedra occurs in animals as well as in man. Lochte,⁹ in Munich in 1937, found piedra on the hairs of 5 out of 7 chimpanzee pelts from the Cameroons, and recently Kaplan¹⁰ has made a considerable study of the infection in primate pelts collected in the American Museum of Natural History in New York. Piedra was found in 57 out of 94 (60.6%) pelts from Asia; in 83 out of 109 (78.1%) from the New World; and in 58 out of 202 (28.7%) from Africa. Only 2 cases were found in 33 primates in zoos in the United States. Since the pelts had been treated for preservation, it was impossible to identify the fungus by culture, but the microscopical findings suggested that black piedra was involved. In contradistinction to piedra of human hair, that in animals causes severe pilar damage with the possibility of breakage at the nodule. With the exception of the primitive lorroids, skins from primates of nearly all the major divisions of this zoological group were found to have piedra to a varying extent.

The rarity of black piedra in humans in Africa is astonishing in view of the enormous reservoir of infection in nature.

CASE HISTORY

The patient, a white girl aged 4 years, is the fourth child of healthy parents who are not blood relations. The first 2 children in the family are healthy, but the third, a boy born in 1954, showed signs of skin disease at birth and died in his fourth week.

The patient also developed a generalized erythema on the day of birth, in 1956, and was referred by Dr. G. de V. Theron of Worcester, Cape, to the Karl Bremer Hospital, Bellville, where she was under the care of Dr. W. H. Opie in the Children's Department. She was first seen by one of us (J.M.) when she was about a week old. The skin was

generally erythematous and there was a typical widespread moniliasis of the skin and the buccal mucosa. Her general condition caused no concern. The monilial infection cleared slowly on treatment with 'mycostatin' and the last remnant disappeared only when she was about 9 months old. At this time the skin remained a little pink.

She was not seen again until 1960. In the interval the skin had become red, scaly and itchy, especially in the flexures. The hair seemed normal at birth but it had soon fallen out in patches until the scalp was denuded; the scalp had always shown fine superficial scaling. Until the age of 2 years there was little scalp hair, but thereafter she had grown a short stubble over the back of the head and rather longer hair in front.

In 1960 she was small for her age, but active and intelligent. The skin was generally a little erythematous and scaly. The erythema was said to vary in intensity and to be occasionally blotchy. Scaling was most marked in the flexures of the elbows and knees, and there was a fine desquamation of the scalp. Follicular keratosis was not noted. On the limbs, particularly the legs, there were rings of superficial scaling, some showing concentric figures; a few such lesions were seen on the trunk (Fig. 1). These rings came and went continually. There was superficial peeling of the fingers and toes, but no palmo-plantar hyperkeratosis. No vesicular or bullous lesions were seen. Secondary coccal infection was obvious about the ears and in the natal cleft, and there was a chronic blepharitis. The teeth and nails were normal. There were no defects in vision, hearing or intellect, and sweating appeared normal. The skin state was one of congenital ichthyosiform erythroderma.

From a description given by Dr. G. de V. Theron it seems likely that the brother who died suffered from the same genodermatosis. The other members of the family show no abnormalities of the skin or epidermal appendages.

The patient's scalp hair was short, coarse, dry, dull and sparse; at the back of the head the hairs were about 1 cm. in length, in front about 5 cm. (Fig. 2). Tiny nodular swellings were visible on most of the short hairs and on some of the longer hairs. The eyebrow hairs and eyelashes were scanty and bore nodules. Body hair was almost non-existent.

Naked-eye examination suggested piedra and hairs were taken for mycological studies, but microscopic examination showed that although some of the nodules were piedra concretions most were bamboo hair swellings, and that yet another abnormality, pili torti, was also present.

Mycology

Some of the hairs received for fungus studies showed small black, hard nodules containing dark, branching hyphae resembling arthrospores and small asci with several fusiform ascospores. The hair shafts themselves were not invaded.

Material was planted on Littman's oxgall agar and on Sabouraud's agar. Most of the nodules gave no growth, but a few produced slow-growing, greenish-black, wrinkled colonies that were fully developed after 22 days. Growth was better on Sabouraud's than on Littman's medium (Figs. 3-6).

The piedra concretion is decomposed by treatment with 10% potassium hydroxide for 10-15 minutes. It is then obvious that the nodule is composed of closely septate, branched hyphae, 4-8 μ in diameter, held together by a cement-like substance. In this case the fungus was clearly ectothrix. Both microscopically and culturally the fungus grown was identical with that described as *Piedraia hortai*.

A fortnight after the child had finished a month's course of griseofulvin (0.5 g. daily), hairs were again planted on agar and a growth of *P. hortai* was obtained. The patient's father, mother, brother and sister showed no clinical or microscopical evidence of piedra, but hairs were planted on agar and in the case of the sister *P. hortai* was cultured on one occasion.



Fig. 1. Back of leg showing intertrigo and circinate scaling.

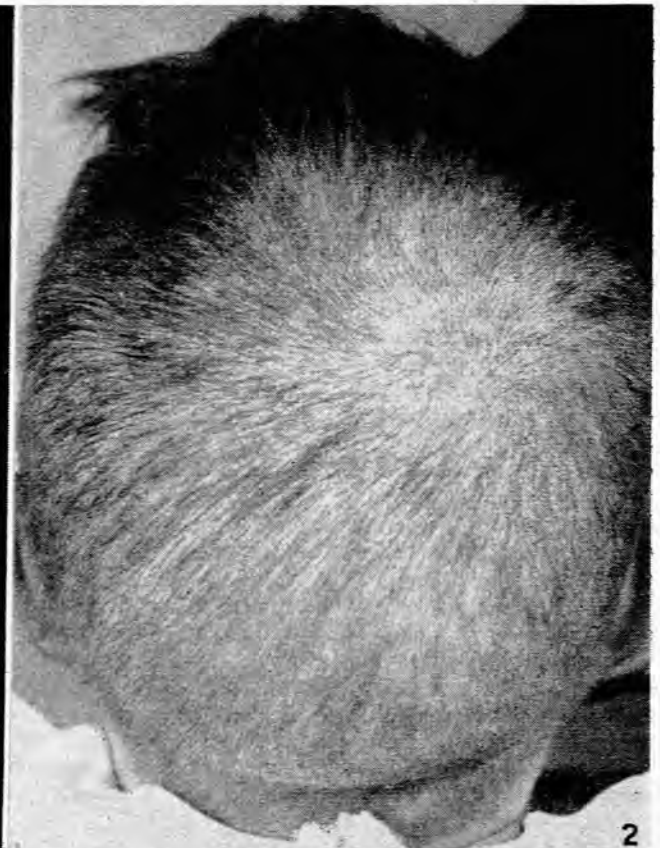


Fig. 2. Stubby hair on scalp.

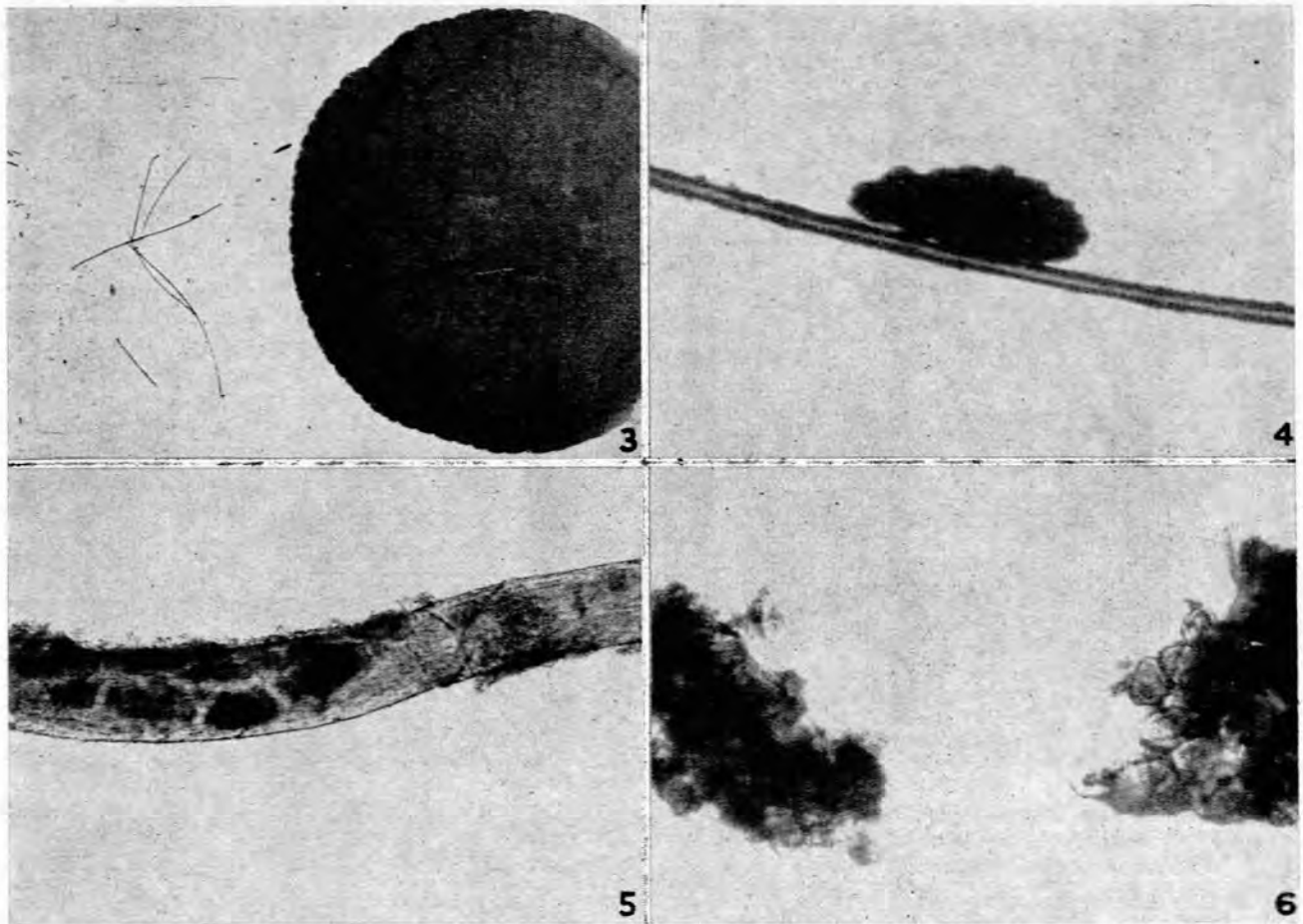


Fig. 3. *Piedraia hortai* growing on Sabouraud's agar (24th day). One of the hairs to the left of the colony bears a piedra nodule.

Fig. 5. Invasion of cuticle revealed by removal of piedra nodule.

Fig. 4. Piedra nodule attached to hair shaft.

Fig. 6. Ascospores in piedra nodule after maceration in potassium hydroxide.

DISCUSSION

Bamboo Hair

Bamboo hair was the excellent term chosen by Netherton¹¹ to describe a unique variety of trichorrhexis nodosa that he found in a girl with congenital ichthyosiform erythroderma. The nodules were found, in his case, on scalp hair, eyebrows, eyelashes and body hair, and the changes he describes are precisely the same as those in our patient's hair. In Netherton's case there was no torsion of the hair shafts deserving the description of pili torti, and cultures for fungi were sterile.

The nodular swellings, which are clearly visible under a hand lens, are dotted irregularly along the hair shaft and may be single or multiple. The abnormality is most frequently found on the shortest hairs, but is sometimes found on longer hairs. There is great variation in the diameter of the hairs. The largest nodules consist of a cup-shaped swelling or socket on the proximal part of the hair shaft into which the distal shaft is inserted, giving an appearance reminiscent of an impacted fracture, and a hair dotted with nodules looks just like a bamboo rod. Breakage of hair occurs at such nodules and most often

leaves a clean socket on the proximal shaft; occasionally a socket is left with a splinter of the distal shaft still emerging from it (Figs. 7 and 8). The least degree of abnormality in the hairs is manifested by ainum-like constrictions of the shaft, and all grades of abnormality between simple sulcation and the final socket formations can be found (Fig. 9).

Many hairs show medullary degeneration manifested by collections of black granules. These show on the illustrations only as dark central masses. Netherton found a dense concentration of these granules in the socket formations, but this was not a notable feature in our case. We agree with Netherton that bamboo hair is a congenital defect and not due to any infective process in the hair shaft. Apart from Netherton's original case and ours only one other has been found. Netherton¹² informs us that Dr. G. Curtis, in the USA, has found a girl with bamboo hair and a history identical with that in his own case.

The defect in bamboo hair is entirely different from that in classical trichorrhexis nodosa. In this fairly common condition the nodules on the hair shafts are caused by longitudinal splitting and the appearance was compared by Sabouraud¹³ to that seen in 'a piece of wicker

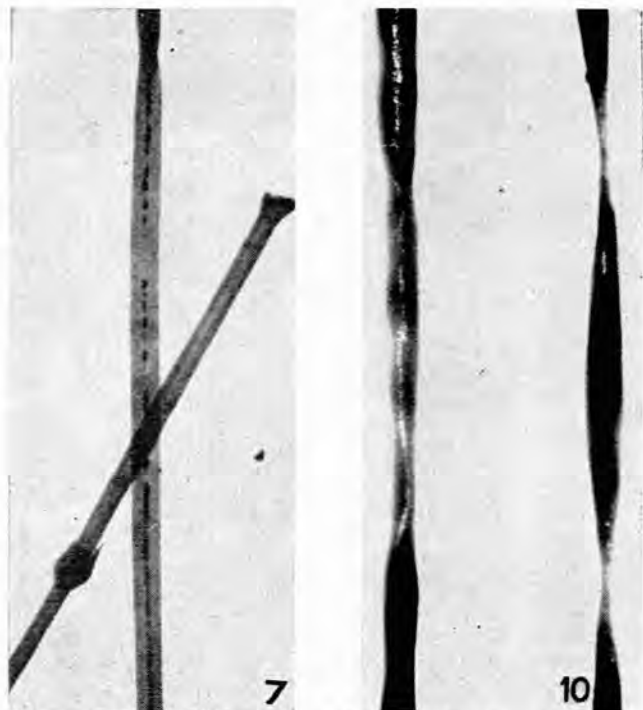


Fig. 7. Bamboo hair. Typical fully developed nodule and terminal socket. The other hair shows medullary degeneration and torsion.

Fig. 10. Pili torti. The irregular twisting of pili torti contrasted with the regular beading of monilethrix (on the right).

that has been bent back and forth a hundred times at the same point', and when a hair fractures the broken ends look like brooms. Trichorrhexis nodosa is usually caused by chemical or physical trauma to the hair (e.g. too frequent shampooing, bleaches, violent brushing), but Touraine¹⁴ mentions some families in which the tendency is inherited in dominance.

Netherton¹¹ entitled his article 'A unique case of trichorrhexis nodosa — "Bamboo hairs"'. In order to avoid confusing 2 disparate conditions we believe bamboo hair should be so described *tout court*, and trichorrhexis nodosa reserved for the commoner state that has for long borne this name.

Pili torti

Pili torti (trichokinesis, trichotortosis, twisted hair, woolly hair, gedrehte Haare) was another defect found in our case. In this condition the hair is flattened and presents from 3 to 10 torsions where the shaft twists through 180° on its axis. Danforth¹⁵ noted that normal hair is frequently ribbon-shaped rather than cylindrical and that torsion may occur, but in pili torti there are usually many twists in a given hair, and the hairs break easily. In our case most of the scalp and eyebrow hairs examined showed multiple twists, and many hairs had both twists and bamboo nodules.

Pili torti has often occurred in families and is then inherited as a dominant trait. Among other abnormalities which have been found in patients with pili torti are milia of the face, keratosis follicularis and classical

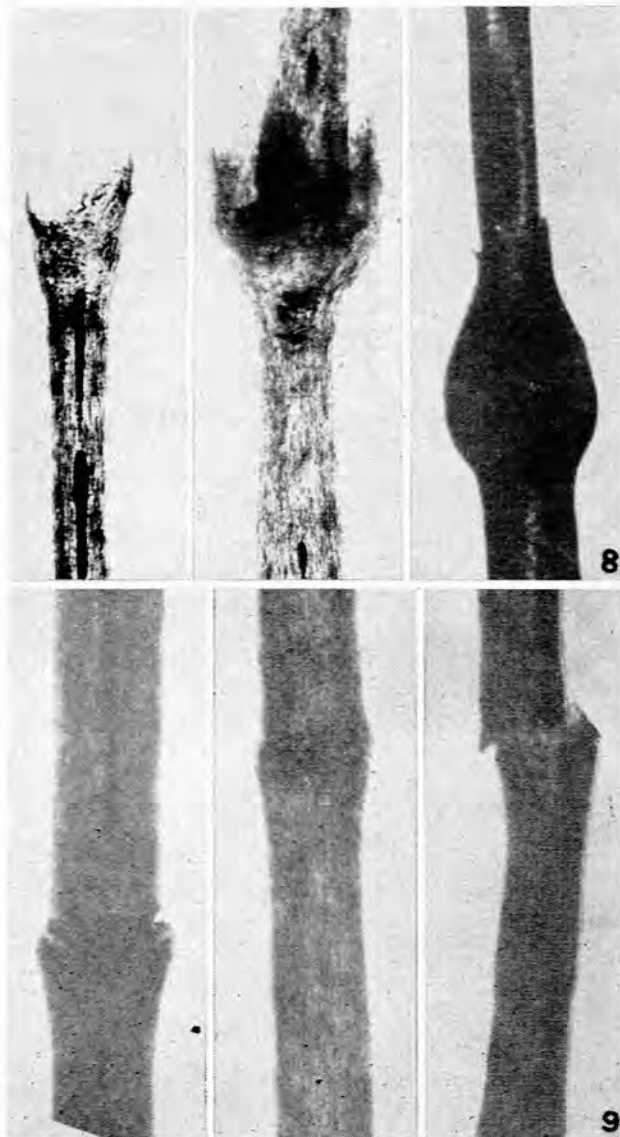


Fig. 8. Bamboo hair. Fully developed nodules and terminal socket.

Fig. 9. Bamboo hair. Minor changes.

trichorrhexis nodosa; ichthyosis has occurred in other members of the family of a sufferer.¹⁴ In a case described by Björnstad¹⁶ the twisted hairs were fluted with longitudinal ridges and fissures. A localized growth of twisted hair followed an infection of the scalp in a case reported by Scott.¹⁷ Children with pili torti are often born with little or no scalp hair, and the abnormality may be recognized only after a year or two when a short stubble grows. In the majority of cases the hair on the occipital scalp is most affected. Twisted hairs are less elastic than normal and break easily with lengthwise splitting (trichoptilosis) leaving a broom-like end as in trichorrhexis nodosa. Breakage only rarely takes place at the first twist above the follicle. Twisting does not occur at regular intervals so that there is little likelihood of confusing pili torti with

the regularly beaded hairs of monilethrix (Fig. 10). The alternating planes give the hair a metallic glint. The eyebrows may be affected and are sparse or entirely absent, and sometimes the eyelashes as well. The scalp hair stays short in childhood, but by the age of puberty some patients produce enough normal hairs to give a presentable appearance and mask the abnormal hairs that remain.

In investigating suspect cases of pili torti it is important to examine dry hairs since the twists are almost unrecognizable in mounted specimens.

SUMMARY

Piedraia hortai was cultured from piedra nodules on the scalp hair of a child with congenital ichthyosiform erythroderma, bamboo hair and pili torti.

Although there is an enormous reservoir of what appears to be black piedra in the primates of Africa, this is the first time that the disease has been positively identified in man on this continent.

The clinical photographs were taken by Mr. Robert Ellis, Department of Clinical Photography, Karl Bremer Hospital, and the photomicrographs by Mr. B. Neiteler, Department of Microbiology (piedra) and Mr. C. van Schaik, Department of Gynaecology (bamboo hair).

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