Cerebral chromoblastomycosis complicated by meningitis and multiple fungal aneurysms after resection of a granuloma

Case report

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Cerebral chromoblastomycosis is a rare intracranial lesion. This lesion was found in a 23-year-old man, who presented with right proptosis and fainting attacks. Computerized tomography revealed a moderately enhanced irregular mass in the right frontal region. Angiography disclosed that the mass was avascular. At surgery, a hard elastic avascular tumor was totally removed piecemeal. Histological diagnosis was a granuloma of fungal origin. Characteristic brown pigments in the hyphae of fungus in the granuloma strongly suggested that the fungus was chromoblastomycosis. The postoperative course was complicated by meningitis and rupture of fungal aneurysms. The patient remained vegetative and died 2½ years later. The literature on such fungal aneurysms is briefly reviewed; no previous case of fungal aneurysms associated with cerebral chromoblastomycosis could be found.

KEY WORDS • subarachnoid hemorrhage • cerebral aneurysm • meningitis • fungal aneurysm • granuloma • chromoblastomycosis

CHROMOBLASTOMYCOSIS usually occurs in the skin and subcutaneous tissue, and the fungi rarely invade other organs, including the brain. There have been about 30 case reports of cerebral chromoblastomycosis, and their chief pathological manifestation is abscess formation in the cerebral parenchyma. We report a case with a large granuloma of chromoblastomycosis in the right frontal region, which was complicated by meningitis and multiple fungal aneurysms following resection of the granuloma.

Case Report

This 23-year-old man was admitted to our hospital on December 10, 1979, with chief complaints of progressive proptosis of the right eye and occasional fainting attacks for a few seconds, both of 6 months' duration. There was no contributory history, and no history of taking antibiotics or corticosteroids. He was a city dweller, but while he was a college student he had worked at a stock farm in Hokkaido for several months.

Examination. The patient was an alert and cheerful young man. Neurological examination revealed a moderate proptosis and anosmia on the right, a slight left hemiparesis, and choked discs on both sides. No cutaneous lesions were found. Laboratory data were all normal including immunological studies. Computerized tomography (CT) showed an irregular high-density mass in the right frontal area, which was moderately enhanced after injection of contrast material. The mass also invaded the left frontal lobe, the right orbit, and ethmoid sinus (Fig. 1). Right carotid angiography demonstrated an avascular mass in the right frontal region (Fig. 2A).

Operation. On December 17, 1979, through a right frontotemporal craniotomy, the tumor was grossly totally removed with piecemeal resection. The tumor was hard, elastic, avascular, and dark yellow in color. Dissection from the brain was quite easy, but the tumor firmly adhered to the dura of the right frontal region, invading the right orbital roof and ethmoid sinus. Total weight of the tumor removed was 168 gm.

Postoperative Course. The immediate postoperative course was smooth and uneventful. Five days after operation, histological examination showed a moderate proptosis and anosmia on the right, a slight left hemiparesis, and choked discs on both sides. No cutaneous lesions were found. Laboratory data were all normal including immunological studies. Computerized tomography (CT) showed an irregular high-density mass in the right frontal area, which was moderately enhanced after injection of contrast material. The mass also invaded the left frontal lobe, the right orbit, and ethmoid sinus (Fig. 1). Right carotid angiography demonstrated an avascular mass in the right frontal region (Fig. 2A).

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Postoperative Course. The immediate postoperative course was smooth and uneventful. Five days after operation, histological examination showed that the tumor was a granuloma of fungal origin. Because of the characteristic brown pigment in the hyphae of the fungi, chromoblastomycosis was strongly suggested (Fig. 3). On the same day that the histological diagnosis was made, the patient became drowsy with intermittent high fever. Lumbar tap revealed cerebrospinal fluid (CSF)
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with marked pleocytosis; the cell count was 1280/cu mm and the protein level 140 mg/ml. No bacteria were found. On suspicion of fungal meningitis, amphotericin B was immediately given intravenously, but the patient's level of consciousness deteriorated gradually. Repeated culture of CSF did not demonstrate any bacteria or fungi.

On December 29, 1979, he became suddenly comatose with decerebrate posture, and CT revealed a right frontal intracerebral hematoma with ventricular rupture. The intracerebral hematoma was evacuated and external ventricular drainage was instituted. He continued to be comatose after the operation and meningitis did not improve in spite of administration of amphotericin B and 5-fluorocytosine. On January 7, 1980, fresh blood spurted through the external ventricular catheter. A CT scan demonstrated a massive intracerebral hematoma in the right frontal lobe with intra-
ventricular hemorrhage. We suspected rupture of a fungal aneurysm as a cause of the repeated intracranial hemorrhage, and cerebral angiography was performed. Right carotid angiography revealed at least four aneurysms; one situated at a proximal branch of the middle cerebral artery and three proximally at the anterior cerebral artery. Vasospasm was also evident (Fig. 2B). Left carotid angiography revealed no aneurysm, and vasospasm was not as severe as on the right.

The intracerebral hematoma was evacuated and the largest aneurysm situated at the branch of the middle cerebral artery was treated with a trapping procedure. The aneurysmal wall was too thin and fragile to obliterate with a clip. The other three aneurysms were left untreated. After the last operation, intracranial hemorrhage no longer occurred. Meningitis gradually subsided. Three weeks after the last operation, right carotid angiography demonstrated a small new aneurysm at the distal branch of the middle cerebral artery, which had not been demonstrated before; the four aneurysms that had previously been visualized had disappeared. The patient remained vegetative and died 2½ years after admission. Repeated attempts to confirm the nature of the fungus by CSF culture failed. Autopsy revealed widespread destruction of the brain, but showed no fungal lesions in the body.

Discussion

Although the decrease of bacterial infection in the central nervous system (CNS) has been paralleled by the increase of opportunistic fungal infection, which reflects the widespread and increased use of corticosteroids, cytotoxic agents, and antibiotics, a primary fungal granuloma in the CNS is still rare.11,19 This patient was quite a healthy young man until the onset of this disease, and there was no evidence of immunological deficiencies or use of any of the above-mentioned agents. The route and opportunity of fungal infection were not confirmed except for his work at a stock farm in Hokkaido during his college life.

There was neither a cutaneous nor a pulmonary lesion that is usually a precursor of invasion of the CNS by fungi, and we did not suspect the diagnosis of fungal granuloma prior to histological examination. Cerebral angiography revealed only evidence of an avascular mass, and CT showed no characteristic findings: only a high-density mass with an irregular margin, which was moderately enhanced after injection of contrast material. Fungal meningitis and rupture of a fungal aneurysm occurred after piecemeal resection of the granuloma. We believe that the surgical treatment was indicated in this case; the tumor was large enough to give a mass effect and there was no evidence of fungal infection in any other part of the body. If granuloma of fungal origin is suspected before or after surgery, it is essential to administer antifungal antibiotics as soon as possible to prevent fungal meningitis and its complications.

Chromoblastomycosis in the CNS is rare, and its chief pathological manifestations are abscess formation, granuloma, and arachnoiditis. The prognosis is poor, probably due to multiple lesions, ineffectiveness of antifungal antibiotics, and immunosuppression.13,17,19 Takahashi, et al.,19 described a case with granuloma of chromoblastomycosis origin involving the cisterna magna and spinal subarachnoid space. Shimura, et al.,17 reported a case with cerebral granuloma from chromoblastomycosis involving the skin, lung, and kidney.
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Both patients developed postoperative meningitis and died, although there was a temporary remission of symptoms. Musella and Collins reported a case with prior occurrence of nocardial brain abscess and immunological deficiency. Fungal aneurysm associated with chromoblastomycosis has not been reported previously in the literature.

Bacterial aneurysms develop secondary to infection of the arterial wall; they usually arise from infected emboli or, more rarely, from the surrounding infection. Fungal aneurysms, on the other hand, commonly develop secondary to fungal meningitis and other surrounding infection such as paranasal sinusitis. Mielke, et al. reviewed 15 cases of fungal aneurysms. The time course for development of fungal aneurysms and their rupture is lengthy (usually several months). Fungal aneurysms are usually found proximally on the major intracranial arteries, while bacterial aneurysms are usually found on the peripheral branches of the middle cerebral artery. Multiple aneurysms occur with about the same frequency as berry aneurysms. Fungal aneurysms are commonly associated with thrombosis in contiguous or remote arteries. Predisposing factors include leukemia, systemic lupus erythematosus, paranasal sinusitis, diabetes mellitus, and fungal endocarditis. In many of the reported cases, antibiotic and corticosteroid therapy was given. In the present case the site of the aneurysms was limited to the right side (the operative side), and only 12 days elapsed from the postoperative fungal infection to the rupture of the fungal aneurysm. Bacterial aneurysms commonly occur about 18 days after the diagnosis of bacterial endocarditis is made, but Bullock and Van Dellen reported a case in which a bacterial aneurysm ruptured 6 days after aortic valve replacement. Bacterial aneurysms may increase or decrease in size rapidly, in days or weeks, and for this reason repeated angiography is recommended. Such rapid change of aneurysm size has not previously been reported in fungal aneurysms.

Postoperative fungal meningitis and fungal aneurysms are rare, and only three cases have been reported. Visudhiphan, et al. reported a case with ruptured fungal basilar artery aneurysm of Aspergillus origin 6 weeks after total removal of craniopharyngioma. Sakaki, et al. described a case with fungal aneurysm of the intradural internal carotid artery and meningitis due to Aspergillus infection following resection of an arteriovenous malformation in the insular region. Mielke, et al. reported a case with ruptured basilar artery aneurysm (Candida and Aspergillus) 10 months after transphenoidal removal of a pituitary adenoma and irradiation for acromegaly.

The results of treatment of fungal aneurysms remain dismal so far. Not only rupture of these aneurysms, associated fungal meningitis, and widespread vascular involvement including thrombosis, but also the ineffectiveness of antifungal antibiotics and an immunosuppressive state may be factors responsible for the poor outcome.

References


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