

Lymphangiectasia haemorrhagica conjunctivae

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Leber (1880) described a condition in which a connexion between a conjunctival lymphatic channel and a conjunctival blood vessel resulted in either intermittent or permanent filling of the lymphatic vessel with blood. Leber named this phenomenon lymphangiectasia haemorrhagica conjunctivae.

Since that first report nine further cases have been described (Bartók, 1917; Conrad and Kühnhardt, 1957; Contino, 1935; Heydenreich, 1956; Leffertstra, 1962; Stepanic, 1958; Zimmerman, 1899). The following is the first report of a case of lymphangiectasia haemorrhagica in the English literature.

Case report

A 44-year-old Caucasian male attended the out-patients department complaining of intermittent attacks of soreness and redness of the left eye.

In the past he had had three attacks of iridocyclitis, which had been treated with ointment, drops, and two subconjunctival injections, the nature of which is unknown. No cause for the uveitis was discovered and there had been no recurrence for 15 years. There was no history of trauma.

His general health was good. His mother suffered from glaucoma, but there was no other significant family history.

Examination The visual acuity was 6/6 unaided in both eyes. The left bulbar conjunctiva was oedematous. In the subconjunctival tissue varicose lymphatic vessels showing sausage-like dilations and constrictions could be seen (Fig. 1). The chemosis was confined to the bulbar conjunctiva.



FIG. 1 *Case 1*
Varicose lymphatic vessels in subconjunctival tissue

in the interpalpebral region. The cornea, anterior segment, lens, and fundus were normal. The ocular tension measured with the Goldmann applanation tonometer was 11 mm. Hg. No lymphangiectasia or other abnormality of the right eye was found. Neither the regional lymph glands nor the parotid gland was enlarged.

Investigations An attempt was made to delineate the dilated, subconjunctival lymphatic vessels of the left eye with fluorescein. 5 ml. of a 10 per cent. solution of fluorescein was injected intravenously and the eye examined with the slit lamp using a cobalt blue filter. Fluorescence first appeared in the tear film and subsequently throughout the chemotic conjunctiva. The main dilated channels filled late, and were poorly fluorescent. In the conjunctiva of the right eye fluorescence was barely visible at any time.

The following day, 0.01 ml. methylene blue was injected adjacent to the chemotic bulbar conjunctiva of the left eye. The limbal lymphatics filled in 2 minutes; within their lumen, valves became visible and appeared to be functioning normally.

After 10 minutes the large vessels began to fill; few valves could be seen and they appeared incompetent. The flow of methylene blue in these vessels could be easily reversed by gentle pressure with a glass rod. At the end of an hour the methylene blue had disappeared from the limbal vessels but the large vessel still showed some colour.

It was thought that pressure might collapse the dilated lymphatics, therefore the left eye was firmly covered with a pad and bandage for 12 hours. However, when the pad was removed the following morning, the oedema was found to have persisted and the dilated lymphatics were filled with blood in a most striking fashion (Fig. 2). The vessels remained blood-filled for the next few days, the blood slowly diffusing into the subconjunctival space. Two weeks later the blood had cleared from the subconjunctival space and from the dilated lymphatics, but the chemosis and lymphangiectasia persisted unchanged.



FIG. 2 Case 1
*Dilated lymphatic
vessels filled with
blood*

Treatment An attempt was made to obliterate the abnormal lymphatics, since it was apparent that a connexion existed between them and conjunctival veins. Coagulation diathermy was applied at two points temporally and nasally.

Result Next day there was a considerable increase in the conjunctival chemosis but the dilated lymphatics had disappeared; 3 weeks later the chemosis had resolved, and apart from one or two isolated segments the dilated lymphatics had disappeared.

This case was presented at a clinico-pathological meeting at the Institute of Ophthalmology in October, 1967, and since that time four further cases of persistent oedema of "recurrent bloodshot eyes" have been referred by colleagues and are briefly reported below.

Case 2

A 22-year-old man complained of intermittent redness of the right eye for 4 months. The redness cleared after a few days but a "jelly-like" swelling remained between attacks. There was no history of previous eye disease or injury.

Examination There was extensive lymphangiectasia in the subconjunctiva, many of the vessels being filled with blood (Fig. 3). After 3 weeks the blood had cleared but chemosis remained.

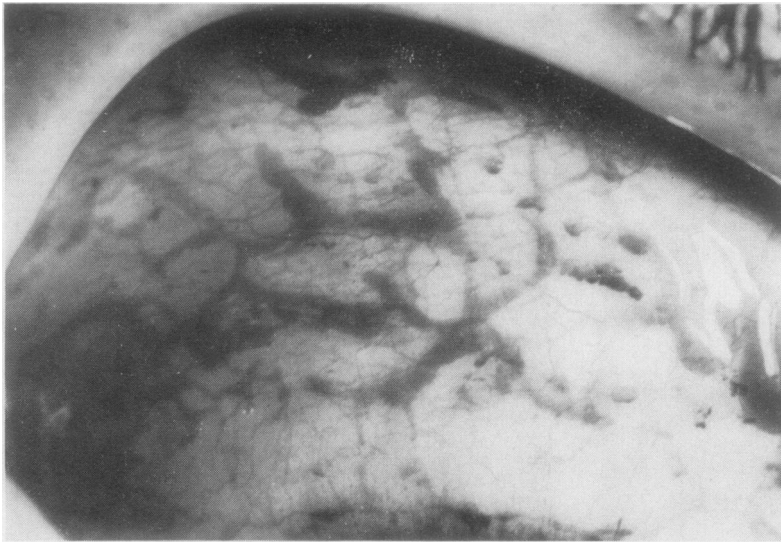


FIG. 3 Case 2
Subconjunctival
lymphangiectasia

Treatment Under general anaesthesia, a small section of subconjunctival tissue was excised for histological examination and other dilated segments were obliterated with coagulation diathermy.

Result Postoperatively the abnormal vessels had disappeared and within a month the eye appeared normal.

Case 3

A 2-year-old girl had had recurrent redness of the left eye since birth, especially after bouts of crying. A red patch appeared at the nasal limbus and gradually cleared over the next 2 or 3 days.

There was no history of direct injury to the eye, although forceps had been used at birth and there had been considerable bruising around the face.

Examination There was a patch of distended lymphatic vessels in the nasal bulbar conjunctiva, some of which contained blood at the time of examination.

Treatment Because of the mildness of the disability no treatment was given.

Case 4

A 49-year-old man complained of numerous attacks of redness of his left eye for "many years". Between attacks there was some "bogginess" of this eye. As a child he had had three attacks of crsipelas around the left eye.

Examination When he was first seen his left eye showed a resolving subconjunctival haemorrhage affecting its lower half, and in this area dilated irregular blood-filled lymphatic vessels could be seen; 2 weeks later the blood had cleared but typical lymphangiectatic vessels were present throughout the previously affected area.

Treatment None was given.

Case 5

A 35-year-old woman had had six attacks of subconjunctival haemorrhage in the previous 2 years. No history of trauma or previous eye trouble was given.

Examination There was an area of resolving haemorrhage with typical ectatic lymphatic vessels filled with blood; 2 weeks later the haemorrhage had cleared but the dilated lymphatic vessels remained.

Treatment None was given.

Discussion

The literature contains nine reports of this unusual condition (Table I).

Table I Previous reports of cases of lymphangiectasia haemorrhagica

Author	Date	Patient		Eye involved	Previous or existing pathology
		Sex	Age (yrs)		
Leber	1880	F	28	R	None known
Zimmerman	1899	F	14	L	Chronic conjunctivitis
Bartók	1917	M	27	L	Angioma of eyelid
Contino	1935	F	50	R	Trachoma
Heydenreich	1956	M	50	R	None known
Conrads and Kühnhardt	1957	F	18	L	Corneal foreign body
Stepanik	1958	F	12	L	Parotid lymphangioma
Leffertstra	1962	F	53	L	Marginal keratitis
		M	60	Not stated	Recurrent marginal keratitis

In five patients there was a history of some previous corneal or conjunctival disease; in three patients no history of previous eye disease was given. In the cases here described (Table II), only one patient had a definite history of ocular disease.

Table II Summary of present series of cases

Case no.	Sex	Age (yrs)	Eye involved	Previous or existing pathology
1	M	44	L	Subconjunctival injections for uveitis
2	M	22	R	None known
3	F	2	R	Forceps delivery. Bruising around eyes
4	M	49	L	Childhood erysipelas × 3
5	F	35	L	None known

Pathological reports are rare, those of Conrads and Kühnhardt (1957) and Leffertstra (1962) being the most recent. The specimens show grossly dilated blood-filled lymphatics surrounded by a minimal inflammatory reaction. In Case 2 in the series here reported a small segment of the abnormal lymphatic was excised, and histologically showed a similar appearance of dilated lymphatic vessels with slight surrounding cellular infiltration.

It is clear that the condition is the result of an abnormal communication between venous and lymphatic channels, but the site and aetiology of such communications are obscure. They may be the result of faulty development as would seem likely in the case of the year-old child who had suffered from recurrent redness of the eye since birth (Case 3) they may result from degenerative or inflammatory processes.

However, if the condition is sufficiently severe to require treatment, it has been proved to be eminently remediable by diathermy coagulation of the distended lymphatic vessels. Excision has also been carried out successfully without undue operative haemorrhage, but this leads to further unnecessary scarring. In those cases where oedema is minimal and symptoms few, no treatment is necessary.

Summary

Five cases of lymphangiectasia haemorrhagica conjunctivae are described. In two, transconjunctival coagulation diathermy resulted in disappearance of the abnormal segment of lymphatic vessel.

It is suggested that this condition is not as uncommon as previously supposed, and that some cases of recurrent subconjunctival haemorrhages may indeed be further examples of this abnormality.

My thanks are due to Mr. Lorimer Fison and Prof. Barrie Jones for encouraging me to report these cases.

References

- BARTÓK, E. (1917) *Klin. Mbl. Augenheilk.*, **59**, 53
 CONRADS, H., and KÜHNHARDT, G. (1957) *Ibid.*, **131**, 670
 CONTINO, F. (1935) *Ann. Ottal.*, **63**, 281
 HEYDENREICH, A. (1956) *Klin. Mbl. Augenheilk.*, **128**, 83
 LEBER, T. (1880) *v. Graefes Arch. Ophthal.*, **26**, pt 3, p. 197
 LEFFERTSTRA, L. J. (1962) *Ophthalmologica, (Basel)*, **143**, 133
 STEPANIK, J. (1958) *Klin. Mbl. Augenheilk.*, **132**, 99
 ZIMMERMAN, F. (1899) *Beitr. Augenheilk.*, **4**, 485 (heft 37, p. 19)