Introduction

Kikuchi-Fujimoto disease is known to be a self-limited systemic disease with fever and cervical lymphadenopathy, and it is confirmed by characteristic histopathologic findings of necrotizing lymphadenitis of the affected lymph node. Although steroids are administered systemically, there is controversy about the efficacy of the steroids, and it is known that most of the fundus abnormalities recover rapidly and normal vision is restored and there is no recurrence. We report a case of Kikuchi-Fujimoto disease associated with bilateral frozen giant vasculitis and report it.

Case report

A 33-year-old man visited ophthalmology clinic due to decreased visual acuity in both eyes 5 days ago. He was diagnosed with acute pyelonephritis in a private physician. The best corrected visual acuity at the time of initial diagnosis was 0.1 in the right eye and 0.2 in the left eye. On fundus, a diffuse vascular sheathing like frosted branch was found in the retinal vessels and retinal hemorrhage was observed, which was found in the posterior and peripheral retina of both eyes. There was no abnormal blood flow in the retina and choroid in the fluorescein angiography. Fluorescein angiogram showed staining and leakage of dye along the vascular sheathing (Fig.1).

The location of the lesion was at risk for resection biopsy, and a fine-needle aspiration biopsy was performed under ultrasound guidance. Necrotizing lymphadenitis was observed on histology (Fig.2).

Figure 2. Histopathological section of cervical lymph node shows numerous phagocytic histiocytes and prominent karyorrhexis (H&E, x100).

After the diagnosis, the antibiotic injection was stopped, and methylprednisolone 500 mg/day was injected intravenously. The onset of fever and headache recovered and the visual acuity recovered. After 1 month, the best corrected visual acuity recovered to 1.0 in the right eye and 0.8 in the left eye, and the fundus findings were normal (Fig.3).

Figure 3. Fundus photographs at 1 month after systemic steroid therapy shows almost normal without any sheathing of retinal vessels in right (A) and left (B) eyes.

Discussion

It was first reported in 1972 by the Japanese doctor Kikuchi and Fujimoto, and it is a histiocyte necrotizing lymphadenitis that occurs in young Asian people under 30 years old. In the present case, the patient had difficulty in diagnosing cervical lymph node hypertrophy at the time of initial diagnosis, because of the fat layer, and because the location of the enlarged lymph node was high near the neck vein. Biopsy was not performed, ultrasonically induced fine-needle aspiration was performed, and necrotizing lymphadenitis was confirmed. Clinical findings include various inflammatory findings in the anterior and vitreous body, and distinctive vascular anastomoses in a relatively uniform shape extending from the posterior to the periphery of the retina are observed. In conclusion, additional examination is needed for patients with Frosted Branch Angitis. Because they may be accompanied by systemic diseases such as Kikuchi diseases.