Cerebellar tuberculoma: a rare disease in an industrialized country

Jens Haase*, Peter Brøgger Christensen, Kirsten Kock and Helge Michaelsen Ingstrup
Department of Neurosurgery, Aalborg Sygehus Syd, DK-9100 Aalborg, Denmark

Abstract. A nine-year-old Somalian boy was treated in Denmark for a posterior fossa tumor, which proved to be a tuberculoma. This disease is virtually unknown in our highly industrialized country, which emphasizes the fact that due to increased international traveling, knowledge of endemic diseases in other parts of the world is necessary. The treatment of tuberculoma and their diagnostic features are reviewed.

Key words: Tuberculoma – CNS tuberculosis.

In 1883, Macewen operated upon the first cerebral tuberculoma [12]. With the turn of the century, Cushing stated that tuberculomas constituted about 30%–40% of all intracranial space-occupying lesions [2]. The fact that intracranial tuberculosis develops from hematogenous spread of extracerebral tuberculous foci explains why improved nutrition and social standards combined with prophylactic BCG vaccinations have caused the incidence of tuberculomas to drop significantly [9]. Today tuberculomas constitute less than 0.15% of intracranial tumors in industrialized societies [4, 6, 9, 13].

However, underdeveloped countries in Africa and Asia still have an incidence of 16%–20% [9, 11]. The Scandinavian countries possess some of the world’s most complete health data files. From these we found that among the Danish population of 5 million people only 3–8 cases of tuberculous meningitis have been encountered each year, and throughout the last 10 years not a single case of intracranial tuberculoma has been observed.

For the new generation of younger neurosurgeons it is thus obvious that intracranial tuberculomas can easily become “a forgotten disease.” This is the reason for presenting this case report.

Case history

A 9-year-old Somalian boy with a life-long history of ataxia, headache, and increased head circumference was referred to the Somali University Hospital in Africa. It was noted that his baby brother had died shortly after birth with a large head. A visiting Italian neurosurgeon performed ventriculography, which revealed severe hydrocephalus and a posterior fossa tumor. A ventriculoperitoneal shunt was inserted, while treatment of the tumor was deferred for technical reasons. It was suggested that his family should have him transferred to a European country for surgical exploration.

Six months later, while visiting his family in Denmark, he was referred to our department because of persistent, staggering gait, headache, and slightly decreased visual acuity. He was found to be short of stature, with a typical hydrocephalic cranium and head circumference of 55 cm. Neurologically, bilateral clumsiness and ataxia were noted, together with bilateral optic nerve atrophy and slightly decreased visual acuity. A CT scan (Fig. 1) revealed, in addition to large bilateral subdural hygromas, a 5×5×5 cm ring-enhancing capsular process with an isodense center in the left cerebellar hemisphere. Angiography showed no tumor blush. Chest radiographs and abdominal ultrasound scans gave no evidence of space-occupying processes in the lungs, liver, or the retroperitoneal space. A Mantoux intradermal skin test (intradermal PPD test) was marginally positive. Blood tests demonstrated hypochromic anemia and eosinophilia, and feces contained hook-worm ovi, which was subsequently treated with mebendazole. The ventriculoperitoneal shunt was revised with insertion of an antisiphon device, and the two subdural hygromas were evacuated through burr holes. No signs of meningeal tuberculosis was encountered.

One week later, with the boy in a sitting position, a left-sided posterior fossa craniectomy was carried out and a huge, firm, avascular and well-demarcated tumor, situated subcortically, was removed in toto from the left cerebellar hemisphere. The tumor was a typical tuberculoma with central caseous necrosis (Fig. 2). Microscopy showed chronic granulomatous inflammation with Langhans giant cells and brain necrosis (Fig. 3). No evidence of fungi or echinococcus cysts could be found using PAS staining. Tubercle bacilli were not identified or cultured.

He was treated with prophylactic antituberculous medication – rifampicin, isoniazid and ethambutol (Myambutol) – without complications. The postoperative follow-up was uneventful and a CT scan performed 2 months following surgery (Fig. 4) demonstrated normalization of the cerebellar vermis and the left cerebellar hemisphere, without signs of remaining tumor or tumor spread. The two subdural hematomas had completely resolved.
Fig. 1. CT scan demonstrating 5 x 5 x 5 cm ring-enhancing capsular process in the left cerebellar hemisphere

Fig. 2. Removed tumor; note central caseous necrosis on cut surface

Fig. 3. Histological investigation showing chronic granulomatous inflammation with Langhans giant cells. G, granuloma; L, Langhans giant cell

Fig. 4. Postoperative CT scan

He was discharged neurologically intact without signs of the previous ataxia or complaints of headache. He returned to his homeland for a further 9 months of antituberculous medication. At no time during or preceding the follow-up were signs of extracranial tuberculosis encountered.

Discussion

Intracranial tuberculosis is derived from hematogenous spread of tuberculous lesions in other parts of the body [9, 11]. Two-thirds of solid tuberculomas are found in the cerebellar area and one-third in the cerebrum [4, 7, 9]. Infraenatorial tuberculomas are prominent among children [4, 7]. Multiple tuberculomas may be seen in up to 30% of the cases [4]. It is usually stated that knowledge of tuberculous infection elsewhere in the body leads to the diagnosis [1, 3, 5, 8]. However, today more than 50% of the patients have no evidence of extracranial tuberculosis [5, 8].

Although solid tuberculomas are the most common form, several other types may be found [11]. These are \textit{incipient tuberculoma} in the form of irregular cerebellar