PURPURA NECROTICA
A POSSIBLE CLINICAL APPLICATION OF THE SHWARTZMAN PHENOMENON*

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This paper describes a disease entity which does not appear to have been previously recorded in Great Britain. It is based on three cases all affecting female children, two of which occurred in 1943, and the third in 1945. These children came from different towns in Staffordshire and had no connexion of any kind with each other.

Case Reports

Case 1. A female, aged five years, was brought to the out-patient department at the Royal Hospital, Wolverhampton, on February 24, 1943, on account of purpura, and was admitted at once.

There was no previous history of illness. The present illness began suddenly on February 17, 1943, with pains in the legs (which lasted for one week). Two days later there was swelling of both ankles which lasted for twenty-four hours and then disappeared, leaving a bruise. The next day bruising was noticed on the inside of the knees and on the buttocks, but the child went to school. When she happened to fall down the bruises were noticed and she was sent home, where she remained till she came to hospital four days later on account of a purpuric rash which had appeared in the meantime.

On admission both legs and buttocks were covered with purpuric patches of a peculiar character, the remainder of the body being free apart from a few small isolated lesions, and the mucous membranes were not affected. When discrete these purpuric patches all had the same appearance. They were of all shapes; some were circular or oval, but there was a definite tendency to angularity, the appearances being precisely similar to those of the next case (see fig. 2). They were raised above the surrounding skin, with a definite edge, and had a black, haemorrhagic bulla at their centre. Their general appearance is best described as a haemorrhagic urticaria. In addition to the discrete lesions there were a few areas of confluence where one simply saw a large purplish-black area. The two largest were on the outside of the left buttock and thigh, and these lesions attracted immediate attention on account of their remarkable shape. That on the thigh was shaped like a rectangle sharpened at one end, having a flat top and sides which converged downwards almost to a sharp point below, the whole appearance being distinctly artificial. The one on the left buttock had an even more artificial shape, being distinctly triangular except that one side was somewhat curved. Both areas were surrounded by a thin red inflammatory margin. All the four small toes on each side were black from their junction with the foot. The child's general condition was better than one would expect from the severity of the local lesions, and the temperature was only slightly raised (99°F.). Both knees and ankles were somewhat swollen and tender, but there was no limitation of movement. Blood count was normal (see below). Routine physical examination was entirely negative.

The disease had reached its maximum by the time of admission and no new lesions appeared thereafter. The subsequent changes in the purpuric areas were unexpected, for instead of gradually fading to a brownish colour and disappearing, an entirely different course of events occurred. In the first place the lesions during the next two or three days became very hard. The black skin covering the small toes was so stiff that the child could not move the toes; while the confluent areas on the left thigh and buttock became so hard that tapping them with the finger-nail gave the same impression that would be given by tapping a plaster-of-Paris bandage. This hardness of the lesions was a very striking clinical feature, and suggests that there had been a coagulation of the tissues involved.

The next and final stage was that of separation. The smaller discrete lesions peeled off as a scale of black epithelium, leaving pink and healthy new skin below. The black skin over the toes split and peeled off in a similar way, and the same sequence occurred in the confluent areas except for the two large ones on the left thigh and buttock. These began to separate at their edges, and it was at once apparent that deep necrosis had occurred. The slough from the oblong lesions on the thigh came away in about ten days, leaving a deep trench which had removed the whole depth of the subcutaneous tissues down to the fascia. The triangular patch on the left buttock went much deeper; the slough, which

* Based on a communication to the Association of Physicians of Great Britain and Ireland on April 12, 1946.
took fourteen days to separate, left a deep dome-shaped cavity extending approximately half way into the substance of the gluteal muscles. These cavities were lined with clean granulation tissue, and the moment the sloughs had separated the child's general condition became quite normal. The wounds then granulated unevenly and became covered with skin, though owing to their size this process took three months, and although the activity of the disease had ceased at the end of February, the child was not discharged from hospital until the end of May, 1943. There were no symptoms after this date, but owing to the size and depth of the lesions there was extensive and permanent scarring of the left buttock and thigh, which is shown in fig. 1, p. 17. They cause no disability.

Case 2. A girl, aged three years, was admitted to the Hallam Hospital, West Bromwich, on March 20, 1943. At eighteen months she had been immunized against diphtheria. In 1942 she had a generalized 'dermatitis' and also impetigo. She had then been well until February, 1943, when she had measles complicated by a right otitis media, from which she made a good recovery. She had been up and about for a fortnight before the present illness appeared. This began suddenly on the morning of March 17, 1943, when the mother noticed haemorrhagic areas on the back of the right lower leg, and the child was obviously not feeling well. On the evening of the same day a further haemorrhage appeared in front of the right ankle. Nothing further happened for thirty-six hours, when (March 19) a large haemorrhagic area appeared on the left buttock, and next day she was admitted to hospital.

On admission she was obviously a very ill child, pale and fretful, and in addition to the haemorrhagic areas already mentioned there were numerous smaller purpuric patches which, as in the previous case, affected almost exclusively the buttocks and legs. They were precisely similar to the individual lesions of case 1, being either rounded or having the same straight-sided geometrical or triangular outlines, were raised above the surrounding skin, and had the same central black haemorrhagic bulla. An isolated lesion on the shoulder is illustrated in fig. 2, p. 17. The temperature was normal, and routine physical examination revealed no abnormalities. The disease was still active, for the next day the right buttock became involved, a number of violet-coloured areas appearing and rapidly increasing in size to coalesce into one large area. This involved the whole right buttock and part of the adjacent thigh and contained a small islet of healthy skin which remained unaffected. This lesion had a distinctly artificial appearance, being more or less square-shaped, and its upper margin was formed by a particularly straight horizontal line. That on the left buttock was rather smaller and more rounded. On both sides there were prolongations passing inwards towards the perineum under the ischial tuberosities. As in the previous case, these large confluent areas were surrounded by a thin red inflammatory border. A week later, on March 28, 1943, the eyelids on both sides, the dorsum of the left hand, and the left heel became oedematous, and by March 30 both hands and the dorsal surfaces of both feet were also involved. On the same day the fronts of both knees became covered with haemorrhagic patches. These were thought to have resulted from pressure, for owing to the state of the buttocks the child could only lie on its face, and when awake it liked to rest in the knee-elbow position. The period of active illness now ceased, no further lesions appeared, and the child's general condition began rapidly to improve. The process of recovery went through the same stages as in case 1. There was the same preliminary hardening of the lesions, to an almost stony hardness, which then healed by desquamation of a patch of hard black epithelium, leaving healthy new skin beneath, while the larger areas which extended down into the deeper tissues healed by the separation of a large slough. This phase gave rise to great anxiety, for it soon became obvious that the sloughs in the buttocks went very deep indeed, and the risk of haemorrhage appeared sufficient to call for the constant presence of a special nurse provided with a bell and a tray of appropriate instruments. (The nursing difficulties were increased by the necrosis simultaneously present on the front of both knees, and the child's recovery under these circumstances is a tribute to the care of the nursing staff.) Separation was, however, quite uneventful, though it took twenty-one days to accomplish. This stage of the illness is shown in fig. 3 from which the great size of the necrotic areas can be realized. When the sloughs had come away, large clean cavities were exposed, covered with healthy granulation tissue, which from their size suggested large gunshot wounds. That on the right buttock was of enormous size and went very deep into the substance of the gluteal muscles, reaching almost to the bone. It is probable that the outer aspect of this buttock was only saved from complete destruction by the islet of unaffected skin; this was supported on a cone of tissue rising from the depths of the wound, and it thereby reduced the volume of tissue destroyed. The wounds gradually granulated and became covered with new skin as in case 1, except that owing to the larger size and greater depth of the lesion, skin-grafting had to be undertaken on the right buttock. Difficulties were experienced in this owing to the amount of fibrous tissue in the scar, with a resulting poverty of blood supply, and it has taken two years for the area to become completely healed. Ultimately six permanent scars have been left, one on each buttock and one at the back of the right ankle, which are illustrated in figs. 4 and 5. The scar on the right buttock covers an area of approximately eighteen square inches (see fig. 5). On the front of the legs there is a wide but superficial scar over each patella, and above the inner
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aspect of the left knee is a small rounded scar resulting from a deeply penetrating necrosis which reached almost to the femur. The child has been quite healthy since her illness.

These two girls obviously suffered from the same disease. The next case is thought also to be an instance of the same condition, though it was much less severe.

Case 3. A girl, aged eleven months, had been breast-fed for three months, and the previous history was uneventful. On March 5, 1945, she had a nasal discharge resembling that of, and probably due to, coryza, and which her doctor diagnosed as due to a cold in the head and teething. On the afternoon of April 16 the child became very miserable and moaned a good deal, so that the mother thought she had been too much in the sun, it being a very hot day. On April 17 a transitory rash appeared, which the mother says consisted of 'red, mottled marks,' affecting the face, back, arms, and legs. The mother thought this might be measles, but the doctor thought it was a heat rash; the spots appear to have been raised. When the napkin was changed on the morning of April 17 the buttocks were normal, but when they were later changed at midday the mother noticed a black bruise high on the inside of the right buttock. At the same time the child became even more fretful, she cried whenever she was touched, and, according to the mother, 'every limb seemed to ache.' On the next day (April 18) she lay still in bed, which was very unusual, and on April 19 the fingers and dorsum of both hands were swollen, and she was admitted to the Royal Hospital, Wolverhampton. Deep in the upper part of the fold of the right buttock was a small purplish-red area approximately half an inch in diameter, and there were red, tender swellings on the right palm and at the base of the right ring finger. There was no rise of temperature; the child was very fretful, had some photophobia and marked general muscular hypotonia. No further incidents developed, and in the course of the next week the area on the right buttock became black and hard as in the two previous cases, and during this period subcutaneous haematomas formed in the swellings on the hand. Whereas, however, the lesions on the hand disappeared by absorption, that on the buttock separated as a slough, leaving a surprisingly deep wound reaching to the muscle, and lined, as before, with clean granulation tissue. The slough took twelve days to separate, and after this there was an immediate improvement in the child's general condition, which rapidly became normal. The wound healed satisfactorily, leaving a scar with a number of lateral prolongations, and is shown in fig. 6, p. 17. Since then the child has been healthy.

In none of these three cases did routine physical examination reveal any abnormalities, the mucous membranes were unaffected, there was no enlargement of the spleen or lymphatic glands, and, apart from a rise of temperature to about 99-4° F. at its height, the disease was afebrile.

Laboratory investigations. These were necessarily limited, owing to prevailing war conditions.

Urine. Urine was normal in cases 1 and 3; in case 2 it contained a slight haze of albumin at the height of the illness.

Bullae. Cultures of the haemorrhagic bullae were sterile.

Blood counts. The blood counts and differential counts are shown in the table.

In case 2 the bleeding time (March 22) was 2½ minutes, and the clotting time 5½ minutes.

Changes in the blood picture were secondary in nature, there being merely an anaemia at the time of the haemorrhage and a leucocytosis during the phase of sloughing.

Summary of Clinical Symptoms

These three cases clearly suffered from the same disease, though the illness of the baby was much the least severe. The disease appears to begin without warning, for there is nothing common to the preceding history of the children; one had had no previous illnesses at all, one was convalescent from measles and had had an attack of impetigo a year previously, while the baby's only illness was a coryza a fortnight before. The acute phase lasted from approximately four to fourteen days, and was characterized by a group of symptoms typical of anaphylactoid purpura. Between them these children exhibited varying combinations of pains in the limbs (all cases); swelling in knees and ankles, followed by bruising (case 1); subcutaneous swellings affecting the eyelids, hands, and feet (cases 2 and 3)—followed once by a subcutaneous haematoma (case 3); and transitory red mottled rash (case 3). The two elder children had a very severe purpuric rash, which was limited to the buttocks and legs apart from a very few scattered lesions over the shoulders. The detailed appearance of the purpuric patches was characteristic of anaphylactoid purpura; they were raised above the surrounding skin, with a distinct edge, and showed a thin red margin at their periphery, while the centre of each had a black haemorrhagic bulla. Their only remarkable feature was a tendency to an unusual angularity of shape. All three cases had one or more areas of confluent purpura, which were remarkable for the geometrical shape of the larger areas, especially the triangular patch in case 1 and the rather square-shaped area in case 2. This acute phase of the illness was virtually afebrile, had no visceral manifestations, no affection of the mucous membranes, and no significant changes.
in the blood picture. The precise order of appearance of the clinical features varied somewhat from case to case; cases 1 and 3 began with pains in the limbs and general constitutional disturbance, while in case 2 areas of haemorrhage were the first manifestation.

After the acute phase recovery began at once, and took the same course in all cases. The initial stage was one of hardening of the lesions, which was a striking clinical feature and presumably indicated a coagulation of both the effused blood and the tissues concerned. After this the discrete purpuric lesions peeled off as a flake of hard black skin, leaving healthy new skin below. The larger areas of confluent purpura separated as sloughs, taking from ten to twenty-one days in the process, and leaving healthy cavities—lined with healthy granulation tissue—which in the more severe sites had extended deep into the underlying muscle. The moment the sloughs had come away the children were back in good health, and all that remained was for the cavities to become filled up with granulation tissue and covered with new skin. This process took six weeks in the case of the baby, three months in case 1, and two years in case 2. Case 2 has the largest scar, a patch on the right buttock covering some eighteen square inches. The children have been quite healthy since. The general picture was, therefore, one of an acute and short illness followed by a prolonged period of repair; except for large and permanent scars there was no permanent disability.

**Cases from the Literature**

There appear to be only two cases of a similar nature reported in the literature. The condition was first described by Martin de Gimard in 1884, and a further case was reported by Beinhauer in 1929.

**Martin de Gimard’s case.** The original account has not been available, and the following précis is taken from a transcription by P. Chevallier (1937). A child, aged eight years (de Gimard’s case 10), suddenly developed oedema over the malleoli; six days later there was a swelling on the face and left shoulder, and as the oedema disappeared the right side of the face became violet. By the eighth day the right side of the face and neck was occupied by an enormous ‘wine-coloured’ swelling, which in places was violet and contained islets of healthy skin. There were similar large swellings on both arms. On the eleventh day violet patches appeared on the ear, and on the twelfth day there was swelling of the right thigh. By the thirteenth day gangrene had appeared on the cheek and upper lip, and the upper parts of the arms were covered with black scabs. The area of gangrene on the upper lip separated on the eighteenth day, and that of the cheek on the twentieth day; in the succeeding days further areas of necrosis separated from the chin, the upper eyelid and the left forearm, removing the muscles and extensor tendons and exposing the olecranon. The child made a complete recovery, but was left with permanent and extensive scarring of the face and arm.

**Beinhauer’s case.** Under the title ‘Purpura haemorrhagica gangrenosa,’ Beinhauer (1929) reports the case of a male aged twenty-eight months.

The illness began with pain in the left ear. Two days later the child had pain in the left ankle, with a diffuse bluish-red discolouration of the whole foot, and during the next two days he developed haemorrhagic areas on both thighs, the left elbow, and the right hand. On admission to hospital the entire left foot, from the toes to four centimetres above the ankle, was covered by a purplish-red haematoma which was stony-hard, and above it there were smaller haematomas. The whole antero-lateral aspect of the left thigh and the postero-lateral aspect of the right thigh were covered with similar stony-hard purplish haematomas. There was a similar lesion on the left elbow and the right hand and fingers; this had a fusiform appearance owing to haemorrhage into the joints. Both buttocks were involved in a haematomatous infiltration which was continuous with the lesions on the thighs and penetrated inwards to the perineum. All these lesions had bullae filled with effused blood. During the next few days the lesions became gangrenous and began
to separate, removing the muscles and exposing the underlying bone. (A photograph taken at this stage shows exactly the same process as that depicted in fig. 2.) In view of an absence of thrombocytes from the blood splenectomy was then undertaken, together with amputation of the left foot and two fingers, but septicaemia developed and the child died three weeks after the operation.

Beinhauer lays stress on the following features, all of which are duplicated in the cases reported in this paper: (1) the stony-hard consistency of the necrotic areas, (2) their sharp definition from the surrounding healthy skin, and (3) the absence of any signs of absorption, the haemorrhage progressing directly into necrosis. The condition appears to have been afebrile until the septicaemia supervened.

There can be no doubt that both these cases are examples of the same condition as that reported in this paper. The term ‘purpura haemorrhagica gangrenosa’ which was used by Beinhauer has also been applied to cases in which a purpuric process followed by local gangrene has formed part of a septicaemic process. Martin de Gimard’s case 8 was of this type, and a further instance was reported by Chevallier (1937). Michael’s case (1920) was also probably of this type. A child, aged two years and a half, had intermittent attacks of purpura over some five months, associated with a temperature rising to 104°F. The child developed gangrene of the terminal phalanges of seven fingers and also of the left thigh and right buttock, ending with loss of the terminal phalanges. The child made an otherwise complete recovery. In a recent article, Marie and others (1946) have described four children who developed gangrenous purpura in the course of a meningococcal infection. The lesions resemble those under consideration in that they were indurated and had a sharp border, as if ‘traced with a pen,’ and a raised edge; but the necrosis was only subcutaneous, and scarring was not a prominent feature. The nature of these purpuric lesions associated with a septicaemia raises questions of great interest, but the differences appear sufficient to justify their clinical separation from the purpura of unknown origin followed by necrosis extending deep into the muscles which is the subject of this paper. In all five cases there has been no temperature and no evidence of infection; the bullae from the lesions in case 2 were sterile.

Discussion

In attempting to elucidate this condition, it is clear that it begins with an illness which is indistinguishable clinically from allergic purpura, though why all such purpuras do not follow the same course remains a puzzle. It appears also to be a disease of children, the recorded ages being eleven months, twenty-eight months, two and a half, three, five and eight years. It is also clear that the peculiar geometrical shape of some of the larger lesions is a feature which demands clinical explanation, for they are clearly artificial. It is impossible to imagine a general pathological process which, free from all restraint, would exteriorize itself in angular areas independent of all anatomical considerations. The scars shown in figs. 1 and 5 have distinctly artificial appearance, especially the straight sides joined by a right angle at the left upper corner in fig. 5. It would appear certain that they must be artificial in the sense of being self-inflicted though not, of course, deliberately so. There are two indications from the clinical histories which agree in suggesting that relative stasis may be an important factor. These are: (1) the localization of the generalized purpuric rash to the buttocks and legs; and (2) the appearance of areas of confluent haemorrhage on the front of the knees in case 2. There were three such areas, one over each patella, and a deep one just above the inside of the left thigh just above the knee. These did not appear till the eleventh day of the illness, and they coincided with angioneurotic oedema of the eyelids and hands. Their appearance at these sites was undoubtedly due to the fact that owing to the large lesions on the buttocks the child had to be nursed on its face and always liked to get into the knee-elbow position.

There is one possible approach to the geometrical outline of the larger lesions which will at the same time explain their localization. In case 1 we have to explain a triangular scar on the left buttock, and a tapering scar on the left thigh (see fig. 1). It is common to see linear marks on the buttocks of patients caused by pressure from the crumplings and folds of their pyjamas, and such lines at times mark out a triangle. At other times one sees areas of redness on the buttocks due to differential pressure from crumpled and non-crumpled areas of pyjama, and these areas are often geometrical in outline. Is it possible that the triangle on the buttock of this case was marked out in this way while the child was lying asleep on its left side? If so, the tapering scar lower down on the outside of the left thigh receives a similar and ready explanation. Such an explanation involves, however, the further assumption that during the active period of the disease there is a critical phase (or phases) of very short duration within which the sensitizing process reaches a sharp maximum, so that quite slight but continued pressure during this short period can, with the accuracy of a carpenter’s pencil, mark out areas on
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the skin subsequently to be the site of deep necrosis. Such an assumption provides a ready explanation of the general localization of the scars in case 2 (see fig. 4). There are large areas on each buttock—each of which has a process passing inwards under the ischial tuberosities—and a scar at the back of the right lower calf. The general appearance at once suggests that the child was sitting in a chair at the time the lesions were marked out. The chair habitually used was an ordinary high nursery chair for feeding the child at table, and the parents were in the habit of putting a large square pillow on the seat. Measurements show that this would have risen round the buttocks of the sitting child to approximately the height of the straight line marking the top of the scar on the right buttock, while the square-shaped outline of the upper portion of this scar is just such as could have been caused by pressure and creasing from a pillow. It is also easy to imagine how such pressure could leave an area of skin unaffected, like that present in the scar on the right buttock. The parents further state that the child used to like to loll on one side with one foot on the foot-rest and the other hanging down and resting against the upright of the chair. Such a position would account for the greater depth of the lesion on the right buttock, and for the presence of a scar at the back of the right ankle and the absence of one from the left foot. A similar explanation is also applicable to the small scar high up in the fold of the right buttock in the case of the baby. The child was very fretful during the first two days of the illness, and the lesion in this situation may well have derived from pressure of the mother's hand or fingers while nursing it. The localization of the lesions in Beinhauer's case is difficult to explain, but the necrosis of the right side of the face in Martin de Girard's case could easily have been delineated by pressure from the pillow while asleep.

It is, therefore, a reasonable clinical deduction that both the localization and the peculiar shape of the larger lesions in all three cases could have been due to slight but continued pressure acting during a critical phase—lying asleep on the left side in one case, sitting in a feeding-chair in another, and being nursed in the third. The lesions on the front of the knee in case 2 appear to be in a somewhat different category; they arose as the result of pressure continued over several days towards the close of the active period of the illness, when the child liked to be in the knee-elbow position owing to the state of its buttocks. Those on the knee-caps left quite superficial scars, while slightly above on the inside of the left knee was a small but deep area of necrosis. If the explanation advanced above for the larger areas is the true one, it has to be supposed that in the later stage of the disease any critical periods were of less intensity, and that is why long-continued pressure did not produce lesions of the same severity.

The only pathological process which will in any way meet these clinical requirements appears to be the Shwartzman phenomenon (1937). In this, a haemorrhagic necrosis is produced by two consecutive injections of a bacterial filtrate from a suitable organism. The first is given intradermally and the second intravenously some twenty-four hours later. There then develops at the site of the original intradermal injection a lesion which Shwartzman describes as follows: 'Four hours after the intravenous injection there appeared severe haemorrhagic necrosis at the prepared skin site. In the gross it was dark blue, swollen, with an angry red periphery, and histologically it showed disruption of the venules, extensive haemorrhage, thrombosis, and necrobiosis of all the cells. The reaction extended from the superficial layers of the skin through the entire thickness of the abdominal wall to the peritoneum' (p. 29). 'In typical strong reactions the early appearance is that of a crop of petechiae which continuously increase in size until there results an extensive confluent haemorrhage forming a sac filled with blood. The colour rapidly changes from blue to violet and almost black. There is an angry red zone at the periphery. The entire process may be so rapid that the petechial stage is indistinguishable' (p. 12). 'The healing of the strongly haemorrhagic lesion is slow. Sloughs which may form in about forty-eight hours after the intravenous injection are followed by gradual separation and scarring. The complete process of healing takes about ten days.'

The description of these experimental lesions coincides closely with the clinical appearances found in the three cases described. The initial development of purpuric patches in the skin which rapidly become confluent, followed by a deep necrosis involving the underlying muscles leading to the separation of a deep slough with subsequent scarring, presents a sequence of events of striking similarity in both the experimental and clinical phenomena. No histological examinations were made in the three cases under description, but fortunately Beinhauer examined the necrosis in the amputated foot of his case. This was characterized by inflammatory changes, mainly seen as a perivascular infiltration of white cells, together with widespread vascular dilatation and thrombosis; the deeper veins of the corium were completely occluded by thrombosis. These changes are closely similar to those described by Shwartzman. Shwartzman
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considers that an essential feature of his phenomenon is the escape of active material in the preparatory injection from the circulation into the tissue spaces, so rendering the tissue cells sensitive. The state of the blood vessels in the skin and subcutaneous tissues during the early phase of the disease—marked by such phenomena as angioneurotic oedema and swelling of the joints—would provide a ready mechanism for this. Shwartzman found that a definite time interval was required between the two injections, the skin not becoming reactive until eight hours after the first injection and becoming inactive again after thirty-two hours. The optimum period was twenty-four hours, and it seemed a possible inference that such an optimum period corresponded with the demarcation of the sites of maximum necrosis in the cases recorded. In further experiments, however, Shwartzman found that it was possible to replace the initial intradermal injection by an intravenous one, provided there was local stasis and hyperaemia. In the relevant experiments (p. 197), intravenous injections were made into the vein of a rabbit’s ear clamped at the base and exposed to heat to induce hyperaemia. Such ears invariably gave a marked reaction to the second provocative injection, and the appropriate state could be reached in a period of time from one-half to two hours (p. 336). Such a time-interval is of the appropriate length for the clinical requirements, which suggest that the larger areas of necrosis may have been delineated by local stasis from lying in bed on one side or sitting in a feeding-chair.

In view of the close agreement between the clinical and experimental data, it is suggested as a working hypothesis that these cases may represent naturally occurring instances of the Shwartzman phenomenon, for which the title of 'purpura necrotica' appears to be suitable.

Summary

Three cases are described of an illness which appears mainly, if not exclusively, to affect children. It begins with features typical of an attack of purpura; the purpuric lesions are associated with necrosis of the tissues affected, and healing is accompanied by separation of the resulting sloughs. These vary in severity from necrosis of the superficial layers of the skin to lesions penetrating deep into the underlying muscles, resulting in severe and permanent scarring. It is suggested that the disease may be connected with the Shwartzman phenomenon, and that in the early stages it may be characterized by short, critical phases during which the operation of slight but continued pressure may determine areas subsequently to be the site of deep necrosis. The illness leaves no sequelae apart from the scarring.

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References


(For Illustrations of this Article see page 17)
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