Relapses and Remissions in Brain Stem Tumours

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Summary: Three cases of brain stem astrocytoma and one of a para-stem vascular tumour are described, each showing a history of relapses and remissions. The diagnoses were confirmed by necropsy, and it is suggested that the relapsing course may have been due to oedema and necrosis in both the tumour and the surrounding brain tissue. Features indicating repeated recurrence of a lesion at the same site in the brain stem justify full neuroradiological investigation before a diagnosis such as multiple sclerosis can be accepted.

Introduction

Though brain stem gliomas are more common in children than in adults, all age groups may be affected. They usually present as a progressive disorder, consisting of oculomotor palsy, defects of conjugate ocular deviation, and other scattered cranial nerve palsies. Pyramidal or cerebellar defects may appear early or may be long delayed, but signs of raised intracranial pressure are uncommon until the later stages. It is not widely appreciated, however, that these tumours may show a relapsing and remittent course, the remission sometimes being so complete and over such a long period that a diagnosis of multiple sclerosis may have been made, and regarded as correct until the final deterioration with headache, vomiting, coma, and respiratory failure brings the true diagnosis to light. Ford (1966) mentioned two children with brain stem glioma who had complete and spontaneous recovery and Sachs (1949) described a case in which the diplopia cleared up for several days before recurring. Apart from these reports, no other record of remission has been found in the literature.

The following four cases illustrate the course that some brain stem and para-brain-stem tumours take, and are recorded in the hope that they will emphasize the importance of keeping this diagnosis high among the differential possibilities when signs and symptoms continuously and repeatedly point to a lesion in this one part of the nervous system.

Case 1

A woman aged 21 gave a five-month history of right-sided dysesthesia followed by diplopia and right internal strabismus. This recovered completely, but was followed by a left internal strabismus. She had had left-sided ataxia for eight weeks, left deafness and tinnitus for three weeks, early morning headache with occasional vomiting for four weeks, and amenorrhea for three months.
Examination showed a left seventh-nerve palsy, nystagmus to the right, slight left-sided deafness, and a left-sided cerebellar ataxia. All tendon reflexes were exaggerated, right more than left, the abdominal reflexes were absent, and the right plantar response was equivocal.

**Investigations.**—Blood count, blood serology, cerebrospinal fluid (C.S.F.) examination, electroencephalograph (E.E.G.), and x-ray examination of the chest and skull showed nothing abnormal. Erythrocyte sedimentation rate (E.S.R.) was 25 mm. Audiogram showed slight nerve deafness. Caloric tests were normal.

During investigation all symptoms and signs completely cleared but four months later she developed headache and vomiting and became unconscious, with marked neck stiffness. Examination showed stupor, anaphoria, neck stiffness, impaired conjugate movement to the left with left sixth-nerve palsy, left lower motor neurone facial paralysis, and deafness in the left ear. Pain sensation was impaired on the left side of the face and the right side of the body. There was generalized hyperreflexia and both plantars were extensor.

Reinvestigation showed a polymorphonuclear leucocytosis, E.S.R. 100 mm., and a xanthochromic C.S.F. at 220 mm. pressure containing 490 mg. of protein per 100 ml. Air ventriculography showed hydrocephalus due to obstruction in the fourth ventricle.

She died five days later, and necropsy showed the pons and medulla almost completely replaced by an anaplastic astrocytoma which surrounded the seventh and eighth nerves in which were grooves made by the vertebral and anterior inferior cerebellar arteries.

**Case 2**

This patient was a man aged 34. At the age of 18 his head flexed to the right for six weeks and recovered. Nine months before admission the left hand became weak for three hours and recovered. Six months later the left arm and leg developed progressive weakness.

Examination showed dysarthria, horizontal nystagmus, right-sided palatal weakness, and wasting with fasciculation of the right side of the tongue. The left arm and leg were spastic, with exaggerated reflexes and extensor plantar.

**Investigations**—Blood count, E.S.R., blood and C.S.F. serology, and C.S.F. analysis were normal. C.S.F. pressure was 210 mm. E.E.G., x-ray examination of chest and skull, and vertebral arteriogram showed nothing abnormal. Air encaphalography showed bialateral displacement of the fourth ventricle.

Ten hours after vertebral angiogram he developed respiratory distress, became comatose, and died. Necropsy showed the medulla enlarged and distorted by a well-differentiated astrocytoma which almost completely surrounded the right vertebral artery.

**Case 3**

A girl aged 41 years developed headache, vomiting, and drowsiness after a minor head injury, and she walked with a broad-based atactic gait and had a right hemiparesis. All the signs and symptoms cleared up except for a slight limp. Twelve months later she developed progressive weakness of the right side, unsteadiness, headache, and vomiting. Her left upper eyelid gradually drooped. Examination showed bilateral papilloedema, partial left third- and mild sixth-nerve palsy, right hemiparesis, and cerebellar ataxia on the left. Tendon reflexes in the lower limbs were exaggerated and the right plantar was extensor.

**Investigations**—Blood count, E.S.R., ventricular C.S.F. analysis, blood, and C.S.F. serology were normal. E.E.G. showed bilateral slow waves. X-ray examination of chest showed nothing abnormal. X-ray examination of skull and neck showed angiogram showed features of hydrocephalus. Air ventriculogram revealed enlarged lateral ventricles due to an obstruction by a space-occupying lesion of the fourth ventricle which had displaced the lower end of the third and fourth ventricles backwards and to the right.

Her condition gradually deteriorated and she died eight weeks later. Necropsy showed enlarged lateral ventricles. The left side of the fourth ventricle and the posterior end of the third ventricle were replaced by cystic astrocytoma which extended up to the optic chiasma and anterior internal capsule.

**Case 4**

A girl aged 13 gave a history of left occipital headache for four to five years. Ten days before admission she developed severe headache followed by vomiting, diplopia, and unsteadiness, with falls to the left.

Examination showed horizontal nystagmus to the right, lower motor neurone weakness of the left face, and left-sided cerebellar ataxia with exaggerated left-sided tendon reflexes. Blood pressure was 130/100 ml.

**Investigations.**—Blood count, E.S.R., and blood and C.S.F. serology were normal. X-ray examination of the chest and skull showed nothing abnormal. The C.S.F. showed lymphocytosis with total cells of 23 and 27/cu. mm. at two subsequent examinations and protein of 65 and 52 mg./100 ml. E.E.G. showed slow waves on the right. Air and iophendylate (Myodil) ventriculography revealed dilatation of the lateral and third ventricles.

She gradually improved, and ataxia, double vision, and headache disappeared. She was seen on five occasions at intervals of six months; each and every time the only detectable sign was nystagmus. Four months after her last examination she was readmitted with severe headache, vomiting, pain and stiffness of the neck, and double vision.

Examination revealed marked spinal rigidity, bilateral papilloedema, motor and sensory impairment of the left fifth nerve, bilateral sixth-nerve weakness, lower motor neurone left facial weakness, complete deafness in the left ear, horizontal nystagmus, and left-sided ataxia.

**Investigations**—Blood count, E.S.R., and blood and C.S.F. serology were normal. X-ray of the skull showed a large, rounded, single meningioma of the skull, and x-ray examination of the chest showed an intracavitary mass of the right lung. Bronchopneumonia was present.

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**Discussion**

If a young patient shows the symptoms and signs of a brain stem lesion which, having been present for some days or weeks, remit completely, multiple sclerosis becomes the most likely diagnosis. It is, however, a diagnosis which should always be made with caution, for there is a very real danger that if this "label" is attached to a patient it will remain, and may well be used to explain any and every subsequent clinical manifestation even if the new features are unusual in demyelinating disease. Particular care should therefore be taken if the signs of brain stem disturbance are accompanied by significant headache and vomiting, and any alteration of conscious level should be viewed with added caution. Again the value of the well-established term "disseminated" sclerosis was that it emphasized the need for the lesions to be disseminated in the nervous system, preferably in both time and place, and when signs repeatedly pointed to a lesion in the same area such cases warrant full investigation.

The finding of a brain stem glioma may worsen the prognosis, but the possibility arises that an extrinsic tumour impinging on the brain stem might be found which might be capable of surgical eradication. In investigating this type of case the value of both introducing air or iophendylate into the ventricular system and of carrying out vertebral arteriography is very considerable, for, whereas both intrinsic tumours of the stem and those lying anterior to it displace the aqueduct and the fourth ventricle backwards, intrinsic tumours press the basilar artery
Role of Candida albicans Infection in Napkin Rashes

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Summary: Skin scrapings, mouth swabs, and faecal specimens from children with eruptions in the napkin area and from a series of normal infants were examined for the presence of Candida albicans.

This was found in 41% of all napkin eruptions but in only one of the 68 normal infants. While C. albicans is a common secondary invader of all types of napkin eruption, primary candida infection of the skin in the napkin area is probably uncommon.

No evidence was found that generalized psoriasiform or eczematous eruptions occurring in association with napkin rashes are due to an allergic response to the fungus. C. albicans is more likely to be present in a napkin rash if the organism has been found in the alimentary tract.

Introduction

Four main types of napkin eruption are usually described, and include the common napkin rash due mainly to irritation by ammonia, seborrheic (irritigenous) eczema, a psoriasis-like eruption (napkin psoriasis), and an eruption reported as a primary Candida albicans infection. C. albicans is well recognized as an opportunistic organism growing where general and local conditions are favourable, and among these conditions moisture and a pre-existing skin disorder are important. C. albicans has been recovered from the alimentary tract of up to 31% of infants under the age of 9 months (Bear, Cheeseman, and MacKenzie, 1958), and it is therefore not surprising that the organism has been isolated from napkin rashes of all types. When so discovered C. albicans may well be accepted as having a primary role in the napkin eruption, and psoriasiform or eczematous lesions which may develop in other sites of the body have often been considered to be either a spread of the infection or an allergic response to the fungus.

In the present investigation we compare the prevalence of C. albicans infection of the skin and alimentary tract in children with different patterns of napkin eruption and in a series of normal controls. Our purpose was to determine, firstly, whether infections of the skin in the napkin area by C. albicans were more likely to be primary or secondary in origin, and, secondly, whether there is any evidence for a causal relationship between C. albicans infection and generalized psoriasiform or eczematous eruptions.

Material

The cases included in the investigation were either referred by their doctors to the dermatological outpatient department of one of the Bristol hospitals or were attending certain of the local authority's child welfare clinics. Very little further selection was made. All children with a napkin eruption who attended the dermatological clinic at the Royal Hospital for Sick Children during the period of the investigation were included. Occasionally in child welfare clinics time did not permit the inclusion of some children with a napkin rash, but this happened only rarely.

Group 1. Napkin Psoriasis.—Of the 27 cases in this group 24 were referred to the Royal Hospital for Sick Children between April 1967 and June 1968 and three were seen at other hospitals. The eruption usually starts in the napkin area, often mainly on the napkin contact sites, and consists of patches of brown-red skin with some scale and well-marginated figurate edges often enclosing areas of normal skin. Lesions of up to 10 cm. in diameter or larger, all with well-defined edges, then develop on the trunk and limbs and usually the scalp,