Takayasu Disease on the Centenary of Its Discovery

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Abstract

Takayasu disease was first reported in 1908 by Mikito Takayasu as “a case of peculiar changes in the central retinal vessels.” Because in these patients the pulse of the radial artery is impalpable, investigations focusing on the ischemic symptoms of the upper body were conducted. In 1948, Shimizu and Sano named this pathological condition “pulseless disease.” Since then, the lesions of Takayasu disease have been detected not only in the aortic arch and its main branches but also in various vessels, including the abdominal aorta and renal arteries. The ocular symptoms of Takayasu disease are considered to be due to ischemia in the retina and choroid. The typical wreath-like arteriovenous anastomosis around the disc reported by Takayasu is observed at a relatively late stage of the disease. The characteristic fundus findings of Takayasu disease include tortuosity and dilatation of the central retinal artery and vein, retinal arteriovenous anastomosis, prominent retinal vasculature, microaneurysms in the capillaries, occlusion of retinal arterioles, soft exudate, choked disc, and optic atrophy. Fluorescein angiography reveals retinal microaneurysms, sludging, slower blood flow, dilatation of retinal vessels, leakage of fluorescein dye due to increased vascular permeability, and arteriovenous anastomosis. Arteriovenous anastomosis initially appears in the periphery at the early stage, and in the arteriovenous crossing at the advanced stage. Systemic administration of corticosteroids is required to prevent vascular stenosis during the early stages of Takayasu disease. Reconstruction of the carotid artery may improve subjective symptoms and fundus findings.

Key Words: arteriovenous anastomosis, pulseless disease, retinal microaneurysm, Takayasu disease, Takayasu’s arteritis

Introduction

Takayasu disease was first reported by Mikito Takayasu (Fig. 1), a Japanese ophthalmologist, at the 12th Annual Meeting of the Japanese Ophthalmological Society in 1908 as “a case of peculiar changes in the central retinal vessels.”¹ At the same meeting, Yoshiakira Ohnishi presented “a case with similar changes in the fundus accompanying impalpable pulse of the radial artery.”¹ In the same year, Takayasu’s original article was published in the Journal of the Juzen Medical Society, Kanazawa Medical School.² In 1921, Nakajima³ proposed “Takayasu disease” as a new disease based on findings reported by Takayasu and Ohnishi as well as his own experiences. Since then, Japanese ophthalmologists have reported many cases of this disease.

In 1948, two neurosurgeons, Shimizu and Sano, named this pathological condition pulseless disease,⁴ focusing on the ischemic symptoms of the upper body. Subsequent studies showed the lesions of Takayasu disease to be present not only in the aortic arch and its main branches but also in various vessels, including the abdominal aorta and renal arteries. The ocular symptoms of Takayasu disease seemed to be caused by chronic ischemia in the retina and choroid resulting from poor blood circulation associated with either stenosis or occlusion of the aortic arch and carotid arteries. Recently, early diagnosis and treatment of Takayasu disease by physicians and surgeons has resulted in a substantial reduction of advanced cases. In this article, we review the history and recent advances in Takayasu disease and consider the current status and future directions of this disease.
First Case Report

Case: A 22-year-old woman (married). Initial consultation: 8 May 1905.2

Medical History

The patient had noticed a gradual loss of vision in both eyes, starting the previous September. She occasionally suffered redness of the conjunctiva. In February, she received treatment and recovered her vision. In mid-March, however, she noticed a gradual loss of vision OD, followed about 10 days later by poor vision OS. She had no other medical or gynecological problems.

Findings

There were no particular findings in the anterior segments. The pupils were slightly dilated and the light reflex sluggish.

OD, the ocular media were normal although marked abnormalities were found in the retinal vessels. The retinal vessels branched 2–3 mm away from the disc, and the branches anastomosed to one another to create a wreath around the disc (wreath-like anastomosis). The vessel radially branches at the anastomotic site, and thin branches running for some distance can be vaguely observed. In the area near the disc, some vessels reach the vitreous body. Blot hemorrhages are observed near the injected disc.

Figure 1. Portrait of Mikito Takayasu. He was born in Saga Prefecture, Japan, in 1860. He graduated from Tokyo Imperial University in 1887, became chief ophthalmologist of the Fourth Advanced Junior High School Department of Medicine in Kanazawa the next year, and was appointed head of the hospital of the school in 1894. He went to Germany to study in 1899, then returned to Japan and was appointed head of the Kanazawa Medicine Special School in 1901. He became Dean of Kanazawa Medical University in 1923 and retired from the university the next year. He died at the age of 79 years in 1938.

Figure 2. Inverted image of fundus schema of the right eye from “A case with curious change in the central retinal vessel” described by Takayasu.2 The retinal vessel is branched 2–3 mm away from the disc, and the branches anastomose to one another to create a wreath around the disc (wreath-like anastomosis). The vessel radially branches at the anastomotic site, and thin branches running for some distance can be vaguely observed. In the area near the disc, some vessels reach the vitreous body. Blot hemorrhages are observed near the injected disc.