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 intracerebellar 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 angiogram revealed digital subtraction angiogram revealed a patent circle of Willis. A standard transcranial Doppler was performed which revealed an abnormality in the anterior cerebral artery. She was discharged home in good condition.

On review at 3 months, the patient was found to have left hemiparesis, nystagmus, and an extensor plantar response. She had had a severe stroke and she had a score of 9/100 at the follow-up on January 1993.

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

Histological examination of the frontal cortex and white matter was unremarkable. The pituitary gland appeared normal. Some hypothalamic 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 haemangioblastoma with focal necrosis and extensive haemorrhage.

Olfactory aplasia is rarely encountered as an isolated abnormality, and was not observed in this normal patient. 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 dento-ovarian dysplasia.5 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, ethmocelephaly, cebocephaly, and cleft palate.6 A wide spectrum of other cranial abnormalities has been found, particularly when holoprosencephaly is associated with chromosomal anomalies,7 but also in other conditions where olfactory aplasia occurs in the sole macroscopic cerebral abnormality, as in Kallmann’s Syndrome. This syndrome is characterised by hypogonadism and anosmia with similarities to the olfactory bulbs, which may be associated with diastrophic dysplasia, lip and palate, sensorineural deafness, and skeletal anomalies.8

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.

KRISTY E. DUNDAS
ROSS MC D ANDERSON
Department of Anatomical Pathology, Alfred Hospital, Commercial Road, Prahran, 3181, Australia


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

Since its first description in 1849, there have been several reports of cerebral amoebiasis.9 To our knowledge only nine adequately documented cases of cure have been described.10 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 2 Cerebral cortical tissue showing trophozoites of Entamoeba histolytica (periodic acid-Schiff stain, originally x 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 periureal 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 suppurrative 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 36 cases in 1977. The average age of their patients was 32 years. Headache or alteration of mental state were the most common initial presentations. Symptoms and signs of meningitis occurred in 45% of the patients. Abnormalities in the CSF were noted in eight of 10 patients tested. Cranial nerve involvement was frequent. Fever and neutrophilia was noted in most cases; 40% of the patients had no history of amoebic dysentery.

Diagnosis is now facilitated by amoebic serology and CT scanning as well as histopathology. The most sensitive test available is IHA; others are latex agglutination, countercurrent immunoelectrophoresis, indirect immunofluorescence, radioimmunoassay, and ELISA. An IHA titre of 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 Ting 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 amoeba 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.

Letters to the Editor
Department of Neurosurgery
HIZBULLAH SHAHIR
Department of Pathology
MICHELE KARIM
Department of Medicine
The Aga Khan University Medical Centre
Stadium Road, PO Box 3500, Karachi 74806, Pakistan

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