Complete avulsion of the optic nerve

A clinical, angiographic, and electrodiagnostic study

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The optic nerve may be damaged in trauma to the head either with or without fracture, and sometimes after an apparently trivial injury. The intracranial portion of the nerve is rarely involved because of its relative mobility but optic atrophy from damage to the intracanalicular or intraorbital portions is well documented. The mechanism of damage in the canal is by extension of bone fracture, direct penetration of a foreign-body, or by shearing forces tearing the pial covering of the nerve. Damage in the orbit usually follows the penetration of a foreign-body.

Damage may also occur at the disc, the suggested mechanisms being by anterior luxation of the globe, retropulsion of the nerve, forced globe rotation, or a sudden explosive rise in intraocular pressure blowing the nerve off the sclera into its dural sheath like a cork from a bottle. A case of *partial* rupture of the lamina cribrosa after blunt injury was reported by Lang (1901). There have been a number of similar case reports and we have reviewed papers by Goldsmith (1903), Hesse (1907), Birkhäuser (1910), Pichler (1910), Gonin (1912), Treacher-Collins (1914), Juler (1916), Loewenstein (1943), Spizziri (1964), Kommerell (1966), Hart and Pilley (1970), and Park, Frenkel, Dobbie, and Choromokos (1971). The last two papers reported fundus fluorescein angiography findings.

The first full clinical description of *complete* avulsion of the optic nerve was by Salzmann (1903) who adopted the term 'Evulsio nervi optici' and who gave credit for the first case presentation to Aschmann (1884). Further case reports have been made by Gagarin (1904), Weinstein (1908), Stoeber (1910), Lister and Hine (1919), Gruber (1959), and Kovács (1971). The true incidence of both partial and complete avulsion of the optic nerve is undoubtedly greater than is suggested by these cases reported over 90 years. Extensive vitreous haemorrhage usually delays or prevents clinical diagnosis in the early stages and the late

stage may be confused with developmental anomaly (Frost, 1903; Stanton-Cook, 1953). The absence of major haemorrhage allowed a definite diagnosis of complete avulsion to be made in the accident unit in this case, which is presented as the first case of complete avulsion of the optic nerve followed-up clinically and by serial fluorescein angiography, electroretinography, and visually evoked cortical responses to investigate the effects upon retinal circulation and function.

Case report

1 APRIL 1974

A 26-year-old woman was the front passenger of a car involved in a collision. She hit her head against the dashboard as she was not wearing a seat belt and became unconscious. On arrival at the general accident unit she was fully conscious with bruising to the left shoulder, upper arm, and ribs. X rays confirmed minor fractures of the left malar bone, second rib, and the little finger. The left cheek was swollen and tender, there were lacerations to the side of the mouth, and the left eye was noted to be blind.

Ophthalmological examination 14 hours after injury revealed a normal right eye but a left eye with bruising of both lids, ptosis, and no perception of light. The cornea was bright, the anterior chamber clear, and the pupil semidilated exhibiting brisk consensual but absent direct light reflexes. The vitreous was clear and the disc replaced by a deep round hole with a clear sharp rim surrounded by an apparently bared ring of sclera one disc diameter wide, Fig. 1. Despite a little preretinal haemorrhage the retina was seen to be flat with patent branch retinal vessels which could not be traced back across the bared area into the disc hole. There was no sign of the central retinal vessels which appeared to have retracted into the hole. The intraocular pressures were normal and equal to digital assessment, there was no proptosis, and the external ocular movements were full.

4 APRIL 1974

The posterior pole of the left fundus was oedematous and the circulation in the branch retinal vessels was

visibly reduced with fragmentation of the blood columns ('Cattle-trucking').

9 APRIL 1974

The left fractured zygoma was raised *via* a Caldwell-Luc approach.

17 APRIL 1974

The disc hole was unchanged, the retina flat, and the branch retinal vessels appeared to be normal.

Fluorescein angiography

The circulation was normal in the major retinal vessels but appeared to stop at the edge of the bared area without continuity into the disc hole. The bared area presented a collection of irregular choroidal vessels, one of which joined the superior temporal artery at right-angles (Fig. 3). Leakage of fluorescein occurred from these irregular vessels staining the bared area and the disc hole (Fig. 4).

Electroretinography

Devices 3182 photic stimulator, Lovac scleral electroretinography contact lens, Devices 3542 high gain amplifiers frequency range 1.0 Hz to 100 Hz.

Right eye—Normal response, a-wave 190 μ V, b-wave 440 μ V.

Left eye—Reduced and delayed response, a-wave 170 μ V, b-wave 200 μ V.

(Measurements from the 10-min scotopic high intensity flash recordings (Fig. 5)).

Visually-evoked cortical responses

Devices 3182 photic stimulator, Medelec MS6 Unit, amplifiers frequency range 0.8 Hz to 500 Hz.

Right eye—Within normal limits.

Left eye—No response. (Fig. 6).

3 JUNE 1974

The disc hole was almost completely filled with white connective-tissue obscuring the previously-sharp rim. A pigment ring had appeared along the outer lip of the bared area except between 4 and 6 o'clock where the retina was locally tented in a white veil-like formation.

Fluorescein angiography

The retinal vessels filled normally and then curled proximally around the edge of the torn retina to disappear into the choroid around the edge of the connective-tissue mass. This mass showed fluorescein staining

centrally and leaking fine irregular choroidal vessels peripherally (Fig. 7).

Lid electroretinography

Lower lid skin electrode, Devices 3182 photic stimulator, Medelec MS6 Unit, amplifiers frequency range 0.8 Hz to 500 Hz.

Right eye—Normal response, a-wave 10 μ V, b-wave 40 μ V.

Left eye—Reduced response, a-wave 10 μ V, b-wave 10 μ V.

(Measurements from the 10-min scotopic high intensity flash recordings).

Visually-evoked cortical responses

Right eye—Within normal limits.

Left eye—Suspicion of low amplitude 3 μ V positive component around 110 msec.

1 JULY 1974

Retinal haemorrhages had disappeared leaving the retina flat apart from local tenting beside the disc hole.

20 JULY 1974

Extensive retinal detachment radiating from the edge of the disc connective-tissue mass between 3 and 7 o'clock.

Fluorescein angiography

This showed patent retinal vessels. Profuse staining of the disc connective-tissue mass with fluorescein could be seen.

Electroretinography

Right eye—Normal response, a-wave 160 μ V, b-wave 410 μ V.

Left eye—Reduced response, a-wave 25 μ V, b-wave 65 μ V.

(Measurements from the 10 min scotopic high intensity flash recordings, Fig. 8).

Visually-evoked cortical responses

Right eye—Within normal limits.

Left eye—No response.

9 SEPTEMBER 1974

Connective-tissue completely filled the disc hole and thick retinal folds radiated from its edge between 2 and 7 o'clock. The retinal vessels were patent, Fig. 2.

FIG. 1 *Fundal appearance 17 days after traumatic complete avulsion of optic nerve. Disc replaced by round hole surrounded by area of apparently bared sclera*

FIG. 2 *Fundal appearance 5 months after complete avulsion of optic nerve. Disc hole filled with connective-tissue. Retina lying in thick folds*

FIG. 3 *Fluorescein angiography (arterial phase) 17 days after complete avulsion of optic nerve. Retinal vessels show normal circulation. Irregular choroidal vessels in bared area with one vessel (arrowed) communicating with superior temporal artery*

Electroretinography

Right eye—Normal response, a-wave $230\mu V$, b-wave $510\mu V$.

Left eye—Reduced response, a-wave $50\mu V$, b-wave $90\mu V$.

(Measurements from the 10 min scotopic high intensity flash recordings).

Discussion

The clinical progress of this case closely followed the first description by Salzmann (1903). The disc was replaced by a hole surrounded by an area of apparently-bared sclera and over the next 2 months this hole filled with white connective tissue. Local tenting of the retina at the edge of the hole produced a white veil-like appearance which has been described in the past as an extension of the connective tissue into the retina. The circulation in the branch retinal arteries and veins appeared to be good apart from an early transient disturbance. This was surprising as these vessels could be traced to the edge of the bared area where they had been divided from the central retinal vessels.

The circulatory changes of the cases reported in the literature are summarized in the Table. Salzmann (1903) and Gagarin (1904) both described branch retinal vessels which were filled with blood despite having lost continuity with the central retinal vessels in the disc hole, as in our case. Salzmann (1903) suggested the possibility of anastomosis between the choroidal and retinal vessels

FIG. 4 *Fluorescein angiography (venous phase) 17 days after complete avulsion of optic nerve. Leakage of fluorescein shown from irregular choroidal vessels. Disc hole and bared area staining with fluorescein*

FIG. 5 *10 min scotopic electroretinogram to high intensity flash 17 days after complete avulsion of optic nerve. Retina flat.*

Right eye: Normal response a-wave $190\mu V$, b-wave $440\mu V$.

Left eye: Reduced response a-wave $170\mu V$, b-wave $200\mu V$

to explain the maintenance of the circulation, and Stoewer (1910) supported this theory. Pichler (1910) reported seeing a vessel communicating between the choroidal vessels in the bare area and the retina.

Fundus fluorescein angiography has been reported in two cases of *partial* avulsion of the optic nerve. Hart and Pilley (1970) found delayed filling and emptying of veins in the affected quadrant with leakage of fluorescein from the damaged nerve and Park and others (1971) found no abnormality in their case. Angiography in our case of *complete* avulsion confirmed normal circulation in the branch retinal vessels and a communication was shown between peripapillary choroidal vessels and

FIG. 6 Visually evoked cortical responses. Right eye (RE) normal response. Left eye (LE) no response. RH shows trace from right hemisphere, LH shows trace from left hemisphere

FIG. 7 Fluorescein angiography 9 weeks after complete avulsion of optic nerve. Disc hole filled with connective-tissue. Retinal vessels shown curling around edge of retina

the superior temporal retinal artery. Although it has been suggested that both circulations may contribute to the blood supply of the optic disc, the existence of communication between the choroidal circulation and branch retinal arteries is not traditionally accepted. With the disappearance of the central retinal vessels such communication appears to be the only possible route for the maintenance of retinal circulation.

It has been suggested that the optic nerve may

FIG. 8 10 min scotopic electroretinogram to high intensity flash 8 weeks after complete avulsion of optic nerve. Retina detached.

Right eye: Normal response a-wave $160\mu V$, b-wave $410\mu V$.

Left eye: Reduced response a-wave $25\mu V$, b-wave $65\mu V$

contain centrifugal fibres which exert an inhibitory effect upon the retina. Many studies have been made of electroretinography after optic nerve transection and the results have been conflicting. Jacobson and Gestring (1958) found increased electroretinogram amplitude after optic nerve transection in lightly-anaesthetized cats. Jacobson and Suzuki (1962) transected cat optic nerves and found that the electroretinogram response to the second of a double-flash stimulus exceeded that to the first flash, the reverse of that with intact optic nerve. Brindley and Hamasaki (1962) found that optic nerve transection with or without anaesthesia in cats had no effect upon the electroretinogram. Gills (1966) investigated two cases of old optic nerve transection in man and obtained higher electroretinogram responses than in the intact fellow eyes. Nagaya, Oishi, and Kuno (1962) studied optic nerve transection in 20 cats by double-flash electroretinography and obtained results in support of centrifugal optic nerve fibres. Brindley and Hamasaki (1966) sectioned cat optic nerves and found no histological evidence in support of centrifugal fibres. Horsten and Winkelman (1969) obtained normal serial electroretinograms for $1\frac{1}{2}$ hours after stereotactic destruction of the optic nerve in a newborn puppy. van Hasselt (1969) studied double-flash electroretinograms after optic nerve transection in unanaesthetized rabbits and found results supporting the existence of centrifugal fibres.

The different results obtained in these studies may reflect the use of different experimental animals, electrodiagnostic equipment, or techniques, and van Hasselt (1972) reviewed the literature extensively in an attempt to reveal possible sources of error or misinterpretation. In our case of complete optic nerve avulsion electroretinography on the seventeenth day showed a normal a-wave but a b-wave amplitude reduced by 54.5 per cent compared with the intact fellow eye. At this

Table Review of retinal vascular disturbances in cases of partial (P) and complete (C) avulsion of optic nerve reported in literature

<i>Paper</i>	<i>Date</i>	<i>Degree</i>	<i>Injury</i>	<i>Retinal vascular disturbance</i>
Aschmann	1884	C	?	Pathological report only
Lang	1901	P	Clothes-prop	Nil
Salzmann	1903	C	Revolver shot	Normal calibre vessels stopping at edge of disc hole
Goldsmith	1903	P	Wood	Vessels continue into hole
Gagarin	1904	Rt C	Horsekick	Vitreous haemorrhage—no view
		Lt C	Horsekick	Normal calibre vessels not continuing into hole
Hesse	1907	P	Stick	Hyphaema—no view
Weinstein	1908	Rt C	Revolver shot	Vitreous haemorrhage—no view
		Lt C	Revolver shot	Vessels attenuated
Birkhäuser	1910	P	Cow's horn	Arteries thin—veins dilated
				Vessels not continuous with disc
Stoewer	1910	1 C	Umbrella	Arteries collapsed—veins dilated
		2 C	Stick	Peripheral vessels patent
Pichler	1910	P	Horsekick	Vessels not continuous with hole
Gonin	1912	1 P	Ski	Nil
		2 P	Ruler	Nil
Treacher-Collins	1919	P	Bat	Affected vessel convoluted and not reaching disc
Juler	1916	P	Stone	Arteries narrow—veins normal ending some distance from the disc
Lister and Hine	1919	C	Shrapnel	Vessels attenuated—fundus drawing shows vessels failing to reach hole
Loewenstein	1943	P	Nail	Not recorded—? nothing abnormal discovered
Gruber	1959	C	Road accident	Normal calibre vessels traced to hole
Spizziri	1964	P	Stick	Tortuous arteries—dilated veins bending over rim into the hole
Kommerell	1966	P	Hook	Vessels normal, continuous with disc
Hart and Pilley	1970	P	Hand	Vessels normal, continuous with disc
Kovács	1971	C	Explosion	Haemorrhage—poor view
Park and others	1972	P	Finger	Vessels normal calibre, continuous with disc

stage the retina was flat with circulation present in the retinal vessels. A further fall in the amplitude of the electroretinogram a-wave (84.4 per cent) and the b-wave (84.1 per cent) compared with the fellow eye occurred after 8 weeks when the retina detached. Our electroretinogram results do not support the theory of the existence of centrifugal fibres in the optic nerve of man, although it is possible that we may have missed an early transient supernormal response or that the retina may have been damaged by the transient vascular disturbance seen on the fourth day.

Summary

A case of traumatic complete avulsion of the optic nerve is reported together with fluorescein angi-

ography and electrodiagnostic findings. Despite the disappearance of the central vessels, circulation remained in the branch retinal vessels and angiography demonstrated communication between peripapillary choroidal vessels and the superior temporal artery. The visually-evoked cortical response was abolished. Electroretinography showed a normal a-wave but reduced amplitude b-wave, not supporting the theory of the existence of centrifugal retino-suppressive fibres in the optic nerve of man.

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