As this case seems nearly unique, I felt justified in publishing it, especially as it may be of use to other surgeons who are dealing with a similar case.

W. W. H., aged 74 years, consulted me in March, 1942, in reference to his eyesight. On examination of his eyes I found as follows.

Right eye, a well marked cataract; under homatropine and cocaine the pupil was well dilated and, though no details of the fundus could be seen, the disc appeared normal; the vision was less than 6/60. Perception and projection of light were very good.

Left eye.—Vision with glasses 6/36; under H. and C. pupil dilated well. Macular region was clear and no haemorrhages were seen. C. Sph. — 5'00 Sph. ⊕ Cyl. 1'50/17° = 6/18.

On questioning the patient the following history was elicited. In December, 1940, he consulted an ophthalmic surgeon, who made the following report:—

Received for publication, June 24, 1944.
Right eye.—Early cataract. Left eye.—Very suffused disc, ? hypertensive.

It was suggested he should have thorough examination. His blood pressure was 160/90. Urine showed no sugar or albumen.

Apparently the sight became worse. In January, 1941, he was examined again and a retinal haemorrhage at the macula was found in the left eye. It was suggested he should have a blood count done; the report was:

Red blood corpuscles 7,250,000. White blood corpuscles 12,000.

Stained Slide no abnormal white cells seen.

Diagnosis Polycythaemia.

As the sight became worse, the question of the right lens being extracted arose, but the surgeon thought this was unwise, as he felt there might be some lesion in the right fundus which might prevent a good visual result. The patient was sent to a physician in reference to his blood condition, and it was decided that no special treatment was necessary.

I was asked by the patient in March, 1942, whether I felt justified in operating and, as the projection of and perception to light were good and the pupil dilated fully under H. and C. and tension was normal, I said that I felt an operation for removal of the lens could be done—though I could not promise what the visual result would be.

In June, 1942, I extracted the right lens, the operation being extrascapular with a small peripheral iridectomy. Except for a slight attack of gouty conjunctivitis (the patient had an attack of gout on his finger), the convalescence was uneventful, the visual result C. +11·0 Sph. +3·0 Cyl.=6/6. Add +3·5 Sph. =J.1

On examination of the fundus, just below and internal to the macular region, was a patch of old choroiditis, in appearance resembling Tay’s choroiditis, but fortunately it was clear away from the macular region.

On February 21, 1943, the vision of the right eye was still 6/6 and I was asked, as the sight had greatly deteriorated in the left eye, whether I could extract the left lens. I said, as there had been active mischief in this eye and a retinal lesion at the macula, I could not be sure what the result would be, but that I would extract the lens if the patient wished it.

The second operation was undertaken in June, 1943. The result was perfect and there were no complications.

Vision C. + 11·0 D. sph. =6/6; +3·5 D. sph. added =J.1

Having become interested in this condition, I looked up numerous papers and abstracts from foreign journals, especially with reference to operative procedure. I could only find one instance of any operative interference and this was a case of Professor Ballantyne’s
who was obliged to do a trephining operation in a case with secondary glaucoma.

As I have been kindly allowed to reproduce the paintings of the fundi in a case of polycythaemia vera under the care of Mr. Nutt, of Sheffield, I feel a short description of the condition would not be out of place.

Polycythaemia is a clinical condition caused by a primary hyperplasia of the erythroblastic elements of the bone marrow. Its origin is unknown; on examination of the fundus special lesions are present. These are so characteristic that Ascher used the term fundus polycythemicus.

There are two types of polycythaemia:

1. The primary form, or polycythaemia vera, is characterised by the increase in number of red blood cells, the haemoglobin content, and the volume of blood, and is accompanied by headaches, malaise, nervous symptoms, general cyanosis and enlargement of the spleen. It generally occurs between the ages of 35 and 65.

2. The secondary or compensatory form is usually due to congenital heart disease, may be due to emphysema or stenosis of pulmonary vessels, and has occurred in cholera and dysentry. It may occur at any age; in the secondary type the cyanosis is more marked. Either type may be associated with mild or severe fundus changes.

Another type of polycythaemia is known as polycythaemia hypertonica or Gaisböck disease. This type is associated with arterio-sclerosis, cardiac disease and renal changes, and is accompanied by high blood pressure without splenomegaly.

The characteristic blood picture of polycythaemia is a high increase in the number of red blood cells—from 7,000,000 to 10,000,000, with an increase in the haemoglobin from 110 to 126 per cent. The colour index is low, but the volume and viscosity of the blood are increased.

The characteristic findings in the fundus in polycythaemia are marked distension, and engorgement of the retinal veins, which appear purplish. The distension is due to the increase of the blood volume and thinning of the venous walls.

In the secondary type, the retinal arteries are also moderately dilated and purplish. Other changes which have been described are retinal thromboses, small scattered haemorrhages, round shaped in appearance.

Hyperaemia of the disc is often seen. Some authors have also described a bluish cupping of the disc, and have stated that in typical cases of polycythaemia without fundus lesions, but with interference of vision, a retrobulbar neuritis may be present. With the slit-lamp the vessels of the iris are sometimes found dilated, with a brown area of dust-like pigment.
External changes in the eyes have been noted in some cases, such as congestion of ocular and tarsal conjunctivae, but these changes are shown generally in the severe congestive types of secondary polycythaemia.

Visual acuity in polycythaemia. In reading through numerous cases one is struck by the good vision these cases have—even after severe changes have been noted; and in the cases where the vision is poor, there are generally added complications, which do not seem due to the actual blood condition, but secondary conditions which have arisen due to the patient’s general state of health.

The paintings which the editorial committee of the Brit. Jl. of Ophthal. have kindly permitted to be reproduced in colour show the cardinal signs of polycythaemia very plainly, namely, the very engorged, tortuous and swollen retinal veins, the hyperaemic discs and the marked venous haemorrhages.

The paintings are from the fundus of a woman, aged 36 years. The vision was:—Right eye 6/9, Left eye 6/9. Blood count (August, 1928), 19 million red corpuscles, haemoglobin 140 per cent.

History. 1921, after the birth of twins, first noticed a high colour on her face. She began to have attacks of dyspepsia and headaches, later vertigo and vomiting. Spleen, easily palpable, colour index, 0.66.

Treatment. Venesection (repeated) from October 28 to November 21, 1928, with administration of phenyl hydrazine, which reduced the blood count to red cells 5,170,000.

On January 13, 1929, vision with glasses was 6/5, Right and Left. Visual fields were contracted, especially to red and green.

I am greatly indebted to Professor Ballantyne for the help he gave in connection with cases he had, and also for the numerous references to principal papers written on the subject; also to Professor Cohen, of Liverpool, who especially helped me in cases with so-called Gaisböck disease. Apart from the numerous references given, I should like to mention the admirable article on polycythaemia vera in the third volume of Duke-Elder’s Text-book of Ophthalmology.

Conclusion

(i) Polycythaemia is much more common than one thinks, but does not come so often into the hands of the ophthalmic surgeon.

(ii) Though the case is often severe, as seen in the case reported, the eye condition does return to normal.

(iii) It is justifiable to carry out operations, if required.

(iv) Care must be taken not to confuse this condition with that of a cerebral tumour, associated with headaches and papilloedema.
CASE OF POLYCYTHAEMIA VERA

REFERENCES


A CASE OF CORNEA PLANAS *  

BY  

MAJOR CHARLES COCKBURN  

R.A.M.C

FROM a perusal of the literature on the condition known as cornea plana, it is evident that this malformation is one of very considerable rarity—in fact, in English Literature only two descriptive papers have been published, Swett (1924), and Barkan and Borley (1936). The latter authors, in an exhaustive article, review the literature on cornea plana and add three cases occurring in one family—mother and two daughters. They discuss the diagnosis, emphasising the distinguishing features between the condition and microphthalmia, and the possibility of glaucoma as a complication. According to these authors, "cornea plana is that condition in which the radius of curvature of the anterior portion of the eyeball, specifically the cornea, is the same as that of the eyeball."

I believe that the case which I describe and illustrate photographically, is one of this rare malformation, and as such consider it worthy of record.

A Greek soldier, aged 28 years, reported to Eye O. P. D. of a General Hospital on Sept. 4, 1942, complaining of defective vision. An accurate history was difficult to obtain, but apparently his visual acuity had always been poor, although, significantly enough, he stated it had deteriorated since enlistment one year previously.

On examination, it was noted that both corneae were definitely smaller and considerably flatter than normal. R. cornea was clear; L. cornea had fairly dense central nebulae. In both eyes the corneo-scleral junction was ill-defined, the sclera seeming to encroach on the cornea. (Barkan and Borley speak of "indistinct corneal opacity extending from the corneo-scleral margin into the corneal tissue"). The anterior chambers were shallow.

Under cycloplegic.
A CASE OF POLYCYTHAEMIA VERA—EXTRACTION OF BOTH LENSES. SATISFACTORY RESULT*

J. Cole Marshall

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