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Case report

A case of bilateral vasculitis associated with pineal germinoma

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ABSTRACT

Purpose: To report a rare case of bilateral periphlebitis associated with a pineal germinoma.

Observations: A 17-year-old male teenager presented at a local clinic complaining of blurred vision in both eyes. The treating physician identified bilateral uveitis, and prescribed the patient with a local steroid treatment. However, the inflammatory findings did not improve with the treatment, and the patient was referred to our hospital for further examination. At the first visit, his best-corrected visual acuities were 0.3 for the right eye and 0.06 for the left eye; we found no inflammation in the anterior ocular segment, but observed bilateral retinal periphlebitis and a proliferative membrane from the papilla to the macula in the ocular fundus. In addition, we found a tractional serous retinal detachment in the macula. We suspected tuberculous uveitis clinically and initiated treatment with an antituberculous drug. However, the condition of the patient did not improve. Two months after our initial examination, left optic neuritis appeared, and we initiated a steroid pulse therapy. Although the periphlebitis remained, the left optic nerve findings and the visual acuity of both eyes improved. Thus, we reduced the oral steroid dose gradually. However, two months after initiating the dose reduction, the patient suffered a consciousness disturbance, and we detected a pineal tumor by magnetic resonance imaging (MRI). The patient was diagnosed as having a germ cell tumor by pathological examination and underwent radiation and chemotherapy. We noted marked improvements in both the periphlebitis findings and in the visual acuity following the treatment for the pineal tumor.

Conclusions and importance: Cases of pineal tumor accompanied with retinal periphlebitis have been reported rarely. Because juvenile retinal vasculitis cases of unknown cause can be associated with pineal germinomas, we recommend brain MRI examinations for such cases.

1. Introduction

Intracranial germ cell tumors develop preferentially in the pineal gland of young men, with the suprasellar region being the next most commonly affected site.[1] Different symptoms appear in accordance with the affected sites, and hydrocephalus occurs when the pineal body is affected. Although the suprasellar regions are more likely to lead to visual impairments, pineal regions may cause Parinaud's syndrome.

Here we report the case of a male teenager diagnosed as having a pineal germinoma during treatment for bilateral retinal periphlebitis.

2. Case report

A 17-year-old male teenager with no significant medical history visited a local clinic due to the loss of visual acuity in both eyes. Bilateral uveitis and macular edema were noted, and treatment with topical steroids was initiated. Because no improvement occurred following treatment, he was referred to our hospital. At the initial examination, the eye movements and light reflexes were normal; his best-corrected visual acuity was 0.3 for the right eye and 0.06 for the left eye, and we found no abnormalities in the anterior ocular segments. The mean critical flicker frequency values for the right and left eyes were 32 and 23 Hz, respectively, with a decrease in the left eye. We observed optic disc swelling and retinal periphlebitis in the fundi of both eyes, the formation of a proliferative membrane around the optic nerve, and tractional serous retinal detachment in the macula (Fig. 1A and B, E, F). Using optic coherence tomography (OCT), we found hyperrefractive foci in the retina and in the vitreous cavity of both eyes (Fig. 1E and F). The fluorescein fundus angiography showed dye leakage from the optic nerves and dye staining of the vein wall, and perivascular dye leakage throughout the retina in both eyes (Fig. 1C and D). Blood tests were normal. The levels of serum angiotensin-

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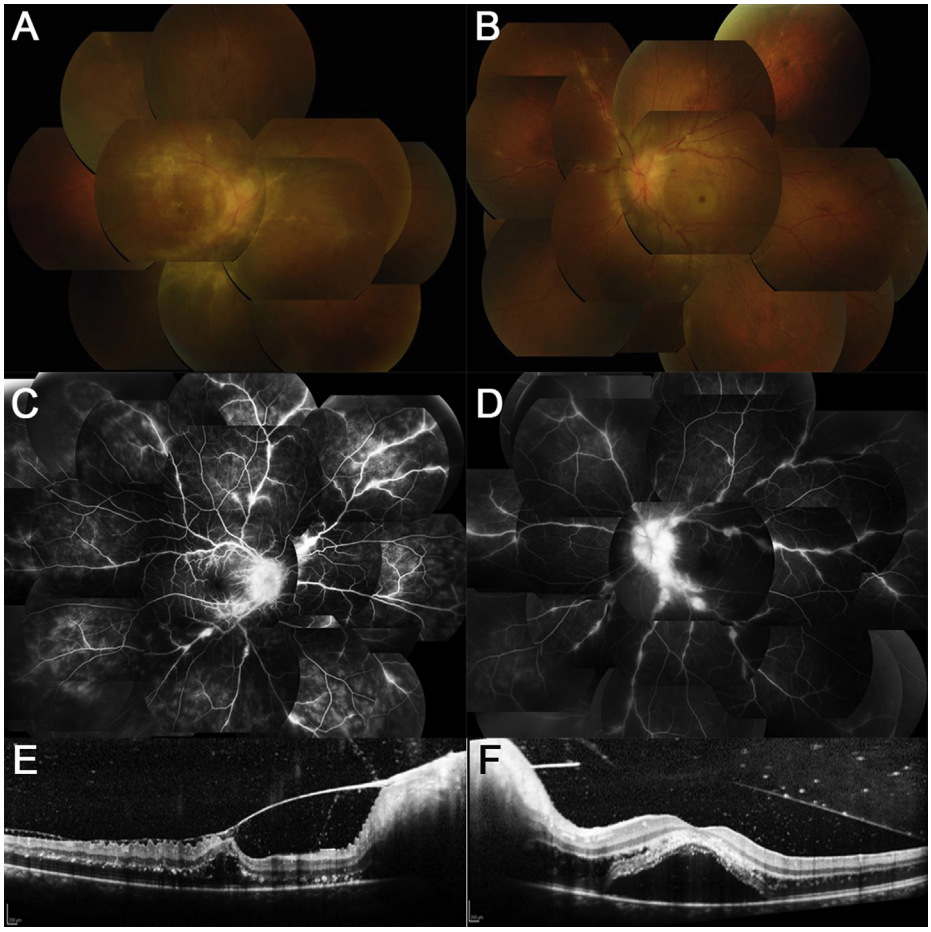

Fig. 1. Color fundus photographs (A, B), fluorescein angiography (C, D), and optical coherence tomography (E, F) images of both eyes at initial presentation. (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)

converting enzyme activity and serum lysozyme, which are high in sarcoidosis, were normal. The *Bartonella* and cardiolipin antibody tests were negative; and the QuantiFeron test (QuantiFERON®TB Gold In-Tube; Qiagen, Germantown, USA) was also negative. The chest radiography and electrocardiography images were also normal.

We thought sarcoidosis was unlikely based on the examination findings; however, tuberculous uveitis was possible based on the lack of response of the patient to steroid treatment and the fundus findings. Thus, we prescribed oral isoniazid (an antituberculous drug). However, the ocular symptoms did not improve with the treatment, and we observed the exacerbation of the swelling of the left optic nerve papillary and reduced left visual acuity (best-corrected visual acuity 0.03) 2 weeks after the initiation of treatment. We diagnosed the left optic neuritis and started an intravenous steroid pulse therapy (methylprednisolone 500 mg × 3 days). Although the retinal periphlebitis findings remained the same after the therapy, the optic nerve swelling and the visual acuity improved (Fig. 2A and B). The oral steroid dose was then gradually decreased; however, a disturbance of consciousness occurred 2 months after starting the dose reduction. A brain MRI at the time confirmed a pineal tumor (Fig. 3). We performed an endoscopic ventriculostomy and obtained a pineal tumor biopsy, and the diagnosis of germinoma was made based on the pathological examinations. Further examinations revealed no signs of metastasis, and chemotherapy and radiotherapy were initiated. At the post-treatment re-examination, the periphlebitis findings in the fundus of both eyes had markedly improved (Fig. 2C–F), and the visual acuity was further enhanced.

3. Discussion

Intracranial germ cell tumors develop preferentially in the pineal gland and the suprasellar regions of male teenagers or young adults. In a report on cases of intracranial germinoma accompanied with ocular symptoms, the tumors were located in the suprasellar region in two out of the three cases and in the suprasellar region and the pineal body in the remaining case.[2] In all the three cases, atrophy of the optic nerves of both eyes was observed in the fundus examinations, causing decreased visual field disturbances and loss of acuity in both eyes.[2] In that report, both the decreased visual field disturbance and loss of acuity were attributed to the direct invasion of the tumor of the optic chiasm and optic nerve.

We found two reports of cases in which germ cell tumors developed in the pineal body.[3,4] In both the cases, patients were also male teenagers (14-year-old), in whom pineal germinoma caused optic disc swelling with retinal periphlebitis, and the fundus findings improved markedly after treatment for germinoma.

The treatment course of our patient suggests that in cases of combined bilateral optic disc swelling and retinal periphlebitis of pineal germinoma, the following two pathological conditions exist: a direct tumor invasion and a paraneoplastic syndrome due to the remote effects of cancer. Because the lesions involved the optic nerves and blood vessels, we could not sample them. Our patient experienced a disturbance of consciousness after the steroid pulse therapy. The inhibition of the immunoreaction against the germinoma itself by the applied pulse steroid might have caused the episode. For cancer-associated retinopathy (CAR), known as a paraneoplastic syndrome, antiretinal antibodies are known to be involved in the pathological condition, and the