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A Case of Craniopharyngioma Presenting as Korsakov's Syndrome

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The particular mental state described by Korsakov (1890, 1891) is, in most cases, a sequel of Wernicke's encephalopathy, which itself is the result of alcoholism. Korsakov, however, also observed the same mental condition in persons, not alcoholic, who had been exposed to carbon monoxide, to phosphorus poisoning, or to profound toxic states. Within a few years, Mönckenmöller (1899) and Meyer (1899) had each described the Korsakov state appearing late in the natural history of sarcomata of the sella turcica and third ventricle respectively. Delay *et al.* (1964) ably reviewed 108 instances in the literature of the association of cerebral tumour and Korsakov state, and concluded that cerebral tumour is most likely to lead to the Korsakov state if it leads to a bilateral lesion in the hippocampo - mamillo - thalamo - cingulate pathways, an observation previously made by Grünthal (1939), Kleist (1934), and Orthner (1957). By virtue of their possible situation, craniopharyngiomas are so placed anatomically that they are associated with the Korsakov state with disproportionate frequency compared to other intracranial tumours. However, in some of the reported cases the Korsakov features were tenuous, were a near terminal or post-operative event, or were outweighed by gross endocrine disturbances. Even so, only 3 out of 28 such cases described in the literature were diagnosed during life. Early diagnosis is thus extremely difficult, for the neurological signs are sparse and then are mainly ophthalmological, whilst the onset is insidious and accompanied by mental changes which deceptively suggest a functional basis to the whole picture.

We therefore propose to describe a case of Korsakov's syndrome due to a craniopharyngioma which re-emphasizes the need to undertake full investigation of any amnestic syndrome that is without a history of alcoholism.

Case History

Mrs. C. R., a 52-year-old married woman with one son, was transferred from the Bethlem Royal Hospital to the Guy's Maudsley Neurosurgical Unit on 14 November, 1965, for exploration of a suprasellar lesion. She came from a happy but impoverished family remarkable for the extreme frequency of neoplastic disease in its members; thus 3 of 6 maternal siblings, 4 of 8 paternal siblings, and 3 of 10 siblings had died of neoplastic disease. There were minor neurotic traits in childhood, but she had worked regularly until the age of 20, when she married. The menarche was at 12 years. She bore a son soon after marriage and her periods stopped at the age of 50. All sexual relationships with her husband ceased in her mid-thirties. She had worked for most of her life, but the home had been run by her mother-in-law until she died in 1961. The patient's general health was described as excellent; she was a lively, pleasant person, much attached to her family.

In September 1964, she changed her job, and soon gave the new one up on the grounds that her employer had exposed himself to her. Then in November it was noticed that she was repeating herself in conversation, continually asking irrelevant questions, and not paying attention to the answers. She was forgetful, and in her shopping she would buy the wrong things; she would also, having taken her dog out for a walk, repeat the performance a few minutes later. In December she visited an optician, complaining of misty vision. He made some vague statement about her fundi, but gave her glasses which corrected her vision whenever she could remember to put them on. Gradually, listlessness and apathy, the repeated forgetting of appointments, and diurnal somnolence confined her more and more to the home. She became irritable, wept frequently, berated her husband for his lack of affection and impotence, and blamed the menopause for her condition. The general practitioner diagnosed menopausal depression, but there was no response to his treatment. In February, 1965, she called upon one sister when in fact she was due to see another. Having done this, she broke down in uncontrollable sobbing; she then spoke as if she was in her own home, and asserted that a friend, long since dead, was in the house. She asked questions which implied that her niece was still courting a man who had in fact married someone else two years before. This episode led to her admission to the Bethlem Royal Hospital.

On admission, her memory for older events was better than that for recent ones. She was disoriented in time and place, repeated herself in conversation, and was agitated and restless. She wandered away on several occasions,

wore clothing unsuitable to the temperature, and frequently became lost on her way to the toilet. She blamed her husband for stinginess, struck him when he visited, and confided to the doctors an unlikely story of her own infidelity, and an account of her sexual relations with her husband which he denied.

All investigation directed at an organic cause for her symptomatology were negative (e.g. X-rays of skull were reported normal as was the EEG). She gradually improved in her mood and her orientation, and became more helpful in the ward. At the time of discharge it was believed that her depressive illness had released hysterical behaviour. However, the psychological tests at the time were compatible with Korsakov's psychosis, in that she had good immediate recall, but impaired short-term memory. W.A.I.S. full scale 87, verbal scale 91, performance scale 83.

During the next month she remained able to do her shopping and housework, but in June 1965, when away from home on holiday, she became very disoriented, agitated, and confused. Her somnolence was marked, and her husband noted her voracious appetite. She gained 8 lb. in weight in two weeks. On return home she was either cooking her joint for ten minutes, or forgetting to cook it at all, or else she wandered around all day doing nothing. Her sister might come and do all the work, but the patient would claim that she had done it all herself. She spoke of her deceased mother-in-law as though she was still alive.

In September, 1965 the first drop attack was observed. In this she suddenly fell to the ground and remained there looking dazed but immobile for 15 minutes. Further attacks continued, gradually accompanied by incontinence until late October, when she had a major convulsion.

Following this she was again admitted to the Bethlem Royal Hospital. She was now very disoriented, had no recollection of her previous admission, was continuously restless, while her talk was sporadic and telegraphic and she perseverated. Her general information was very poor for recent events, and she believed the Queen was in Norway to celebrate the birth of her son Paul. Her immediate digit span was eight digits forwards and five backwards, but she was unable to remember a simple name for two minutes. Psychological tests showed a Wechsler Bellevue Full Scale I.Q. 85, V.S. 89, P.S. 82, similar to those taken before. She was unable, however, to learn any new material. For example, in Meyer's paired associate tests she persistently made clang or habitual associations instead of the correct ones.

Physical examination now revealed early papilloedema and bilateral extensor plantar responses, but no other physical sign, including normal visual fields. The skull X-rays now showed that the dorsum sellae had become eroded and that there were flecks of calcification above it. The lumbar cerebrospinal fluid was normal.

On admission to the Neurosurgical Unit she was still alert at interview, but became somnolent when left alone. She would wander restlessly around, becoming lost at every turn. She could be made to confabulate with ease. The air encephalogram confirmed that there was a small tumour above the dorsum sellae, and a pre-operative diagnosis of craniopharyngioma in the interpeduncular fossa was made. At operation a loculated cyst was sucked out and biopsied. It lay in the expected site in the interpeduncular fossa but in front of the brain stem and above the sella. It was inoperable. The pathological report was of a craniopharyngioma. During the first few post-operative days she was more alert, oriented, and recognized her doctor. Thereafter she gradually became more stuperose. She remains in this state.

Discussion

The clinical features exhibited by this patient are those of Korsakov psychosis (Lewis, 1961; Hoenig *et al.*, 1962; and Delay *et al.*, (1964). In cases where this association exists a distinction has to be made between the "Korsakov" symptoms, those due to hypothalamic dysfunction, and symptoms of cerebral tumour. Of particular note in this case was the rapid development and early presentation of the amnestic state in the history of the tumour. A review of thirteen cases (Table I) in which

Table I

Author Year Case	Age Sex	Symptoms in order of Presentation (1)	Symptoms in order of Presentation (2)	Physical Signs	Operation	Time of Diagnosis	Length of illness (months)
1. Lang 1924	38 F	Severe headache Amenorrhoea Disorientation	Memory deficit Confabulation Somnolence	Torpor Weight loss Vomiting	No	P.M.	15
2. Almeida Dias 1933	32 M	Irritability Obscene behaviour Headaches Vomiting Blurred vision Somnolence	Adiposity Polyuria-dypsia Memory disturbance Confabulation Hallucination	Nil	No	P.M.	36

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Author Year Case	Age Sex	Symptoms in order of Presentation (1)	Symptoms in order of Presentation (2)	Physical Signs	Operation	Time of Diagnosis	Length of Illness (months)
3. Grünthal 1939	40 M	Localized headache Nausea Disorientation	Insomnia Lability of affect Incontinence	Ataxia	No	P.M.	15
4. Wagner (O. M.) 1942	27 M	Headaches Fall off in work Absent libido Confusion	Failing vision Thirst Retardation Memory impairment	Pallor of discs Hemianopia	No	P.M.	8
5. Wagner (J. H.) 1942	45	Irritability Mild hypomania Visual disturbance	Disorientation Confabulation Inverted sleep rhythm	Central scotoma, Optic atrophy, Spastic lower limbs	No	P.M.	7
6. Wagner (K.) 1942	38 M	Tiredness Lying Sweating Somnolence Hypomania	Forgetfulness Headaches Vomiting Disorientation Confabulation	Optic atrophy	No	P.M.	6
7. Riese 1950	57 F	Weakness of limbs Hallucinations Disorientation Memory disturbance	Excessive thirst Excessive appetite Confabulation Euphoria	Bilateral Hoffman's+ Skull Percussion painful on R.	No	P.M.	8
8. Williams (2) Pennybacker 1954	51 F	Memory lapses Confusional episode Headaches and vomiting Somnolence	Euphoria Disorientation Poor memory Confabulation	Nil	Yes	Post-op.	4
9. Sivadon 1957	58 M	Disorientated Confabulation	Memory Euphoria	Bilateral Hoffman Rossolimo	No	P.M.	?
10. Russell (10) Pennybacker 1961	60 F	Irritability Disorientation Headaches	Incontinence Somnolence	Nil	Yes	X-ray biopsy	24
11. Russell (6) Pennybacker 1961	67 F	Memory loss Poor concentration Somnolence Visual loss	Confabulation Disorientation Depression	Nil	No	P.M.	12
12. Russell (11) Pennybacker 1961	57 M	Tiredness Somnolence Depression	Memory loss Disorientation Confabulation	Nil	Yes	P.M.	24
13. Russell (20) Pennybacker 1961	66 F	Headaches Visual loss Somnolence Thirst	Memory disturbance Confabulation Disorientation	Anosmia Hemianopia	Yes	Biopsy	52

mental symptoms occurred early on in the history shows that there is a commonly recurring group of symptoms in these tumours. We agree with Sivadon (1957) that where mental symptoms predominate the correct diagnosis is often delayed.

The Korsakov state in our patient was of interest since in addition to the embarassment confabulation of Bonhoeffer (1904) which is nonspecific in the organic psychosyndrome (Bleuler, 1954) she showed true confabulation such as false memories (Delay *et al.*, 1964) which emerged clearly in the Meyer's paired associate test. True confabulation was often in the sphere of temporal relationships and evidence of this was also in her case history, a point emphasized by van der Horst (1932). Disorientation at home and inability to cope at an early stage were probably related to her recent move of house and to the fact that housework was for her a recently acquired skill. This probably accounts for her early presentation to the Emergency Clinic with purely mental symptoms. It seems to us that much of the apparent difference in the cases as they present has been due to the stage in the natural history at which they have been referred. Thus Russell and Pennybacker's Case 6 had been at home for nine months "gradually losing her sight and her senses" before she was admitted.

The most common hypothalamic symptom is somnolence and reversal of sleep rhythm, but polydipsia and polyphagia occur often late in the natural history. Changes in libido are also mentioned. In our case the patient had had no sexual relationship with her husband for sixteen years, but then within a space of six months she had complained of an indecent exposure, claimed an extramarital affair (whether due to increased libido, phantasy, or to confabulation is unknown), and also claimed resumption of marital relationships. Since her periods ceased at this time, all this was attributed to the menopause. The position of the tumour was such that it lay within the interpeduncular fossa in a situation where it could compress the hypothalamus and the mamillary bodies. There were no visual field changes in our case, but these do occur because of chiasmatic pressure. Headache and mood changes, papilloedema and optic atrophy, when they occur, are all non-specific changes. Complaints of visual disturbance are frequent as in our own case, but are very often unaccompanied by any signs. X-ray signs are noted in about 50 per cent. of craniopharyngiomas, but in this patient, retrospective examination revealed only the faintest trace of calcification in the X rays taken on the first admission. The air encephalogram is often more valuable.

Summary

A case of craniopharyngioma and Korsakov psychosis is presented to emphasize the need to consider alternatives to alcoholism as the cause of Korsakov psychosis. Where mental symptoms present early with minimal endocrine changes the diagnosis is likely to be missed. In a case of clear-cut Korsakov psychosis without alcoholism or previous history of anoxia, a neoplastic cause should be presumed until disproved by full investigation and follow-up.

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