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Arachnoid hyperplasia in optic nerve glioma

SIR, In the September 1979 issue of the *BJO* Cooling and Wright¹ have attempted to account for the curious fact that in their experience only 1 of 20 patients with meningiomas of the optic nerve was a child, whereas from the combined case material in the Registry of Ophthalmic Pathology of the Armed Forces Institute of Pathology, and at the University of California, Karp *et al.*² reported that 7 of their 25 cases involved children who were 11 years old or younger, and 10 patients (40%) were younger than 20 years. Cooling and Wright offer as their explanation for this difference in experience the possibility that the pathological diagnoses in the paediatric cases of Karp *et al.* were incorrect. They base this suggestion on their own unfortunate experience in having had to operate on a child 3 times before the correct diagnosis of glioma was established.¹ Biopsies at the first 2 operations had led to a diagnosis of meningioma, but in fact it was subsequently shown that arachnoid hyperplasia associated with a glioma had been mistaken for meningioma. The Editor of the *BJO* was stimulated to prepare an editorial on the subject which was carried in the same issue.³

That exuberant meningeal hyperplasia may be observed in association with gliomas of the optic nerve is a fact that has been well appreciated by ophthalmic pathologists for over half a century. Two of the authors of the original paper in question (Zimmerman and Spencer) have written on this subject⁴⁻⁷ and have for many years in their lectures cautioned about making this error in pathological diagnosis. Curiously, Cooling and Wright even cited some of these publications^{4,5} in support of their argument, including the fact that Hogan and Zimmerman⁴ pointed out that there is 'the possibility of diagnostic confusion between arachnoid (sic) hyperplasia accompanying a glioma, and orbital meningioma'.

To document further our concern about correct histopathological interpretation I would like to take this opportunity to comment on another strange case report that appeared in the *BJO* in 1977.⁸ The authors interpreted their tumour as a mixed meningioma and astrocytoma, though judging from their published illustrations I suspected they had misinterpreted arachnoid hyperplasia for meningioma. At my request the senior author kindly permitted me and my colleagues to review a histological section of this tumour, which confirmed to our satisfaction that the 'meningioma' was in fact arachnoid hyperplasia. We are therefore well aware of the possibility for such errors in histopathological interpretation, but they are not attributable to the pathologist's absence from the surgeon's side in the operating room.

The glib implication that in the series by Karp *et al.*² the pathological diagnoses must have been made by inexperienced pathologists so removed from the surgical scene that they couldn't possibly make the correct diagnosis cannot be allowed to stand without challenge.

Any pathologist who has had any experience in the differential diagnosis between arachnoid hyperplasia and meningioma should recognise at a glance that the illustrations contained in the paper by Karp *et al.* (their Figs. 1 through 6)² cannot be accounted for by hyperplasia. It is customary among pathologists and clinicians of mature judgment to request an opportunity to review histopathological preparations before publishing such allegations as Cooling and Wright have done. Karp and his colleagues have never been asked to give Cooling or Wright an opportunity to study sections of the tumours from their paediatric cases. There was no correspondence relative to this problem between Cooling and Wright, on the one hand, and Karp *et al.* on the other.

In retrospect it is unfortunate that we did not include in our paper a section on differential diagnosis between meningeal hyperplasia and meningioma. This experience will serve as a stimulus for a more detailed future report on the subject, but for the present I would like to point out some basic differences that even surgeons who are not knowledgeable about ophthalmic pathology should be able to appreciate. (1) In arachnoid hyperplasia the proliferation remains intradural, whereas in most meningiomas there is invasion through the dura into the surrounding orbital tissues, as was so clearly evident from Figs. 1 and 4 in the paper of Karp *et al.* (2) Orbital recurrences with or without intracranial involvement are common complications after operations on meningiomas of the optic nerve in children (one of the main points of the paper by Karp *et al.* ignored by Cooling and Wright), but this never (in our experience) occurs with meningeal hyperplasia. (3) Meningiomas are potentially lethal tumors, especially in children, whereas meningeal hyperplasia is not.

To document further and augment the validity of the data presented by Karp *et al.* I can, through the courtesy of Dr Melvin G. Alper,⁹ who restudied and updated the Registry case material last year in preparation for his Francis Heed Adler lecture on orbital meningiomas, give some additional follow-up information on the paediatric cases in Karp *et al.*'s series:

Case 2. This patient, who was 6 years old at the time of enucleation and whose massive meningioma was illustrated in Fig. 4, had further recurrences with intracranial involvement and died at age 31.

Case 3. This child, who was 6 years old at the time of initial surgery, was the only prepubertal child who was living without recurrence at the time of the initial report. However, she developed an orbital recurrence and died of a viral infection at age 13. Invasion of the eye was also observed postmortem.

Case 8. Five years after the initial excision of the meningioma this patient had an exenteration followed by a transcranial exploration because of a recurrence of the tumour.

The aggressiveness of these meningeal tumours in young patients clearly reflects the fact that they are truly neoplastic and not merely reactive processes as suggested by Cooling and Wright.

In summary, the clinical course and the typical histopathological features of the paediatric tumours reported

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by Karp and coworkers establish beyond reasonable doubt that these were meningiomas, not gliomas with meningeal hyperplasia. While we have no explanation for the almost total absence of meningiomas in British children, I can report that since the publication of Karp *et al.* we have seen additional cases in the United States. Messrs Cooling and Wright must seek a better explanation for their different experience.

The opinions or assertions contained herein are the private views of the author and are not to be construed as official or as reflecting the views of the Department of the Army or the Department of Defence.

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SIR, We were very interested to read Professor Zimmerman's comments about our paper. The purpose of our communication was to highlight the difficulties facing the pathologist receiving histopathological material without adequate clinical information. Our presumptive clinical diagnosis was an optic nerve glioma, when the child detailed in the case report was first seen. However, when the normal intracranial portion of the optic nerve with its surrounding abnormal tissue was examined by an experienced neuropathologist, the neurosurgeon failed to inform him that the main part of the tumour lay within the orbit. The pathologist incorrectly diagnosed a primary optic nerve meningioma.

We fully accept Professor Zimmerman's statement that all the patients reported by Karp and his associates¹ were meningiomas. His expertise in this and other fields is unquestioned, and we acknowledge that he has selected patients with tissue that he has diagnosed correctly as meningioma. We therefore withdraw our statement that some of his patients may have had arachnoid hyperplasia remote from an unsuspected glioma.

However, the reported incidence of primary optic nerve meningioma in childhood is very low in all recently reported series of this lesion²⁻⁵ except that of Professor Zimmerman and his colleagues. There are 2 possible explanations which may account for this discrepancy. (1) The 10 children they recorded all had a primary optic nerve meningioma. It is possible that the increased incidence may be due to selection. They reported 21 specimens out of a total of 81 dating back to 1927. (2) All patients had a meningioma but a proportion of these were secondary in origin, i.e., there was extension into the orbit from the middle or anterior cranial fossa. This is a distinct possibility for, no matter how expert the pathologist, he is dependent on the accuracy of clinical information supplied by surgeons in centres of differing excellence, often using the primitive diagnostic facilities in an earlier era. Therefore one cannot be absolutely sure of the origin of the meningiomatous tissue in order to correctly classify the meningioma as a primary optic nerve tumour.

The expertise of Professor Zimmerman and his associates is beyond question, but they have in many cases been dependent on clinical details sent them with the excised specimen. As an example of this, a recent report from the A.F.I.P.⁶ detailed their experience of optic nerve gliomas. The authors found that the incidence of associated neurofibromatosis was only 10%, one of the lowest reported in any series. Their explanation was that the low incidence was probably due to the inadequate clinical information provided by those referring the specimens to the A.F.I.P.

Our experience and that of others differs from that of Professor Zimmerman and his coworkers. During the past 5 years there has been 1 case of primary optic nerve meningioma occurring in a child in the combined clinical London experience of Moorfields Eye Hospital, the Hospital for Sick Children, Great Ormond Street (David Taylor, personal communication), and the National Hospital for Nervous Diseases (W. I. McDonald, personal communication). During the same period 12 primary optic nerve meningiomas were seen in adults.

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Obituary

Hugh Campbell Orr, MB, ChB, FRCP(Glas)

Hugh Campbell Orr, formerly consultant ophthalmic surgeon to Wolverhampton and Midland Counties Eye Infirmary, died on 30 April aged 81. He was born at Hamilton, and during the first world war enlisted in the Royal Artillery, serving 3 years in France. He graduated in medicine from Glasgow University in 1924 and shortly afterwards was appointed house surgeon to Wolverhampton Eye Infirmary. He had originally intended to take up neurology, but Ophthalmology claimed him and he never regretted his change in direction. He was appointed assistant surgeon in 1925 and, after qualifying FRFPS (Glasgow), honorary surgeon in 1934. His desire for progress frequently led him into dispute with the governing council of the hospital, but the force of his argument and his determination usually won the day. With the introduction of the National Health Service he became consultant surgeon, and after his retirement from the service in 1963 he continued in his private practice, school clinics, and general ophthalmic services work.

His professional career spanned years in which tremendous changes occurred in ophthalmology, with progressive refinement of surgical instruments and technique, and it says much for his ability that he always kept abreast of these developments, maintaining that surgeons could not mark time, only go forwards or backwards. His early interests were in industrial injuries and retinal colour photography, but in later years he became more interested in vascular and degenerative diseases of the retina and was a firm believer in the value of vasodilator drugs. Greatly troubled by the increasing number of patients referred with senile macular degeneration, against current thought he foresaw the possibility that it might eventually yield to treatment and merited thorough investigation. Although he did not have the facilities to do research into the condition himself, he encouraged others to do so and the renewed interest of recent years gave him considerable satisfaction.

During the postwar years he travelled much in Europe

and was held in high regard by many ophthalmologists whose clinics he visited. Apart from the strictly formal rounds on these occasions he valued as much the free discussions on ophthalmic matters and interchange of ideas during the social gatherings. He was never afraid to defend his often controversial views, but after a hard-fought disputation would often wryly quote from his favourite Omar Khayyam: 'Myself when young did eagerly frequent, Doctor Saint, and heard great argument About it and about; but evermore Came out by the same door as in I went'.

He had a reputation as a strict disciplinarian towards both his patients and his staff, but he took great pains to teach surgery to his juniors, and although they went in daily fear of his disapproval he and Mrs Orr were charming and generous hosts when they entertained successive house surgeons and registrars to an evening's relaxation at the theatre at Stratford-on-Avon. His work dominated his whole life, and practically his only leisure activity was reading, but in later years much time was devoted to his only daughter, disabled by chronic illness, whose death came only a few weeks before his own.

For many years he was the secretary of the Staffordshire Branch of the BMA. He was president of the Midland Ophthalmological Society in 1947–9 and in 1971 served as deputy master of the Oxford Congress, where he was a familiar figure.

The theme of Hugh Campbell Orr's life was to work and to serve. He served the local community well as an ophthalmologist and his name was a byword in Wolverhampton; and he worked until only 5 days before his death—truly a life-time of dedication to his calling.

B.R.K.

Notes

Courses at Institute

The following courses will be held at Institute of Ophthalmology: Revision course in eyelid and lacrimal surgery, 27–29 October 1980 (fee £50). Recent major advances in the management of ocular trauma, 26–28 November 1980 (fee £75). Indications and techniques in the ophthalmic management of retinovascular disease, 1–3 December 1980 (fee £75). Applications to the Dean's Office, Institute of Ophthalmology, 17–25 Cayton Street, London EC1V 9AT.

Laser workshop

A laser workshop for ophthalmic diseases will be held at the Singapore General Hospital on 26–28 September 1980. It is organised by the Academy of Medicine, Singapore, and the Australian College of Ophthalmologists. Details from Laser Workshop, 606–610 Singapore Polyclinic, Specialists' Centre, Orchard Road, Singapore 0923.