Incidental olfactory aplasia: a case report

We report a case of olfactory aplasia with no other cerebral malformation in an elderly woman, of high intelligence. The patient was 73 years old and first presented at the age of 69 complaining of forgetfulness. On review by a neurologist at this time, history and examination were unremarkable apart from information volunteered by the patient herself that she had never had any sense of smell.

Neuropsychological testing was also performed with standard tests from the Wechsler Intelligence Scale. The patient achieved a memory quotient well in excess of 143, which is the highest score in the published norms. Her pro-rated verbal and performance IQs were also in the high range at 134.

She was admitted to the Alfred Hospital, Melbourne, in January 1992 after collapse. A cerebral CT scan showed an intracerebral haemorrhage with extension of blood into the fourth, third, and lateral ventricles. She was taken to theatre where the blood clot was evacuated and a Rickham’s reservoir was inserted. A postoperative CT scan showed no detectable abnormalities of the mid-brain and pons. Review of the cerebral CT scan showed no detectable abnormalities of the skull.

Histological examination of the frontal cortex and white matter was unremarkable. The pituitary gland appeared normal. Some hypoxic change was noted. There was extensive haemorrhagic necrosis of the mid-brain,pons, and the periventricular region of the medulla. Sections of the cerebellum showed a haemangiomatosis with focal necrosis and extensive haemorrhage.

Olfactory aplasia is rarely encountered as an isolated abnormality in an otherwise normal patient.1 More commonly it is associated with a wide range of other malformations that may be both cerebral and somatic. The cerebral malformations are usually categorised by the broad term arhinencephaly, and include holoprosencephaly, agenesis of the corpus callosum, abnormalities of the optic tracts, cerebellar hypoplasia, and dentoooliary dysplasia.2 These abnormalities may range from mild to severe in degree but mental retardation is usually present. Somatic malformations are also common. These include facial dysmorphism, which may take the classical form of midline hypoplasia, especially when associated with holoprosencephaly, with cyclopia, hypotelorism, ethmencephaly, ceboccephaly, and cleft palate.3 A wide spectrum of other systemic abnormalities has been found particularly when holoprosencephaly is associated with chromosomal anomalies,4 but also in other conditions where olfactory aplasia occurs. The sole macroscopic cerebral abnormality in this patient was the absence of the olfactory bulbs.

This case is unusual not only because other features of the arhinencephaly spectrum that are often associated with olfactory aplasia were absent, but particularly because this patient was of superior intelligence.

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Amebic brain abscess: a rare but serious complication of Entamoeba histolytica Infection

Since its first description in 1849, 1 there have been several reports of cerebral amoebiasis. 2,3 To our knowledge only nine adequately documented cases of cure have been described. 4,5 We present a case of cerebral amoebiasis with no evidence of disease elsewhere who made a complete recovery after surgery and treatment with intravenous metronidazole.

Figure 1 CT scan of the brain with contrast showing an ill defined mass in the right parietal region with ventricular compression.

A 57-year-old man presented to the emergency room with a three day history of progressive weakness of the left half of the body associated with confusion, headache, and vomiting. He had had a sore throat and fever for one week before admission. On admission, he was febrile (39°C) and disoriented. He had a left hemiplegia with reduced sensation, increased tone, and an extensor plantar response on the left side. The rest of the examination was unremarkable. His white cell count was 13·9 with 82% neutrophils and his erythrocyte sedimentation rate was 77. Tests of liver function, blood urea, and creatinine were normal. He was negative for HIV by enzyme linked immunosorbent assay (ELISA) and serum protein immunoelectrophoresis was within normal limits. A chest radiograph showed no abnormality. CT of the head showed a large mixed density lesion in the right posterior parietal region. There was compression of the posterior half of the right lateral ventricle (fig 1). A burr hole biopsy revealed necrotic material, and a provisional report suggested a fungal infection. Treatment with amphotericin B was started.

Figure 2 Cerebral cortical tissue showing trophozoites of Entamoeba histolytica (periodic acid-Schiff stain, originally × 400).
As the patient continued to deteriorate an emergency craniotomy was made the same evening. The mass consisting of dark brown necrotic material was removed by suction. Widespread cortical thrombophlebitis was noted. Examination of the material removed confirmed the presence of amoebae (fig 2). Treatment with metronidazole (500 mg 6 hours intravenously) was started and continued for three weeks. Postoperatively the patient was ventilated for 48 hours. Thereafter his condition began to improve. Subsequent bacterial and fungal cultures of the biopsy material were negative, whereas ELISA and indirect haemagglutination assay (IHA) titres were 1:3200 and 1:512 respectively. Investigations to find a primary focus included several stool examinations, which were negative and an abdominal ultrasound, which was normal. Abdominal CT disclosed an irregular, lobulated centrally necrotic mass arising from the upper pole of the right kidney. A needle biopsy of the lesion under ultrasonic guidance was inconclusive. The patient gradually made a complete neurological recovery and was discharged home.

Subsequently the patient underwent a radical right nephrectomy and para-aortic lymphadenectomy for a well differentiated multicystic renal cell carcinoma without extension into perirenal or perineural vascular tissue, or the lymph nodes. There was no evidence of hepatic or renal amoebiasis.

The classic description of the clinical presentation was given by Armitage. The patient is usually a male in the prime of life who has had dysentery, acute or chronic, with suppurative hepatitis. After liver abscess drainage and while apparently improving, cerebral symptoms and signs develop. The average duration from the first neurological symptom to death is 10 to 15 days. Orbison et al, in an extensive review of publications, were able to collect 83 cases in 1961 and 1967 (fig 2). Treatment with metronidazole is 1:128 or an ELISA ranging from 1:1000 to 1:50 000 is suggestive of extra-intestinal amoebiasis. Negative IHA, ELISA, and indirect immunofluorescence make the diagnosis of invasive amoebiasis unlikely. We have found only three previous reports of CT. Becker et al described a poorly defined lesion without any zone of reactivity as in our patient. Schmutzhard et al and Tikly et al described small or large ring-like enhancing lesions involving predominantly the white matter with surrounding oedema and midline shift.

Direct observation by light microscopy with an unstained wet preparation will show motile amoebae. Tissue is best obtained from the periphery of the lesion. Permanent Wright stain, Feulgen reaction, and immunofluorescent labelled antibodies directed against the amoebae in tissue may be used or amoebae isolated and cultured, the last requiring considerable experience. Surgical decompression with removal of necrotic and infected material may be required for the control of intracranial pressure. Intravenous metronidazole is essential and achieves adequate CNS penetration when the history of illness is relatively short. This report describes only the 10th case documented to have been cured of cerebral amoebiasis with the use of a combined surgical and medical approach.