CASE NOTES

HYDROA VACCINIFORME AFFECTING THE EYE*

BY

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A CASE which has been observed over a period of 6 years is presented as a rare condition with certain features distinguishing it from previous cases described.

Case Report

A boy first attended the High Holborn Branch of Moorfields Eye Hospital in July, 1952, at the age of 6, on account of redness and discharge of the right eye accompanied by photophobia, these symptoms having been present for one week. He was seen to have a rash on the face and, on inquiry, the mother told us that it had been first noticed in early May of that year; furthermore, she thought there had been some transient rash during the Spring of the previous year. There was no similar trouble in the family.

Examination.—There was a marked papular and vesicular eruption on the exposed portions of the face, mainly the cheeks, bridge of nose, and forehead. A few pustules were present and there was some flaking of the superficial epidermis. Considerable blepharospasm was present, and in the right eye there was ciliary injection, corneal oedema, and infiltration in an interpalpebral distribution with some ingrowth of deep vessels.

Treatment.—The eye improved on atropine drops over the next fortnight but the trouble recurred in September, when both eyes became painful and injected; the patient was then found to have a bilateral keratitis which responded to local atropine and cortisone in 2 weeks.

Progress.—During the winter the eyes remained quiet. At that time a diagnosis of interstitial keratitis was considered but blood Wassermann reactions on the child and mother were negative.

The boy was next seen on April 7, 1953, when he had a central corneal ulcer in the right eye and an associated vacciniiform lesion on the right lower lid margin. The cornea healed within 3 weeks on homatropine with cortisone drops and aureomycin ointment.

There is no record of his attending again until April 16, 1955, when he was found to have an active corneal infiltration in both eyes which responded to similar local treatment in 3 weeks. At that time he was having chloroquine tablets from a dermatological clinic.

He attended again on April 21, 1956, with an injected left eye, and was seen to have a marginal infiltrate at 8 o'clock on the cornea with some deep-vessel formation. This responded well to local treatment but he was kept on cortisone until October. That year it is noted that the onset of the facial eruption preceded the eye symptoms by 3 weeks.

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Recent Examination.—On May 3, 1957, he attended with a recurrence of activity in the left eye (Fig. 1). The visual acuity was recorded as right 6/9, left 6/36. In the right eye there was an old deep opacity in the central portion of the cornea with ghost vessel formation (Fig. 2). In the left eye a central superficial corneal abscess was present, also a few deep vessels, and a faint flare with a few cells in the anterior chamber (Fig. 3).

FIG. 1.—Facial eruption in hydroa vacciniforme (May, 1957).

FIG. 2.—Right eye showing residual deep opacity of cornea (May, 1957).

FIG. 3.—Left eye, showing superficial abscess, pannus, and deep opacity of cornea (May, 1957).

Result.—This condition in the left eye gradually improved on gutt. atropine, with oculentum hydrocortisone, and chloramphenicol.

Laboratory Investigations:

1952	Blood Wassermann reactions and Kahn test (patient and mother)	..	Negative
	Repeat blood Wassermann reactions (patient)		Negative
1952-54	Blood and urine tests for porphyrins		None detected
	Screening test for porphyrins		None detected
	Ehrlich's test for porphyrilinogen		Negative
1954-57	Blood count normal		
	Van den Bergh's test negative to direct and indirect method		
	Serum alkaline phosphatase	} Within normal limits	
	Serum proteins		
	Thymol turbidity		
	Urine: Urobilinogen	} None detected	
	Urinary porphyrins		

General Clinical Characteristics

The general condition has been variously named in the past, being first described by Bazin (1862) as hydroa vacciniforme. Later, Hutchinson (1879, 1889) and other authors termed similar conditions summer eruption, prurigo aestivale, prurigo adolescentium, and acne prurigo. The tendency has been for very mild cases to be referred to as summer eruption, less mild cases as hydroa aestivale, and more severe cases as hydroa vacciniforme. Senear and Fink (1923) used the term hydroa aestivale for cases showing no scar formation and hydroa vacciniforme for those developing scars. It has also been suggested that these two conditions may vary in age and sex distribution. However, Möller (1900), using experimental methods, produced lesions of different intensity by altering the exposure to ultra-violet irradiation, and most authors agree that the apparent difference in the two conditions is mainly one of degree. The sex incidence is usually described as greater in males, but, though this is true of the familial cases, they comprise only some 10 per cent. of the total and in the largest published series a preponderance of females is found in the proportion of two to one.

The condition is that of a recurrent vesicular eruption, situated on the exposed cutaneous surfaces especially the cheeks, bridge of nose, ears, and backs of the hands. Usually commencing the third or fourth year of life, it gradually decreases in intensity up to puberty and then tends to disappear. It is found mainly in temperate climates and is most active in summer, occurring in successive outbreaks each lasting some 2 or 3 weeks. The causative agent is agreed to be actinic rays, with exposure to wind and extremes of temperature acting as subsidiary factors; some cases show recurrences in the winter during sunny weather when there is snow on the ground. The association with porphyrinuria has been shown in many cases, but more recently this has come to be regarded as a less important factor (Mathews, 1937). If it is present the urine may be wine-coloured and the teeth pink (erythrodontia).

The eruption is preceded by an itching sensation, then a red macule appears with consequent vesicle or bullous formation. The larger vesicles often become depressed in the centre and form a crust resembling a vaccination vesicle; later pustules may develop. The duration of such a lesion from its commencement to crust formation is 3 to 4 days.

Ocular Manifestations

There have been few descriptions of the ocular manifestations, the most detailed paper being that of Stokes (1940). Lesions of the lids usually occur as part of the facial eruption and, if severe, cicatricial ectropion may develop (Friede, 1921; Wendleberger and Klein, 1938). Chemosis may appear with vesicles on the conjunctiva which, on rupture, may expose the sclera, and sometimes even staphylococci may form. The corneal conditions previously described have either been associated with adjacent lid lesions or have developed as a primary type showing fine vesicles on the interpallebral cornea leading to denuded areas of epithelium and consequent vascularization.

In the differential diagnosis, dermatitis herpetiformis, lupus erythematosus, erythema multiforme, and pemphigus must be considered, but the characteristic distribution, age incidence, and limitation of the recurrent attacks to the late spring and summer should make for an easy diagnosis.

Treatment

This is a self-limiting disease. During the active phase protection from sunlight and hot winds by means of special clothing is important; wide-brimmed hats, high-necked dresses, and long sleeves are all helpful. Various protective lotions and creams have been devised, varying from simple oily lotions and lanolin to ointments containing quinine or aesculin to arrest actinic rays. Also creams containing disodium naphthol sulphonate, which absorb ultra-violet radiation, may be useful. Mepacrine and chloroquine administered orally can be effective, and more recently local corticosteroids have been used, but are not always so successful as one would hope.

Discussion

The case described demonstrates certain interesting features. The regularity of onset each year can be explained by the normal seasonal increase in exposure to actinic rays. It was proposed that one dermatological clinic should attempt to identify the exact wavelength responsible for the eruption. However, Blum (1941) and Epstein (1942), in their cases, found that there was no specific wavelength as the prurigo was provoked by more than one spectral region. Epstein, in describing the pathological reactions to light, found that in the experimental provocation of prurigo in these cases there must always occur an initial erythema and that the intensity of the eruption was proportional to the degree of erythema. That ultra-violet was not essential he showed by producing a rash with yellow-red light and even by alpha radiation. With regard to the close connexion with porphyrinuria, Blum and Pace (1937) showed that exposure to wavelengths to which porphyrins are known to sensitize the tissues (3,000–4,500 Å) did not generally produce lesions in hydroa vacciniforme. The mechanism of production of lesions, Blum suggested, was that of photo-allergic sensitivity, and he differentiated it from the photo-toxic reaction which is non-allergic and is seen in the sunburn and primary sulphonilamide response.

The hypothesis in the former process is that a pro-antigen is present in the tissues and is converted by light into the antigen; antibody formation is stimulated which may be fixed or circulating and in some cases the antibody can be passively transferred to a donor.

It has already been mentioned that two types of keratitis have been described in hydroa vacciniforme, a primary type and one secondary to adjacent lid lesions. The present patient has shown both these types on different occasions. In the primary type the interpalpebral distribution, together with the fact that the lesions were distributed throughout all layers of the cornea, might support the hypothesis that there is present in the cornea a photosensitizing substance. The deep keratitis was recognized to be very similar to typical interstitial keratitis, in which condition many believe that a "toxin" is elaborated by the spirochaete or other causative organism which sensitizes the cornea to some stimulus, either endogenous or exogenous.

The delay noted in this case between the onset of the eye condition after the appearance of the rash on the face could be due to the protection afforded by the lids; photophobia appears to be present even when the eyes are clinically quiet. In the secondary type of keratitis time would have to elapse for the development and possibly secondary infection of the lesions on the lids. Certain pigments have been found that sensitize normal tissues to light; these include eosin, fluorescein, bengal rose, and haematoporphyrin. It is questionable, therefore, whether fluorescein or bengal rose should be applied repeatedly to the eyes in cases of hydroa vacciniforme to detect epithelial damage of the cornea.

Summary

A case of hydroa vacciniforme is described, which has shown types of corneal involvement over the period of 6 years in which the eruption has been active. At times the appearance closely simulated interstitial keratitis. The condition is reviewed and the aetiology of the separate lesions is discussed.

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