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Introduction

This volume is primarily composed of selected papers reported at the International Migraine and Headache Symposium held in Florence on May 25th to 27th, 1970. The participants of the Symposium are a distinguished group of international investigators working in the field of headache. These papers have been published with minimal editorial changes to preserve the style and thought of the authors.

Three papers not presented at the Symposium have been added to complete this Volume: 'Investigation of Headache', by E. GRAEME ROBERTSON; 'Cluster Headache', by JOHN M. SUTHERLAND and MERVYN J. EADIE; and 'Psychology and Neuropsychology in Migraine', by GERHARD S. BAROLIN.

We hope that this collection of papers will provide not only practical help in the clinical aspects of the headache problem but also knowledge concerning new areas of investigation in the pathogenesis of migraine and headache. I wish to thank the authors and particularly Professor SICUTERI, whose efforts made this volume possible.

ARNOLD P. FRIEDMAN, M.D.
Editor-in-Chief
Research and Clinical Studies in Headache

Contents

Contents

International Migraine-Headache Symposium
May 26–28, 1970, Florence (Congress Palace)
Selected Papers

Res. clin. Stud. Headache, vol. 3, pp. 1–91 (Karger, Basel 1972)

The Investigation of Headache

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Contents

Introduction

The clinical conduct of a patient complaining of headache, sometimes calls for skill in the diagnosis of an intangible, isolated symptom, while at other times the diagnosis depends, not on headache alone, but on other symptoms and signs. The cause of headache may be otherwise innocuous, or it may be serious and even lethal; hence diagnosis entails great responsibility.

The primary need is, of course, to obtain as much information as possible from the patient, his relatives and from his general practitioner. In many instances the psychological background is important; this information sometimes being crucial in diagnosis, and helpful in sustaining the patient's morale during a serious illness.

Headache as an Isolated Symptom

Paradoxically a dissertation upon investigation should also indicate when not to investigate.

The investigation of headache, without other symptoms and signs, is rarely rewarding, for by far the commonest cause of isolated headache is a psychogenic one. The simple statement that most people suffer from headache, often as the result of simple stresses (such as shopping in a crowded city) is often a tangible commencement to therapeutic discussion during the interview. Reassurance is essential; evading the issue, harmful.

The diagnosis of tension headache is reasonable, if tension is used to denote a state of mind which may lead to tension of the frontalis and cervical musculature. Anxieties, conflicts, noise, fatigue and undue stresses may precipitate headache in the predisposed. Some forgotten cephalic sensation may direct attention to the head and establish a vicious cycle compounded of increasing over-sensitiveness, anxiety and awareness of the head, conveying a conviction of abnormality. In some instances the symptomatology is so bizarre that diagnosis is easy; in some it is so difficult that the practitioner may have to decide to take whatever risk there is of being wrong and reassure the patient.

Psychogenic headache is much commoner than headache due to an organic complaint, but occasionally headache is the first symptom of organic disease. In patients in whom suspicion exists, innocuous radiography of the skull, radio-isotope scanning and electroencephalography may be used to lessen the risk of misdiagnosis. When an accurate diagnosis of the cause cannot be made at the time, and must be postponed, the risk of unassuaged anxiety may be lessened by the reassurance which normal investigations may convey. Observation may be enjoined, and this may give chosen patients a sense of security. In other patients it may be best to warn a close relative of the need for observation. Obviously decisions can only be made when all circumstances are understood, and the medical practitioner's own personality will to some extent modify his decisions.

When, in 1788, JOHN WHITE, Esq., Surgeon General to the Colony of New South Wales joined the First Fleet at Spithead he wrote these words in his Journal, which I quote in support of the above advice:

> When I got on board the Alexander, I found there a medical gentleman from Portsmouth amongst whose acquaintance I had not the honour to be numbered. He scarcely gave me time to get upon the quarter-deck before he thus addressed me—'I am very glad you are arrived, Sir; for your people have got a *malignant* disease of the most dangerous kind; and it will be necessary, for their preservation, to get them immediately relanded!' Surprised at such a salutation, and alarmed at the purport of it, I requested my assistant, Mr. Balmain, an intelligent young man, whom I had appointed to this ship for the voyage, to let me see the people who were ill. 'Sir', returned Mr. Balmain, taking me aside, 'you will not find things by any means so bad as this gentleman represents them to be; they are made much worse by him than they really are. Unlike a person wishing to administer comfort to those who are afflicted, either in body or in mind, he has publicly declared before the poor creatures who are ill, that they must inevitably fall a sacrifice to the malignant disorder with which they are afflicted; the malignity of which appears to me to exist only in his own imagination.' Mr. Balmain had no sooner concluded than I went between decks, and found everything just as he had represented it to be. There were several in bed with slight inflammatory complaints; some there were who kept their bed to avoid

the inconvenience of the cold, which was at this time piercing, and whose wretched clothing was but a poor defence against the rigour of it; others were confined to their bed through the effects of long imprisonment, a weakened habit, and lowness of spirits; which was not a little added to by the declaration of the medical gentleman above mentioned, whom they concluded to be the principal surgeon to the expedition. However on my undeceiving them in that point, and at the same time confirming what Mr. Balmain had from the first told them, viz. *that their complaints were neither malignant nor dangerous,* their fears abated. This short conversation had so sudden an effect on those I addressed ... that before we got from between decks I had the pleasure to see several of them put on such clothes as they had, and look a little cheerful. On returning to the quarter-deck, I found my new medical acquaintance still there, and ... he thus once again addressed me—'I suppose you are now convinced of the dangerous disease that prevails amongst these people, and of the necessity of having them landed, on order to get rid of it.' Not a little hurt at the absurd part this gentleman had acted, and at his repeated importunity, I replied with some warmth that I was very sorry to differ so essentially in opinion from him, as to be obliged to tell him that there was not the least appearance of malignity in the disease under which the convicts laboured, but that it wholly proceeded from the cold, and was nearly similar to a complaint then prevalent, even among the better sort of people in and about Portsmouth.

Notwithstanding this he so persisted that I could no longer keep my temper; and freely told him that the idea of landing them was as improper as it was absurd ... And, to make him perfectly easy on that head, I assured him that when any disease rendered it necessary to call in medical aid, he might rest satisfied that I would not trouble him, but would apply to Doctor Lind, Physician to the Royal Hospital at Haslar, a gentleman as eminently distinguished for his professional abilities as for his amiable qualities, or else to some of the surgeons of His Majesty's ships ... most of whom I had the pleasure of knowing, and on whose medical knowledge I was certain I could depend.

This peremptory declaration had the desired effect. The gentleman took his leave, to my great satisfaction.

Radiography

Localized pain is usually the result of local pathology, which may be demonstrated by radiography. A common cause is purulent infection of the nasal accessory sinuses. In these instances localized pain, with perhaps purulent discharge, usually establish the diagnosis although the latter may be absent e.g. in 'vacuum' headache due to block of the ostium of the frontal sinus. In headache due to infection of sinuses, or mastoid air cells, radiography usually establishes the diagnosis. Paget's disease is recognized by radiography alone, and many instances might be recorded in which radiography provides important diagnostic information.

A rare cause of localized pain quickly becoming generalized is reported:

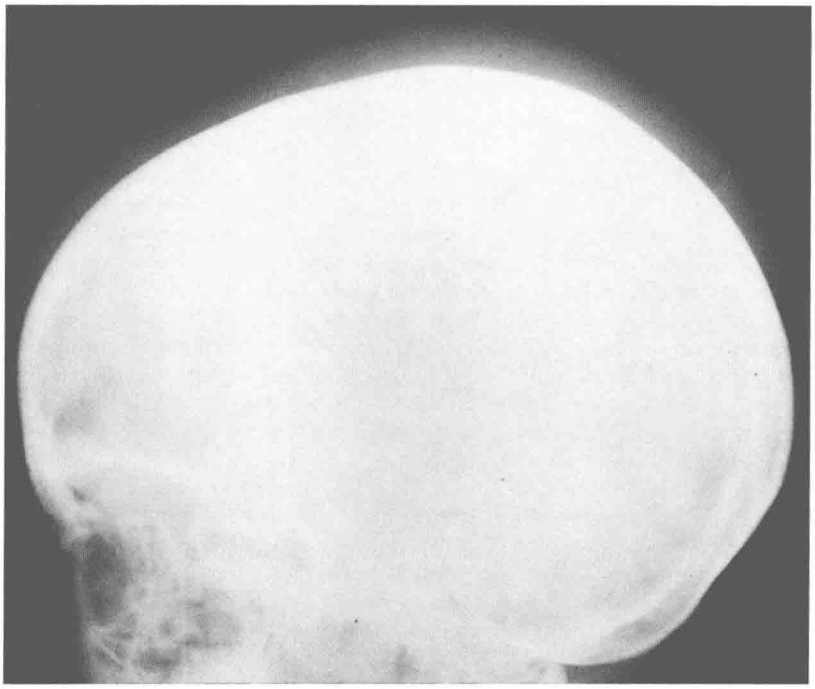

Fig. 1. Case 1: subgaleal haematoma, shown by X-rays of low penetration.

Case 1. A child of seven years complained of right temporal headache 7 days before admission to the Royal Children's Hospital, Melbourne. Two days later a large swelling 7,5 cm in diameter appeared in the same situation and within a further 3 days a fluctuant swelling spread all over the head, limited anteriorly by the supraorbital ridges and posteriorly by the superior nuchal lines. The headache also became generalized.

In 1959 she had experienced attacks of suprapubic pain for about 5 months, and 1 month before the onset of the present headache a knock to the right shin led to a large haematoma. She had always bruised readily. Two of her brothers suffered from congenital spherocytosis.

Radiography of the skull showed a diffuse soft tissue shadow external to bones of the cranial vault which were of normal appearance, (fig. 1). The suspicion of intermittent coagulopathy (von Willebrand's disease) prompted the fullest blood investigation, but no abnormality was found. The circumference of the head remained the same for a week and then it began to decrease, with palpable organization commencing posteriorly.

Subgaleal haematomas are usually caused by trauma in which movement of the scalp is thought to shear blood vessels. It may be treated by aspiration, but this case shows that spontaneous resolution may occur, as with other forms of haematoma.

Examination of Cerebrospinal Fluid

Lumbar puncture alone may be sufficient for full diagnosis, as in meningitis.

Case 2. In 1942 Mrs. A. J., a housewife of 30 years, began to suffer from severe bouts of headache, which lasted from 10 to 21 days and recurred at intervals of 1 to 6 months. The headache seemed to start in the eyes and extend through the eyes to the back of the head, and thence down the neck and up to the vertex. She had many domestic worries, which she thought tended to produce the headache.

She was first seen on 16 November 1950 during a bout of headache which had lasted for 6 weeks. It was the same type as before, but more severe, and she added that it seemed like a skull-cap made of lead. She felt nauseated and, on several mornings, had vomited. She also noticed a roaring noise in her ears, and stooping would increase the headache and roaring. During the latter part of this period she saw double, and found it a little difficult to walk straight.

No abnormality was found on examination. The blood pressure was 120 systolic, 80 diastolic. The pressure of the cerebrospinal fluid was normal and the fluid contained 120 lymphocytes, 14 polymorphs and 2 large mononuclear cells per mm^3. The protein content was 100 mg%, with increased globulin. Sugar was estimated at 0.06 mg%; chlorides, 670 mg%. The colloidal gold curve was 0000144444 and the Wassermann reaction was negative. No torulae were found in this and in two subsequent specimens but on 5 January 1951 cryptococcus neoformans had grown on Sabouraud's medium.

In the meanwhile, radiograms of the chest showed a spherical mass, 5 cm in diameter, lying medially in the left infraclavicular region. Cryptococcus neoformans was seen microscopically in, and on culture of, the mucopurulent sputum. The EEG showed 6–7 per sec waves of variable voltage, with reversal of phase between the left parietal and occipital regions.

On 12 January 1951 the posterior segment of the left upper lobe was removed and an adjacent extra pleural empyema was evacuated [McCONCHIE and HAYWARD, 1958]. Subsequently she developed an apical empyema which had healed by the time of her discharge from hospital on 23 May 1951. After discharge she felt well, except for less severe headache which lasted for one or two days, and recurred over the years at intervals of days to several weeks. The headache seemed to start in one eye, usually the left, and go through to the back of the head. As the pain increased the other eye became sore. She had become frightfully forgetful and she worried a lot. She had a 'little bit of smoker's cough' with sometimes a little pain in the chest, and occasionally she brought up a little phlegm.

In 1953 she had a number of carbuncles, but no cryptococci were found in the pus. On 27 July 1953 the pressure of the cerebrospinal fluid was normal. It contained 5 polymorphs and 18 lymphocytes per mm^3, 80 mg% of protein, with a slight increase in globulin, 738 mg% of chlorides and 0.07 mg% of sugar. No precipitation of colloidal gold occurred. No cryptococci were seen or cultured.

On 10 April 1958 she felt extremely well and had suffered only one severe headache and several mild ones over the previous year. The cerebrospinal fluid contained 1 polymorph per mm^3, the protein content was 100 mg% and the colloidal gold reaction was of meningeal type. Again no cryptococci were seen. Bouts of headache continued and

whenever examined the cerebrospinal fluid contained a small number of cells (e.g. on 27 February 1968, 3 polymorphs and 24 lymphocytes). The protein was always high (around 220 mg%) and the sugar between 20 and 30 mg%. No cryptococci could be demonstrated. Innoculations did not infect mice.

She remained reasonably well, able to bring up her family, do her housework and cope with a turbulant domestic situation, until in January 1969 headaches and giddiness became severe, with nausea and vomiting. She was unable to maintain her balance and fell

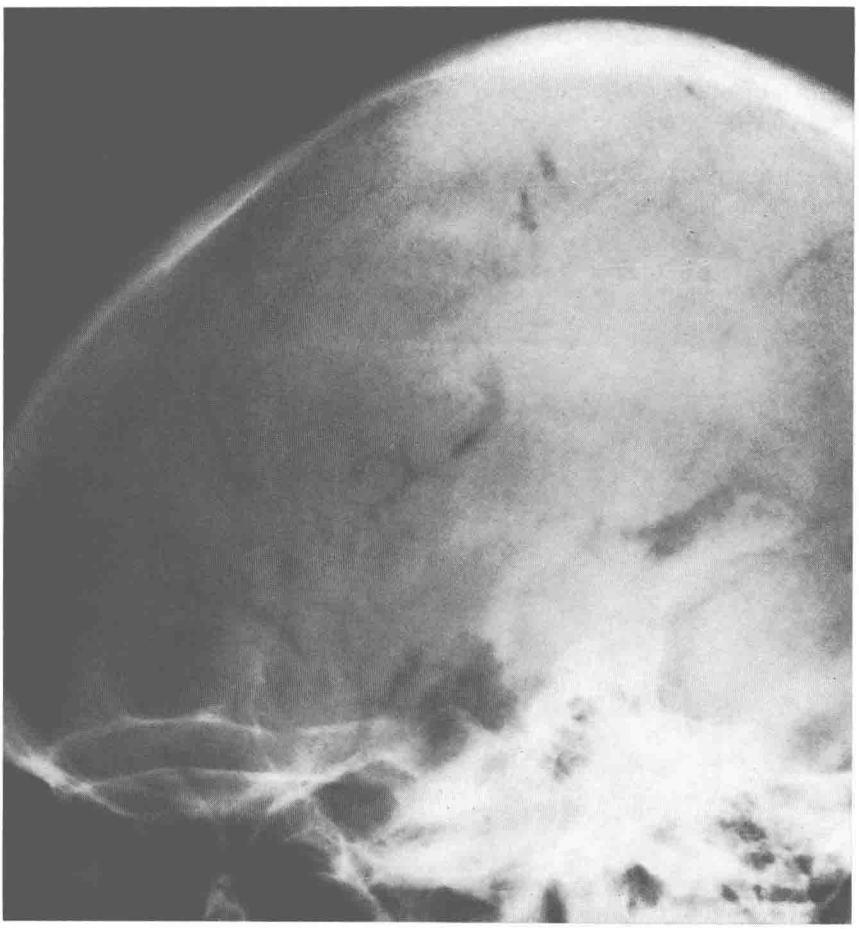

Fig. 2. Case 2: meningitis due to *Cryptococcus neoformans*. Gas entered the skull very slowly, and accumulated in the cervical canal. It is seen in the pontine, interpeduncular and chiasmatic cisterns, in the superior cerebellar cistern and in a few sulci. Several other stages of filling are illustrated to show that important deductions can be made even when only small volumes of gas enter the skull.