

**Three case of xeroderma pigmentosum (Kaposi) or atrophoderma pigmentosum / by H. Radcliffe Crocker.**

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THREE CASES

OF

XERODERMA PIGMENTOSUM

(KAPOSI)

OR

ATROPHODERMA PIGMENTOSUM.

BY

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Read March 25th, 1884.

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Received January 8th, 1884.—Read March 25th, 1884.

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THE disease of which I show examples to-night was first described by Kaposi in 1870, from four cases in Hebra's practice. Since then cases have been recorded in Germany, France, and America, but those now shown are the only cases known to have occurred in England, and were exhibited about a year ago at one of the Societies as examples of lupus. They, therefore, did not attract the attention they deserved and no account of them was published. I am indebted to Mr. Martin, a student of University College Hospital, for having them sent up to me.

The children are the three eldest of a family of four; the father, æt. 42, is a Shropshire farmer, and has always had good health, the mother, æt. 40, was in good health at the time of their birth and suckled them all. The last child, a girl, æt. 6 years, is perfectly healthy, and is described as having a "beautiful skin." This child



was not suckled. There is no history of cancer or any other hereditary disease in the family on either side.

CASE 1.—Alice Elizabeth B—, æt. 12, was admitted with her brother and sister into University College Hospital on December 10th, 1883.<sup>1</sup>

She was quite healthy until she was between twelve and eighteen months old, when the disease commenced without any previous symptoms, with freckle-like pigment spots which appeared simultaneously upon the face, neck, back of forearms, hands, upper arms, and legs below the knees over the same areas that are now affected, and though the spots have increased in number and other lesions have appeared, the father does not think the disease has spread at all from the commencement. The lesions are thus limited to the uncovered areas, the child having worn short sleeves and socks, and the father thinks the disease began in spring or summer because the spots were thought to be sun-freckles.

Except that the lentiginous spots became more numerous, larger, and some deeper in colour, there was no alteration noticed until she was six years old, when ulcers appeared on the right cheek and nose.

*Present Condition.*—The patient is moderately well-nourished, but less so than her younger sister, does not suffer much pain, and her general health is fairly good.

*Positions of Lesions.*—The disease involves the whole of the face, ears, and neck, terminating on the forehead where the hair begins, in front, but in the temporal region the lesions extend in a minor degree back into the hair as far as a line drawn from the insertion of the ear. The scalp is, however, in great part covered with dirty brown scales of small size.

Below it extends to the level of the second rib in front and to the outer third of the clavicle at the sides and behind to the level of the fourth dorsal vertebra; it does not terminate abruptly, the pigments spots becoming gra-

<sup>1</sup> See Plate III.



dually more sparse below the level of the clavicles. It occupies the whole extensor surfaces of the fingers, hands, forearms, and arms as high as the insertion of the deltoid, but the nails are unaffected. On the flexor aspect the whole palmar surface is quite free, but that of the forearms is thickly covered, but more on the radial than the ulnar side, while it terminates abruptly at the wrist.

The inner aspect of the upper arms is nearly free, the affected areas being mapped out by a line drawn from the lowest point of insertion of the deltoid to each condyle of the humerus. On the legs the disease extends from the tubercle of the tibia, to three inches above the ankle in front, and for a corresponding area on the calf, but laterally, it goes up to the level of the upper border of the patella on the outer side.

The affected areas are quite symmetrical and the skin of all parts of the body not mentioned, is perfectly normal. The red part of the lips and adjacent mucous membrane is white, mottled with red streaks, but the tongue and rest of the oral mucous membrane are free. The right eye has a vascular pterygium extending from the inner angle to the cornea. The disease varies in intensity, being most marked on the face, neck, and extensor surfaces of forearms, while the legs are only slightly affected.

The lesions are multiform, consisting of :

1. Pigmented spots from a pin's head to an inch in diameter, many of the smaller being rounded and like ordinary freckles, but the larger are irregular from aggregation. They vary in colour from a light raw umber to a deep sepia, the large spots being the darkest. They are most abundant on the forearms and neck. Scattered amongst these lentigines are white atrophic spots about an eighth of an inch in diameter, in many parts scarcely noticeable, but in the upper part of the cheeks and in the infraorbital regions aggregated into larger areas of very white skin, very smooth and presenting a cicatricial aspect, or covered with thin layers of readily peeling epithelium. That there is actual contraction of the skin is shown by



the presence of some degree of ectropion of the lower lid, which is denuded of lashes; the mouth also is habitually open, though the lips can be closed. The surface of the skin is very finely wrinkled, especially round the mouth. On pinching up a fold it feels very thin and it is less easily effected than normally, but there is no approach to the difficulty experienced in a case of atrophic scleroderma.

There is marked cicatricial contraction of the nose, the nostrils being rounded and widely dilated, this contraction, however, being partly due to the ulcers, to be presently alluded to, and not only to the atrophy. Telangiectases exist as spots slightly raised above the surface from a pin's point to a third of an inch in size, of a bright crimson colour, and with a lens they can be seen to consist of small dilated vessels; they are not numerous, but conspicuous by their bright colour in the white atrophied skin. More abundant but less noticeable are fainter tufts of dilated capillaries.

Superficial ulcers covered with thick yellowish crusts are abundant upon the nose, and there are a few on the right cheek, but only two on the left. There is one as large as a shilling on the lower lip. When the crusts are removed the surface of the sore is slightly above the surrounding skin in some of them, below it in others. Three tumours are present on the right side of the face. The largest began a year ago on the tragus as "a little black lump;" it grew slowly, preserving its colour, and, when it was about the size of the end of the finger, began to ulcerate, six months from the time of its being first noticed. It is now a flattened spheroidal fungating mass four inches in diameter, projecting an inch and a half above the surface. It covers almost the whole of the right ear and is constantly exuding a sanious fluid. The second tumour, an inch and a half long and one inch wide, extends diagonally from the right malar eminence to the upper lip; it is evidently made up of two tumours fused together. The third is one inch in diameter, on the right side of the chin. These smaller



tumours are covered with black crusts of dried exudation. The tumours are tender, but are not often spontaneously painful. There are no granular enlargements.

More easily felt than seen are numerous small warts springing from the pigment spots; they are most abundant about the face and arms, and some of them closely resemble small pigmented senile warts. Insignificant as these warts appear they are of no slight importance in relation to the tumours.

All the viscera seem healthy, and the functions duly performed. The urine has a sp. gr. 1020, is acid, clear, with no albumen or sugar. Since the case was shown to the Society in January, my colleague, Mr. Marcus Beck, has been kind enough to remove the tumours, together with two glands beneath the large tumour, and a small piece of skin from the outer side of the upper arm. The indolent ulcers also were scraped with a sharp spoon. The large tumour, which was extremely friable and had grown considerably since it was shown to the Society, sprang from a comparatively narrow pedicle, about one inch in diameter, in front of the ear, and did not implicate any of the skin beyond the pedicle, which appeared to spring from the subcutaneous tissue.

After hardening in a fluid consisting of equal parts of a one sixth solution of chromic acid and alcohol, sections were made. The large tumour consisted of masses of papillomatous structure made up of elongated cylinders bordered with oval or cylindrical epithelium, enclosing smaller epithelial cells, with very little fibrous stroma (Pl. V). This structure was imbedded in granulation tissue which constituted the greater part of the tumour; some of the cells of this fibro-cellular tissue were spindle-shaped, and the whole tumour was tunnelled with numerous large blood-vessels. There was nothing approaching to a cancerous structure. The glands beneath were slightly enlarged, showing a slight increase of fibrous tissue; in short, only the appearances one would expect in a somewhat irritated gland. The smaller tumour showed more



epithelial structure. It, too, consisted largely of granulation tissue rich in blood-vessels, but the papilloma masses, instead of being congeries of separate cylinders, form digitate processes from a broader base ; fibrous septa passed up between the lobes, so that it presented a gland-like appearance not unlike a molluscum contagiosum tumour in shape. The extremities of the processes lay free in a cavity in the granulation tissue, and in some places the masses had fallen out, leaving a lacuna in the granulation tissue.

How these gland-like structures are formed can be inferred from the small ulcer in which some of the epithelial surface was left.

In this there were only traces of the horny layer. The rete was increased in thickness, while the inter-papillary processes were enormously elongated by down-growth, and in some places they met and enclosed portions of the papillary layer of the corium. These hypertrophied inter-papillary processes apparently get separated from the rest of the rete, the compressed corium between the lobes forms the fibrous septa, and thus form the probable source of the pseudo-glands seen in the smaller tumour, and by still further disconnection with each other produce the papillomatous structure of the large tumour.

The rete-cells in many instances were vacuolated ; there was leucocytic infiltration between the rete processes and through the corium and granulation tissue beyond it.

The skin showed atrophy of the papillary layer, the rete forming a slightly wavy line over the thinned corium. The horny layer was apparently unaltered, but the lowest layer of the rete was notably pigmented in parts with dark brown granules. At the site of a commencing wart, which was included in the section, the horny layers were much increased in number and separated from each other, and dipped downwards into a corresponding depression of the rete where the cells were flattened. In the centre of the wart the rete sent down an obtusely conical process into the corium ; round this process was a scanty infiltra-



tion of leucocytes apparently proceeding from the vessels connecting the superficial and deep plexuses. There was no other alteration in the corium below the papillary layer.<sup>1</sup>

CASE 2.—Alice Amelia B—, æt. 10, closely resembles her sister, but the disease is less advanced. Her nutrition is not at all affected. The disease began at the same age as Elizabeth's, viz. from twelve to eighteen months, and exactly in the same way and in the same situations, and has followed the same course with one exception. The sores began when she was only about four years old, within a month of their appearance in her elder sister.

The limits of the disease are the same as those of her sister except that in front it reaches only to the level of the first rib and behind to the second dorsal spine. On the forearms it is less marked on the ulnar side of the flexor aspect, does not extend beyond the first proximal phalanges of the fingers, and is much fainter upon the legs. There are two freckles on the right palm. The left leg has extensive superficial scars from a scald in infancy.

The pigment spots are about the same as in her sister, but the atrophic and vascular spots are not so numerous, and there are not such large white areas as in Case 1. The ulcers number about half a dozen on the face, the largest about half an inch across near the outer canthus, and they are nearly all on the right side. There is less cicatricial contraction than in her sister, the lips being perfectly mobile, but there is some left ectropion. The lids are granular, there is conjunctivitis, and great photophobia, but no pterygium. There are some verrucose projections to be felt both on the face and forearms, but no tumours at present.

CASE 3.—Benjamin B—, æt. 9, is most like Case 2, but in some respects more advanced (Pl. VI). Beginning at the same age it followed the same course, but the ulcers

<sup>1</sup> The microscopical observations were confirmed by Mr. Marcus Beck, who concurred in the view that the tumours were papillomatous, and not epitheliomatous.



began when he was only three years old; in short, the ulcers began on all three children within a month of each other. The difference in distribution is, that it extends to the level of the clavicle in front and to the second dorsal vertebra behind, that the ulnar side of the flexor aspect of the forearm is almost free, and that the lesions extend to the end of the fingers. There are pigment spots on two of the nails of the right hand; the tip of the tongue has white spots upon it, where the papillæ are atrophied, so that it has a smooth pale aspect like some syphilitic tongues at that part. There are white spots also upon the lips. The pigment spots are like the others, but there are none more than three quarters of an inch across. There is a white smooth area over the right orbit and side of the nose, but the atrophic spots are inconspicuously scattered about amongst the pigment. Telangiectases are fewer than in the other; more than a dozen scabbed sores exist on the left side of the face, below the forehead, and on the nose, the latter being much distorted from cicatricial contraction. There are only a few small ulcers on the right side of the face. There are no verrucæ on the face, but a few on the arms. There are no tumours now, but there was one on the left cheek which grew out to the extent of an inch and a half in a finger-like way; it did not discharge, but the end became dead and was cut off from time to time, by the patient's mother. The stump was red but not ulcerating, and ultimately the tumour dropped off, leaving a slight scar.

There is a pterygium on the outer canthus of the left eye and on the inner of the right. The boy's general health and nutrition appear good.

Adding these to the already published cases, we have in all thirty-four cases on which we may found a general account of the disease. Taking first the etiological factors, we find that the number of males and females is exactly equal; consanguinity, however, plays an important part. The thirty-four cases are distributed over seventeen families, no less than twenty-six cases having occurred in nine



families. Another striking feature is its tendency to attack one sex only in a family.

This is true in seven instances. In Rüder's series, out of a family of eight boys and five girls, seven boys were attacked, while all the girls and one boy remained free. Nothing in the record of the cases suggests any explanation of this selection, and I do not remember any analogous instance where a disease, which is equally liable to attack both boys and girls, yet habitually selects only one sex in a family. Kaposi and myself alone record exceptions to this.

*Age.*—Nearly all the cases have commenced in the first or second year of life; one was only five months old. There are, however, two cases which did not begin until the age of nine and sixteen years respectively. The disease is obviously due primarily to a congenital predisposition, though, like ichthyosis and prurigo, it does not manifest itself for some time after birth, requiring perhaps some further condition for its development.

Heredity does not appear to have any influence. Three instances of cancer in seventeen families are recorded, but I should hesitate to regard that as of etiological significance.

Hygiene does not seem to have any influence. Most of the patients were in fairly good circumstances, and in no instance have the general surroundings been demonstrably in fault.

*Season.*—In a few instances where the time of year of the onset is mentioned it was in spring or summer, and in some others the time of year may be inferred as the parents have at first regarded the pigment spots as sun-freckles, and both Neisser and Vidal are inclined, therefore, to think that insolation may be an exciting cause in a congenitally predisposed child. These few facts are all we know of the etiology of the disease.

*Symptoms.*—When we review the symptoms and course of the disease we cannot but be impressed by the remarkable similarity of all the cases. Up to the time of the onset of the eruption all the parents state, that nothing



abnormal in the skin had been observable, nor were there any general symptoms to attract attention either before or at the time the disease began. Simply, the common history is that, some morning, probably in spring or summer, the face becomes covered with red spots something like those of measles, for which the disease has been mistaken, or the redness may be more diffused, forming patches. This erythematous eruption fades in a few days, but leaves behind pigmented freckle-like spots, or as in Duhring's, my own, and some other cases, the lentigines have appeared without any preceding eruption. Following at varying intervals of months or years, or it may be simultaneously, the other parts of the body which are habitually uncovered, viz. the neck, face, backs of forearms, and lower parts of arms, and sometimes the legs are similarly affected. When once the disease has attained to the dimensions exemplified by the cases shown to-night it ceases to spread, the further development being in the direction of fresh lesions within the self-prescribed limits. The pigment spots enlarge, become of a deeper colour, and others form, even in such unusual positions as the nails and palms, as in my cases, and the palpebral mucous membrane and sclerotic in one of Vidal's. As time goes on the pigment spots prove themselves by no means the harmless freckles they at first appear, but play an important part in the evolution of what are afterwards to become fatal lesions. We do not know how soon the white atrophic spots and areas develop but they are later than the pigmentation. And whether they are consequent upon the telangiectases, as Taylor thinks, or whether the telangiectases are the consequence of the obliteration of the vessels in the atrophic areas, remains to be proved. The atrophy sometimes involves the mucous membranes. At varying intervals, but usually some years from the commencement of the disease, superficial ulcers develop, covered with yellowish brown, or black blood-crusts. Some of the ulcers heal while others spread and fungate. A little later when the finger is passed over



the skin small verrucose projections can be felt situated upon the pigment spots. These warts are in most cases the starting-points of the tumours. Here I would call attention to a point not noticed before, namely, the prevalence of the ulcers and tumours upon the right side of the face. Out of twelve instances in which the side where these lesions began is mentioned, in nine it was upon the right and in only three upon the left side, and when both sides were involved they usually predominated upon the right.

*Clinically*, the tumours may be described as verrucose and fungating, but pathologically, all observers except Taylor, of New York, have described them as epitheliomas, with the epidermic glands abundant in some but absent in others. That they are not always or at all times epitheliomas the present case demonstrates, and Taylor's was not improbably like mine. It is, however, quite possible that if these tumours were left to irritate and be irritated that epithelioma might supervene, for the boundary dividing papilloma from epithelioma is often a very narrow one. Leloir and Vidal thought that the epitheliomas develop in other structures besides the warts, as they found nodules of epithelioma in the middle of the corium, which they thought originated in the glands of the skin. It must be borne in mind, however, that on the Continent they use the term epithelioma in a somewhat wider sense than we do. With regard to the production of the ulcers I believe them to be due to infective pus. I have seen some of them evidently produced by the discharge from the eye, and in such ill-nourished tissues they are easily excited and kept up. The fact that in these cases they followed in the younger children so soon after their appearance in the elder suggests a similar explanation. If untreated they tend to fungate and perhaps become the site of an epithelioma, while some have described them as becoming rodent ulcers.

The appearance of the tumours marks the beginning of the end. Glandular implication and general infection are



rare, though it probably occurred in Hebra's first patient, who died of cancer of the peritoneum when twenty-seven years old ; but as the tumours multiply and fungate the health of the patient, which has hitherto been so remarkably good that no general symptoms belong to the disease, gives way at last from the continuous discharge and he dies marasmic and exhausted.

Vidal divides the disease into three periods :

1. The onset and development of the erythema and pigment spots.

2. The atrophy of the skin with its consequent dryness, wrinkling exfoliation, and accompanying telangiectasis and superficial ulcers.

3. The formation of verrucous projections whence the fungating tumours develop.

*Diagnosis.*—Few will dispute that the disease is *sui generis*, but errors may arise in its diagnosis. At the commencement the red spots have been mistaken for measles and the pigment spots for ordinary freckles ; time and the surrounding circumstances will clear this up. Later on difficulties have arisen from laying too much stress on one lesion to the exclusion of the rest ; thus the telangiectases have led to its being regarded as a rare form of *nævus*. From the scabbed ulcers and cicatricial aspect it has been pronounced to be a form of lupus ; but what was it before the ulceration commenced, and what kind of lupus begins as superficial sores scattered irregularly over the face and without evident preceding infiltration of the skin ?

But the disease that most closely resembles it is the atrophic stage of *scleroderma universalis*. Here we have immobility, white atrophy, telangiectases, and sometimes pigmentation, but in *scleroderma* the immobility is much greater, the telangiectases are rarely so conspicuous, and the pigmentation is not in lentiginous spots but in streaks, while the early history is totally different.

Indeed, error is almost impossible if the existence of the disease is borne in mind and all the lesions and the history of their development known.



The *prognosis* is gloomy enough. It is true that cases have gone on to adult age, and one which began late, viz. at nine years old, lasted for thirty years before the tumours made their appearance, but the majority succumb long before this and many have died under puberty. There is absolutely no hope of spontaneous improvement.

*Treatment.*—Everything yet tried, such as arsenic, tonics, cod-liver oil, and so forth has been absolutely futile. The removal of the tumours as a palliative is an obvious indication, and my experience of these three cases leads me to believe that much may be done for their relief by local treatment. The ulcers have nearly all been healed or are progressing in that direction. The recent ulcers have been treated with a weak ammonio chloride of mercury ointment, followed by boracic acid ointment. The old indolent or fungating ulcers were first scraped with a sharp spoon and then dressed with boracic acid lint or ointment. The eyes, under Mr. Tweedy's direction, have been diligently bathed with boracic acid lotion and greatly improved, and thus fresh ulcers from the irritation of the discharge on the cheeks has been avoided. Those who saw the cases in January will bear witness to their improved appearance, and though no doubt this is only temporary, constant watchfulness and early treatment of fresh lesions would almost certainly delay the progress towards death by saving the strength of the patient.

*Pathology.*—As to the general nature of the disease all Continental authorities are agreed that it is an atrophy of the skin, probably due to a neurosis, which we can only ascribe to a congenital predisposition, though all do not agree that the atrophy is primary.

Granting the existence of a trophic neurosis many of the symptoms are not without analogies in other diseases :

Excess of pigmentation, followed by total absence of it, is often seen in leucoderma, and probably pigmentation always precedes the white patches in that disease. As a degenerative change we see pigmentation in elderly people, and at the present time I have two cases of old persons



where, after removal of eczema, pigmentation has been left which in one of them is distinctly freckle-like. Atrophy with pigmentation and telangiectasis, as before remarked, is seen in scleroderma, but here infiltration of the tissue precedes the atrophy, the telangiectasis, being, probably, due to collateral hyperæmia consequent upon the blocking of neighbouring vessels by the infiltration. The ulcers appear to be accidents so to speak and to a great extent avoidable, but the warts which are the starting-point of the tumours are probably another evidence of degenerative change analogous to the pigmented warts so frequent in senile atrophy of the skin, and these warts too are not unfrequently the seat of cancerous changes. Kaposi and Neisser explain the production of epithelioma at so early an age as due to the active changes going on in the epithelial tissues, in the production and rapid disappearance of new papillæ and epithelium and of the pigment-producing elements. The disease appears to me best explicable by regarding it as one in which the nutrition of the tissue is damaged in the same direction as in senile decay, and therefore prone, like worn-out tissues generally, to resent irritation by the production of morbid and too often malignant growths. The pterygium, a condition rarely seen in the young, is another symptom of this premature sensibility. I certainly prefer the above analogy to the other put forward by Vidal, who asks :

“May it not be a variety of that numerous class of epithelial cancers of which the first and second stages are only the prelude, and the disease is only established at the third period ?

“In superficial epitheliomas from multiple foci, which in various forms we call benign epitheliomas, the cancrroid may be preceded for years by a dry seborrhœa or an epidermic exfoliation of a greyish or brownish colour. Gradually this lesion passes insensibly into a cancrroidal ulceration which, superficial at first, ultimately gets deep. Compare also the affection of the tongue which, after lasting perhaps twenty years, becomes cancerous, the epithe-



lium having gradually got thick and rugose, this tylosis being the first stage of the cancrioid."

One word about the name. Xeroderma is certainly an unfortunate selection, as that term is already in thoroughly established use for the mildest form of ichthyosis, with which this disease has nothing to do, and it can only therefore lead to confusion. Taylor's name, "*Angioma pigmentosum et Atrophicum*," depicts several features of the disease, but is too clumsy an appellation. The disease is admittedly an atrophy of the skin, and I would therefore suggest "*Atrophoderma pigmentosum*," which would call attention to two prominent symptoms, avoid confusion, and not be too complicated for ordinary use.



*Table of Cases of Xeroderma Pigmentosum up to January, 1884.*

No	Age.	Sex.	Consanguinity.	Date of onset.	Age when tumours first appeared.	Nature of tumours.	Observer.	Reference.	Remarks.
1	18	F.	—	Early childhood	?	—	Hebra and Kaposi	Hebra's Dis. of Skin, Syd. Soc., vol. iii, p. 254	Died of cancer of the peritoneum, æt. 27.
2	10	F.	—	"	11 years	Epithelioma	"	Do.	Died of cancerous cachexia, æt. 13.
3	8	F.	2 sisters	1 year	8 years	Sarcoma	Geber and Kaposi	Do.; also Archiv Dermatologie und Syphilis, 1873, Heft i	Tumours began on nose, probably on right side, as it was worse there.
4	6	F.		"	None	—	"	Do., do.	Geber called it a rare form of nevus.
5	5½	F.	Sister and brother	2 years	"	—	Kaposi	Medizinische Jahrbücher, 1882, p. 619	Had meningitis when 6 months old.
6	2½	M.		"	"	—	"	Do.	Not seen by Kaposi; described by parents.
7	17	M.	—	?	12 years	Epithelioma	"	Do.	First tumour on right cheek; became very numerous, and were destroyed.
8	22	M.	—	16 years	20 years	"	"	Do.	First tumour on left lower eyelid; numerous ulcers elsewhere, the first on right ear.
9	?	F.	3 sisters	Very early life	—	Angiomyxoma	R. W. Taylor	Abstract in Archives of Dermatology (American), vol. iv, pp. 66, 343	No details given.
10	?	F.		"	—	"	"	Do.	
11	?	F.		"	—	"	"	Do.	
12	?	F.	2 sisters	"	—	"	"	Do.	
13	?	F.		"	—	"	"	Do.	
14	?	F.		"	—	"	"	Do.	
15	?	F.		—	—	—	"	Do.	
16	40	M.	—	9 years	About 39 years	Epithelioma	Heitzmann	Do., vol. iv, p. 67	First tumour on left cheek.



		F.		6 months	None		Duhring	American Journ. of the Med. Sciences, vol. lxxvi, p. 424	Mother died of cancer of uterus. Has not altered from age of 9 years; scalp was invaded; freckles over the first lesion. Skin rough and flabby on neck and arms only; no mention made of pigment.  Many small warts, pin's-head size, on arms.
17	17		—		None	—			
18	5 mos.	M.	7 brothers	5 months	"	None	W. Rüder	Rüder, Ueber Epithelial-carcinom der Haut bei mehreren Kindern einer Familie, Berlin, 1880 (quoted by Neisser in Viertelj. für Dermat. und Syph., 1883, p. 51)	
19	18 mos.	M.		—	"	—	"		
20	4	M.		—	"	—	"	Do.	—
21	5	M.		—	About 5 years	—	"	Do.	Had a tumour on lower lip, which dropped off some weeks before he came under observation. Only scar left.
22	9	M.		—	8 years	Epithelioma	"	Do.	Tumour on lower lip, excised eight months before, recurred; warts very numerous all over affected area.
23	11	M.		—	11 years	—	"	Do.	An ulcerating wart beginning on right cheek.
24	10	M.		—	About 9 years	—	"	Do.	Extensive cancerous growth in temporal region was the cause of death.
25	24	M.	2 brothers	3 months	22 years	Epithelioma	A. Neisser	Viertelj. für Dermatol. und Syph., 1883, p. 55	Father died of obstruction of oesophagus. ? cancer. Began with red patches after a definite exposure to sun. Ulceration, which became epitheliomatous, began on right lower lid.
26	22	M.		"	None	—	"	Do.	Not seen by Neisser. Said to be exactly the same as brother, but no tumour.



No	Age.	Sex.	Consanguinity.	Date of onset.	Age when tumours first appeared.	Nature of tumours.	Observer.	Reference.	Remarks.
27	11	F.	{ 2 sisters }	3 years	9 years	Epithelioma, fungoid	E. Vidal	Annales de Derm. et de Syph., vol. iv, No. 11	Tumours began on right side of nose. Tumours began on right cheek, and others followed on same side. Died at the age of 11 from exhaustion.
28	11	F.		18 mos.	4½ years	"	"	Do.	
29	15	M.	{ 3 brothers }	2½ years	4 years	Epithelioma, verrucoid	"	Do.	Corneous vegetation on root of nose, right cheek, and upper lip.
30	12	M.		2 years	4 to 6 years	"	"	Do.	Most of tumours on the right cheek; had one which dropped off on right hand.
31	10	M.		"	5 to 7 years	"	"	Do.	Right side of nose showed first tumour; others followed on left side and median line of face; several dropped off. Died, at 10 years.
32	12	F.	{ 2 sisters and their brother }	Began between 12 and 18 months	10½ years	Papilloma, fungoid	H. R. Crocker	—	All the large tumours are on the right side of face, and most of the sores also.
33	10	F.			Sores at 4 yrs.	None	"	—	Has many crusted sores, but no fungous tumours yet; nearly all on right side.
34	9	M.			Sores at 3 yrs.	Papilloma, verrucoid	"	—	Has many crusted sores, mostly on the left side. He had one tumour which projected more than 1½, became strangulated, and dropped off. The sores developed on the two younger children within one month of their appearance on the eldest, their respective ages being 6, 4, and 3 years.



Of 14 cases where site of first tumour is mentioned definitely, 3 began in median line, 9 on right side of face, 3 on left.

*Table of Doubtful Cases (or which have been quoted as Xeroderma Pigmentosum).*

	1	26	F.	9 families. 26 cases.	29 in the course of 1st 2 years.	From 4 to 39 years.		Glax	Arch. für Dermat. und Syph., vol. ii, 1875, p. 114	
34 cases.	1	26	—	—	22 years	None	—	—	Arch. für Dermat. und Syph., vol. ii, 1875, p. 114	This is clearly a case of atrophic scleroderma.
	2	—	—	—	—	—	—	J. Hutchin- son	Lancet, vol. i, 1875	Quoted by Neisser. Mr. Hutchin- son reports it as a case of xero- derma of Wilson = Ichthyosis, which it undoubtedly is.
	3	—	—	—	—	—	—	Tanturri	Il Morgagni, Juli, 1877	Quoted by Neisser. Called "idio- pathic, pigmentary, telangiect- tatic, small-celled sarcoma of the skin." Have not seen original paper, but <i>primâ facie</i> it is not unlikely to be xeroderma pigmentosum.
	4	—	—	—	—	—	Unilateral idiopathic cutaneous atrophy, with pigmentation and hyper- trophy of cutaneous capillaries	Atkinson	Richmond and Louis- ville Med. Journ., Dec., 1877	Duhring, who has seen the original paper, considers this a case of morphœa, or closely allied to it.
	5	28	F.	—	—	—	General atro- phia cutis	E. Wilson	Diseases of Skin, 6th ed., p. 394	Began at 21 years. Closely allied to general Scleroderma.



DESCRIPTION OF PLATES III, IV, V.

Three cases of Xeroderma Pigmentosum (H. RADCLIFFE  
CROCKER, M.D.).

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PLATE III.

Alice Elizabeth Beddoes, æt. 12 years.

PLATE IV.

Benjamin Beddoes, æt. 9 years.

PLATE V.

FIG. 1.—One of many lobed masses embedded in granulation tissue from a filbert-sized tumour on the cheek.  $\times 100$ . Reduced one half.

*a.* Small epithelial cells filling the lobes.

FIG. 2.—Portion of the upper part of a small ulcer from the cheek.

*b.* Lowest layers of horny part; the rest has been excoriated.

*c.* Dilated vessels.

FIG. 3.—A single lobe of the papillomatous tumour.  $\times 350$ .