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COMMUNICATIONS

FAILING VISION CAUSED BY A BONY SPIKE COMPRESSING THE OPTIC NERVE WITHIN THE OPTIC CANAL *

**REPORT OF TWO CASES ASSOCIATED WITH MORGAGNI'S
SYNDROME BENEFITED BY OPERATION**

BY

MURRAY A. FALCONER AND BRIAN E. PIERARD

*From the Department of Neurosurgery, Dunedin Hospital, and
Otago University, New Zealand.*

IN this paper we seek to draw attention to a hitherto undescribed condition, in which an optic nerve is compressed and indented by a bony spike projecting into the optic canal (optic foramen). We have encountered two cases, both of which were benefited by decompressing the optic nerve, and we desire to emphasize the need for recognizing the condition when it is present. The pathological condition underlying both our cases was Morgagni's syndrome. This is a disease of obscure aetiology, which occurs in middle-aged women, and is characterized by hyperostosis frontalis

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interna, obesity, and hirsuties, and sometimes by mental disturbances. It would seem that the bony spikes, situated in our two cases within the optic canals, were of similar origin to the spikes or osteophytes arising elsewhere from the inner table of the skull.

Case Histories

Case 1.—This patient, a 37-year-old single woman, was referred, in June, 1948, by Dr. R. P. Wilson and Prof. F. H. Smirk with a serious loss of vision in her right eye. Since childhood, she had been blind in her left eye as the result of

FIG. 1.—Case 1, general build.

FIG. 2.—Case 1, scalp incision and shaven face on 12th post-operative day.

an injury. Eight years ago she had noticed a slight dimness of vision in her seeing eye, particularly for distant landmarks, and this had remained stationary for six years. Then, during the last two years, her vision had become progressively more blurred, until she had difficulty in discerning even large objects. Throughout this latter period she had experienced attacks of "blacking out" of vision, which would appear without apparent reason and last a few seconds.

She also had several other symptoms. Since childhood, she had suffered from recurrent attacks of "acute articular rheumatism" in various joints, large and small, of her limbs. For ten years, she had complained of severe frontal headaches which appeared in attacks several times a month, and lasted from a few minutes to two days. Two years before admission, she had developed an abnormal thirst, drinking large quantities of water daily and passing correspondingly large volumes of urine, but this had since lessened. During this period of abnormal thirst, she had exhibited a disturbance of her sleep-rhythm, finding difficulty in sleeping by night. She had also complained of ringing noises in her right ear,

which was gradually becoming deaf. Her menses had commenced at the age of eleven years, but her periods had always been irregular and often overdue. Recently there had been an increased menstrual loss. At the age of sixteen years, an ovarian cyst had been removed, shortly afterwards she had rapidly increased in weight. For many years she had taken thyroid tablets 2 gr. daily to keep her weight down, but in spite of this her weight had remained at about 16 stone. Mentally she had always been normal, she successfully ran a second-hand shop, and was by all accounts an astute business woman. Her family history seemed uneventful, except that all the members were "large".

FIG. 3.—Case 1, visual fields charted 2 years before admission.

FIG. 4.—Case 1, visual fields charted shortly before operation.

Examination.—An alert and cheerful woman, of large and heavy build, with gross obesity involving mainly the trunk and proximal portions of the limbs (Figs 1 and 2). Height, 5 ft. 9½ in. Weight 16 st. 8 lb. She shaved daily, and exhibited hair on her chest, although the pubic hair distribution was feminine.

Ophthalmological Examination.—Visual acuity: right eye—4/36; J. 12 (corrected); left eye—blind. The right eyeball presented a low degree of myopia (−1.0 D.S.), its tension was normal, and its conducting media were clear. The optic disk was pale in its nasal half, but otherwise was normal. The retinal vessels were also normal.

Visual field charts taken at intervals during the two preceding years were available, and these showed that during this period her distance acuity had declined from 6/9 to less than 6/60. The decline in reading vision from J.2 to J.12 had been proportionately less, suggesting that an additional factor connecting with her myopia was also operating. The original chart showed constriction of all isopters of the right field, but chiefly on the temporal side and in the upper quadrant (Fig. 3). By the time of admission, this constriction had increased but no sector-shaped defect reaching the blind spot was detected (Fig. 4).

The left eyeball presented an old-standing traumatic cataract, and this was associated with a slightly divergent and concomitant squint. The movements of each eyeball were full. Pupils were approximately equal in size, and reacted to light and convergence.

Other Systems.—Apart from slight bilateral deafness, examination of the central nervous system resulted in negative findings. There was a slight goitre. The cardiovascular system also seemed normal (B.P. 125/85). Her fluid intake averaged 2700 to 4700 cc. *per diem*, while the urinary output averaged 1200 to 2700 cc. Transient swelling and crepitus were noted in both wrists and in the right ankle. Passive movements at the hip-joints were markedly reduced.

Radiological Examinations.—Views of the skull showed numerous osteophytes projecting inwards from the inner table of the vault in the frontal region (Fig. 5). These osteophytes were symmetrically arranged, and in places were confluent. In the midline they projected on either side of the superior longitudinal sinus almost enclosing it. The sella turcica appeared normal.

Views of the right optic canal revealed a blunt spike of bone projecting about 2 mm. into the lumen from its lower and inner part (Fig. 6). By contrast the left optic canal appeared round.

Lumbar Encephalography outlined a normal ventricular system. The basal cisterns filled normally with the air, and the silhouettes of the optic nerves in their intracranial course appeared normal and free from compression.

Plain x-rays of various joints and long bones showed no definite abnormality, although x-rays of the feet, taken four years earlier, had shown some lipping in the mid-tarsal joints. This lipping had since disappeared.

Laboratory Procedures:

- (a) Basal metabolic rate, −9 to −6 per cent.
- (b) Glucose tolerance test (50 g. glucose ingested).

Specimen	Blood sugar %	Urine sugar
Resting	0.16	Nil
½ hour	0.27	
1 hour	0.22	
2 hour	0.17	3 +

- (c) Lumbar cerebrospinal fluid—normal.
- (d) Blood count and sedimentation rate—normal.
- (e) Blood chemistry:

Serum calcium	-	9.9 mg. per 100 cc.
Blood cholesterol	-	330 mg. per 100 cc.
Blood uric acid	-	3.5 to 7 mg. per 100 cc.
- (f) Wasserman reaction—negative in both blood and cerebrospinal fluid.

FIG. 5 (a) and (b).—Case 1, X rays of skull showing extensive hyperostosis frontalis interna.

Summary.—A progressive failure of vision had occurred, the visual acuity having fallen from 6/9 to 4/36 in the preceding two years. X rays had disclosed a bony spike projecting into the right optic canal in its lower and inner part, and the position of this spike could be correlated with the site of maximum impairment of the visual field in the upper temporal quadrant. In the absence of any other demonstrable cause, this spike was therefore presumed to be the lesion responsible for the visual failure. As the other eye was blind, and the visual loss in the seeing eye had reached a very serious level, exploration of the optic nerve within its canal was decided on.

FIG. 6.—Case 1, X ray of right optic canal showing bony spike projecting into canal from its lower and inner part.

Operation.—Under intratracheal anaesthesia, a right frontal craniotomy of the type used for a subfrontal pituitary exposure was performed (M.A.F.). Difficulty was encountered in separating the skull from the underlying dura because of osteophyte-formation. The dural envelope surrounding the frontal lobe was elevated off the anterior cranial fossa, and was incised in front of the sphenoidal ridge, exposing the intracranial portion of the right optic nerve and the chiasm. No abnormality was here seen. Next, a burr-hole was made in the orbital plate of the frontal bone, and from this starting point the roof of the orbit was removed piece by piece in a backward direction towards the optic canal. Then, the wall of the canal itself was removed on its superior and lateral aspects, so

decompressing the optic nerve (Figs 7 and 8). It was not possible to gain access to the floor of the canal below the optic nerve, and so the actual bony spike was not disturbed. After haemostasis was secured, the craniotomy was closed.

FIG. 7.—Diagram indicating amount of bone removed from anterior cranial fossa to decompress the right optic canal.

Post-operative Course—Convalescence was uneventful. Three weeks later, when she was discharged from hospital, her visual acuity had improved to 6/12 and J.6, while her visual field had filled out except for a sector-shaped defect reaching the blind spot from the upper temporal quadrant (Fig. 9). Shortly afterwards, Dr. Wilson reported that her acuity had reached 6/4 partially, and had then deteriorated slightly.

Later Progress.—When seen again by us 14 months later, she said that, apart from a slight and transient impairment which occurred shortly after her return home and was corrected by a change of spectacles, the vision in her right eye had remained good. She could see to read a newspaper, sew, and drive a motor-car, and had resumed all her former activities. Headaches had been slight.

FIG. 8.—Assistant's sketch illustrating operative exposure. The right frontal lobe has been elevated, and the optic canal decompressed. Notice the optic nerve freed within the canal.

Her menses were still irregular. On examination her visual acuity was 6/9 partially and J.4 (Fig. 10). The optic disk was now definitely pale while the visual field showed no essential change from that charted at the time of discharge, although the limits of the sector-shaped defect were perhaps more definite.

FIG. 9.—Case 1, visual fields charted 3 weeks after operation.

FIG. 10.—Case 1, visual fields charted 14 months after operation.

Case 2.—This patient, a 48-year-old single woman, was referred, in March, 1949, by Drs. W. J. Hope-Robertson and A. D. S. Whyte. For four months she had noted a slight but apparently progressive impairment of vision in her right eye. During the last two months she had found herself unable to thread a needle in poor light, and more recently she had become aware of a streak of darkness in the vision of her right eye, which was continuously present above and to the nasal side of the fixation-point. She was, however, still able to read a newspaper with either eye without spectacles.

A year previous to admission she had suddenly developed a severe headache in the right frontal region, which had lasted two days. Simultaneously she had

FIG. 11.—Case 2, general build.

FIG. 12.—Case 2, scalp incision on 14th post-operative day.

noted a "black eye" on that side, as though she had been bruised, but there had been no injury. Since her vision had become impaired, further but milder attacks of frontal headache had occurred. Apart from an attack of jaundice during her late teens, previous illnesses had consisted only of childhood fevers. Since adolescence she had always been of large build. Her menses had been normal until three months previously, when they had become irregular and

associated with menorrhagia. There was a history of rheumatism in her right wrist, but no history of thirst or polyuria. The family history seemed uneventful.

Examination.—A stout woman with a big skeletal frame, who was alert and of normal intelligence. Again the obesity was pronounced over the trunk and proximal portions of the limbs (Figs. 11 and 12). Height, 5 ft. 9 in. Weight, 18 st. There was marked hairiness of her legs and a masculine type of pubic hair distribution. There was no hirsuties of the face.

Ophthalmological Examination.—Visual acuity: right eye 6/9, J.1; left eye, 6/9, J.1. Both eyeballs were emmetropic, the intraocular tensions normal, and

the conducting media clear. In the right eye there was some pallor of the temporal half of the disk, but the retina and the retinal vessels were normal. The left fundus was entirely normal.

The visual field of the right eye showed a sector-shaped defect situated immediately above the horizontal meridian in the upper nasal quadrant, and reaching to the blind spot (Fig. 13). There was a smaller and less distinct

FIG. 13.—Case 2, visual fields charted before operation.

defect below the horizontal meridian. The appearances were those of a nerve-fibre bundle defect.

The left visual field was normal. The eyeball movements were full, and there was no squint or nystagmus. The pupils were equal and they reacted briskly to light and convergence.

Other systems.—No abnormality was detected during the examination of the nervous system, or of the heart and lungs (B.P. 150/100). Her joints were supple, and she had no goitre. The fluid balance was normal.

Radiological Examination.—Views of the skull showed a few broad-based osteophytes projecting from the inner table on either side of the superior longitudinal sinus in the frontal region (Fig. 14).

Views of the right optic canal showed a sharp spike 3 mm. high projecting into its lumen from the outer wall just below its horizontal equator (Fig. 15). The left optic canal was completely circular.

X rays of several joints and limb bones were also normal.

Laboratory Procedures:

- (a) Basal metabolic rate—+9 to +11 per cent.
- (b) Glucose tolerance test—normal, no glycosuria.
- (c) Lumbar cerebrospinal fluid—normal.
- (d) Blood count—normal.

- (e) Blood sedimentation rate—36 mm. for first hour (normal 3 to 10 mm.).
- (f) Serum calcium—8.5 mg. per 100 cc.
- (g) Wasserman reaction—negative in blood.

FIG. 14.—Case 2, X ray of skull showing some broad-based osteophytes projecting intracranially in frontal region.

FIG. 15.—Case 2, X ray of right optic canal showing fine spike projecting into centre of canal from lateral wall just below the equator.

Summary.—The clinical picture in this patient presented many resemblances to that of the first patient. A diagnosis of glaucoma had been suggested by the visual-field changes, but had been ruled out by repeated observations of the intraocular tension, which had always been normal. Although the visual acuity of the right eye was still normal, the visual impairment was progressive, and, as in Case 1, the position of the visual-field defect corresponded nicely with the position of the spike within the optic canal. Furthermore, the same salient features of Morgagni's syndrome were present, although in different degree.

As Case 1 had benefited by decompression of the optic nerve, a similar operation was decided on to prevent further visual loss.

Operation.—A right frontal craniotomy was performed, similar to that in Case 1 (M.A.F.). In this patient, however, after the roof of the optic canal had been removed, the lateral wall was undermined on its outside, and then broken outwards in one piece away from the nerve (Fig. 16). The bony fragment thus obtained included the actual spike which had projected into the nerve (Fig. 17).

FIG. 16.—Case 2, post-operative X ray of optic canal indicating amount of bone removed. The location of the canal is completed with a series of white dots.

FIG. 17.—Case 2, bony fragment removed at operation from lateral wall of optic canal ($\times 5.7$).

Post-operative Course.—Convalescence was uneventful except that, for about two weeks, the patient had a ravenous appetite, without obvious excessive thirst. She was discharged from hospital on the seventeenth day, and about this time the scotomatous defect had lessened at the periphery of the field, and she was no longer aware of a streak of darkness in the vision of her right eye (Figs. 18 and 19).

Later Progress.—When seen again four months later, she stated that her vision remained good, and that she was aware of the dark streak only when tired. She had resumed her normal life. Dr. Hope-Robertson reported having observed an improvement in his visual-field charts from before operation similar to that shown by ours, but no further improvement of vision seemed to have occurred after the first month. Six months after operation the patient confirmed by letter that her vision remained improved.

Discussion

In both cases the evidence indicates that the visual failure was due to compression of the optic nerve by a bony spike. First we have the correlation in each case between the position of the visual-field defect and the position of the spike within the optic canal. Secondly, benefit followed surgical relief of the compression, for not only was the deterioration of vision halted, but in both cases a slight improvement in vision actually occurred.

Cases of failing vision due to concentric compression of an optic nerve within the optic canal have been described in such conditions as osteitis deformans, osteitis fibrosa, and leontiasis ossea,

where the optic canal may become involved in the bony disease (Falconer and Cope, 1942). One of us has recently reported a case, in which an optic nerve was compressed within the optic canal by a depressed spicule of bone projecting from a fracture

FIG. 18.—Case 2, visual fields charted 12 days after operation.

FIG. 19.—Case 2, visual fields charted 3 weeks after operation.

line; an improvement in vision followed decompression of the optic nerve (Falconer, 1949). A search of the literature, however, has failed to disclose any case-reports of optic-nerve compression by a bony spike developing *sui generis*, as in the two cases described in this paper. The development of the bony spikes in our two patients is probably accounted for by the fact that they both suffer from Morgagni's syndrome. The clinical features of this condition might here be conveniently reviewed.

Review of Morgagni's Syndrome

The triad of Morgagni's syndrome consists of hyperostosis frontalis interna, obesity, hirsuties; in some patients neuropsychiatric symptoms are also present. The aetiology of the condition is obscure, but most writers favour a disturbance of the hypothalamus-pituitary mechanism. Morgagni's original description, based upon a single case studied at autopsy, was published in 1765, but recognition of the syndrome has come mainly from the papers of Stewart (1928) and of Morel (1930). The condition is almost entirely confined to the female sex, about 98 per cent. of sufferers being women (Moore, 1935). It is uncommon under the age of 40 years, and occurs chiefly in the 50- to 60-year age-group. Statistics vary as to its frequency, but Pederson (1947) observed hyperostosis frontalis interna in 2 to 3 per cent. of skulls of 485 women *x*-rayed for head injury. Moore (1936) observed the condition in 1.44 per cent. of 6,650 skulls examined radiologically, but his material included both sexes. A higher incidence of this condition is probably met with in mental-hospital inmates.

Bony Changes.—These are chiefly confined to the frontal bone, and tend to be symmetrical. Osteophytes project from its inner table indenting the dura. They consist of dense cancellous bone covered by a layer of compact bone, and as development continues the diploe gradually becomes sclerosed. The outer table is never affected, and the contour of the skull remains unchanged. As the process advances, more and more osteophytes appear, and spread out fanwise from the falx and superior longitudinal sinus, the attachment of which is usually spared. Most authors appear to consider that, apart from the vertical and horizontal portions of the frontal bone, the rest of the skull is usually spared, although Moore (1935), recording a proportion of cases with seventh and eighth cranial nerve symptoms, suggested that the petrous bone may sometimes be involved. Our two cases indicate that osteophytes may also occur in the optic canal (sphenoid bone). The skeleton, outside the skull, is not involved.

Other Endocrine Disorders.—Obesity is seen in most cases. It involves mainly the trunk and proximal portions of the limbs, sparing the extremities, as it did in our two patients.

Hirsutism, rather than true virilism, is a prominent endocrine feature. It consists of an overgrowth of hair especially on the face, but without characteristic male hair-distribution, clitoral enlargement, or other stigmata of masculinization.

Menstrual disorders are common. Thyroid disorders are reported in about 5 per cent. of cases (Carr, 1936). Decreased sugar tolerance is not infrequent. Polydipsia, polyuria, and polyphagia were reported by Morel (1930), but are usually considered rare, although they were observed in our patients. Hypertension and arteriosclerosis seem to occur in a large proportion of cases.

Neuropsychiatric Symptoms.—These are often absent, but since the publication of the papers of Stewart (1928) and Morel (1928), both dealing with mental-hospital patients, a great deal of attention has been paid to nervous and mental manifestations. These may be merely frontal headaches, associated perhaps with memory defects and nervousness, but almost every form of psychosis, as well as epilepsy, has been encountered in or attributed to the syndrome, sometimes without adequate justification. Such features, apart from headache, were absent in our two cases.

Ophthalmological Aspects.—Few authors mention the question of visual failure in their case-material. Van Bogaert (1930), however, records a case of Morgagni's syndrome in which there was bilateral visual failure with optic atrophy; he presumed this to be due to pressure on the optic nerves within the optic canals, but the optic canals were evidently not demonstrated radiologically. Moore (1935) noted visual symptoms in thirteen out of 59 cases in which case-histories were available, but the tabulated summaries are insufficient for proper evaluation of the causes of visual failure. Moore, however, advised that detailed radiological studies of the skull, presumably including the optic canals (optic foramina), should be made in such cases, although he himself did not mention any specific instance. Carr (1936) mentioned a 41 per cent. incidence of visual symptoms, but did not amplify this statement. Since then several other authors have recorded in passing an occasional case with visual failure (Eisen, 1936; Andrews, 1942).

Conclusion

Our two patients thus presented all the requisite features of Morgagni's syndrome (hyperstosis frontalis interna, obesity, and hirsuties) although neither had obvious psychiatric disorders. They would appear to be the only cases of the syndrome at present recorded, in which the visual failure was shown to be due to compression of an optic nerve by a bony spike or osteophyte situated within the optic canal.

It seems likely that many cases of this condition must have been missed in the past, and consequently gone unrelieved. In skilled hands, the surgical decompression of the optic nerve should carry a negligible mortality, and should improve the prospects of sufferers with this condition.

Our two cases also illustrate the value of quantitative visual-field studies and of careful radiography of the optic canals (foramina) in all cases of failing vision, with or without optic atrophy, where the cause of visual failure is obscure (see also Falconer, 1949). The demonstration of a bony spike in the optic canal, the position of which can be correlated with the position of a progressive visual-field defect, is sufficient justification for surgical intervention. Our second case indicates that, if the best results are desired, such intervention should not be delayed until gross visual failure has occurred. The chances of restoring normal vision presumably become less the longer the compression has been in existence.

Summary

Two cases of unilateral visual failure are reported, both due to compression of an optic nerve by a bony spike situated within the optic canal. Both were benefited by decompressing the optic nerve, and, in one case, the bony spike was removed as well. In one instance the improvement of vision was from less than 6/60 to 6/9.

Both cases were examples of Morgagni's syndrome, and the salient features of this syndrome are outlined. The bony spikes within the optic canals had presumably a similar origin to the bony spikes or osteophytes arising elsewhere from the outer table of the skull.

A plea is made for early surgical intervention in similar cases in order to preserve vision. The value of quantitative perimetry and of careful radiography of the optic canals in the recognition of such cases is also stressed.

We are most grateful to our ophthalmological colleague, Dr. Rowland P. Wilson, for his assistance in both cases. In Case 1 the diagnosis of optic-nerve compression by a bony spike was first made by him, and he suggested that we should decompress the optic nerve. We are also grateful to Dr. A. C. Begg for the radiological studies, and to Mr. E. R. Macdonald for the reproductions.

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