

Ruptured pediatric posterior cerebral artery aneurysm 9 years after the onset of Kawasaki disease: a case report

Satoshi Tanaka · Takao Sagiuchi · Ikuo Kobayashi

Received: 14 July 2006 / Revised: 18 August 2006 / Published online: 2 December 2006
© Springer-Verlag 2006

Abstract

Case report A 12-year-old boy who had a history of Kawasaki disease 9 years ago experienced a subarachnoid hemorrhage by ruptured right posterior cerebral artery aneurysm. On day 1 operation, as the aneurysm was very fragile and bled easily, two intraoperative ruptures, including a very premature rupture, were encountered. As a result, a left hemiparesis especially severe in the left hand was caused by the right anterior thalamic infarction due to the occlusion of a thalamo-perforating artery arising near the neck of the aneurysm.

Discussion The histopathological examination of the intraoperative excised aneurysmal dome disclosed the thickening of the endothelial inner due to the endothelial hypertrophy and the invasion of inflammatory cells. This finding of the aneurysm was partially mimicking the finding of the coronary artery of the patients with Kawasaki disease. The combination of cerebral aneurysm and Kawasaki disease has never been reported until now, and the etiology of the aneurysm of this patient is unclear.

Keywords Pediatric cerebral aneurysm · Posterior cerebral artery · Kawasaki disease

Introduction

Kawasaki disease is an acute self-limited vasculitis in childhood, which is characterized by fever, bilateral non-exudative conjunctivitis, erythema of the lips and oral mucosa, changes in the extremities, rash, and cervical lymphadenopathy [11]. Coronary artery aneurysms or ectasia develop in approximately 15 to 25% of untreated children and may lead to ischemic heart disease or sudden death [16].

In spite of the generalized vascular disease, the combination of cerebral aneurysm and Kawasaki disease had not been reported. Almost all cerebral aneurysms detected in childhood seemed to be congenital [7]. The inflammatory cerebral aneurysms had been rare in children [4]. In contrast, the recent reports documented that dissecting aneurysms were frequent in children [13, 19].

We observed the pediatric posterior cerebral artery (PCA) aneurysm ruptured 9 years after the onset of Kawasaki disease. The relation of the cause and effect of this aneurysm and Kawasaki disease is still unclear, although the histopathological examinations of the aneurysmal dome have disclosed the inflammatory feature mimicking the finding of the coronary involvement in Kawasaki disease.

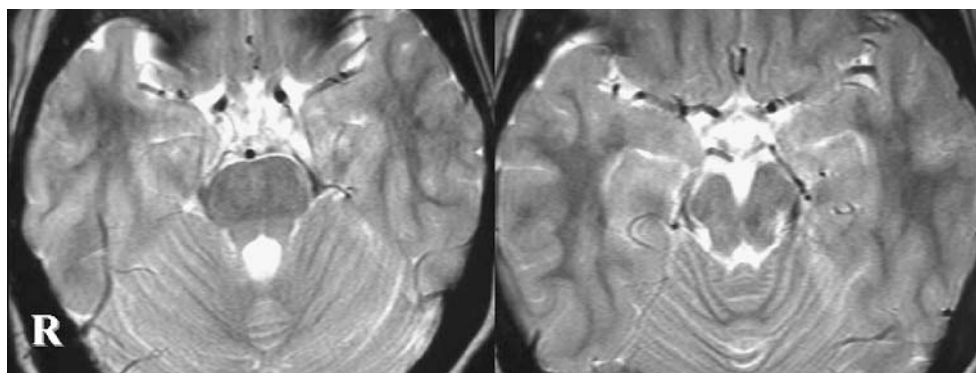
Case report

A boy 12 years and 11 months old visited the emergency room of the Kitasato Institute Medical Center Hospital on April 3, 2005 due to severe headache since 12 h before. He had a history of cerebral palsy since 1 year old and Kawasaki disease since 3 years old. He underwent an operation for spasticity twice, at 7 and at 10 years old. He

S. Tanaka (✉)
Department of Neurosurgery, Kawasaki Hospital,
2040 Kisaki-nicho,
Hitachiohta, Ibaraki 313-8511, Japan
e-mail: stanaka-nsu@umin.net

T. Sagiuchi · I. Kobayashi
Department of Neurosurgery,
The Kitasato Institute Medical Center Hospital,
Kitamoto 364 8501, Japan

Fig. 1 T2-weighted image of magnetic resonance imaging on February 2002 in our hospital showed no abnormal finding especially in PCA territory



had been healthy and had walked to a school for physically handicapped or mentally retarded children by himself before the onset. In February 2002, magnetic resonance imaging (MRI) in our hospital showed no abnormal findings (Fig. 1). In December 2004, he visited our hospital due to a high fever from an unknown etiology for 3 days, and the ultrasound cardiogram (UCG) showed a mild dilatation of the left main trunk (LMT) of the coronary artery at this point (4.6 mm in diameter), but no other cardiac abnormality had been detected.

His computer-assisted tomography (CT) on admission showed a thin subarachnoid hemorrhage in the interpeduncular cistern and right ambient cistern (Fig. 2). The three-dimensional CT angiography disclosed a saccular cerebral aneurysm arising from the right PCA and the right posterior communicating artery junction (Fig. 3). The UCG revealed

that his cardiac function was normal with 73% ejection-fraction rate but with a mild dilatation of the LMT of the coronary artery (4.3 mm in diameter).

The operation for neck clipping was done on April 4, 2005 (day 1) through right subtemporal route. A linear skin incision was made, extending vertically upward originating at the zygomatic process of the temporal bone approximately one finger's width anterior to the ear. The bony opening was about 4 cm across at the base and 3 cm high. After the dural opening, the tight right temporal lobe was gently elevated. Twelve minutes after the introduction of the microscope, just when the bloody cerebro-spinal fluid was removed, the premature rupture of the aneurysm happened. The bleeding continued for 13 min, and the hemostasis was completed by dome clipping under the application of a temporary clip to the basilar artery for 5 min. Then, the

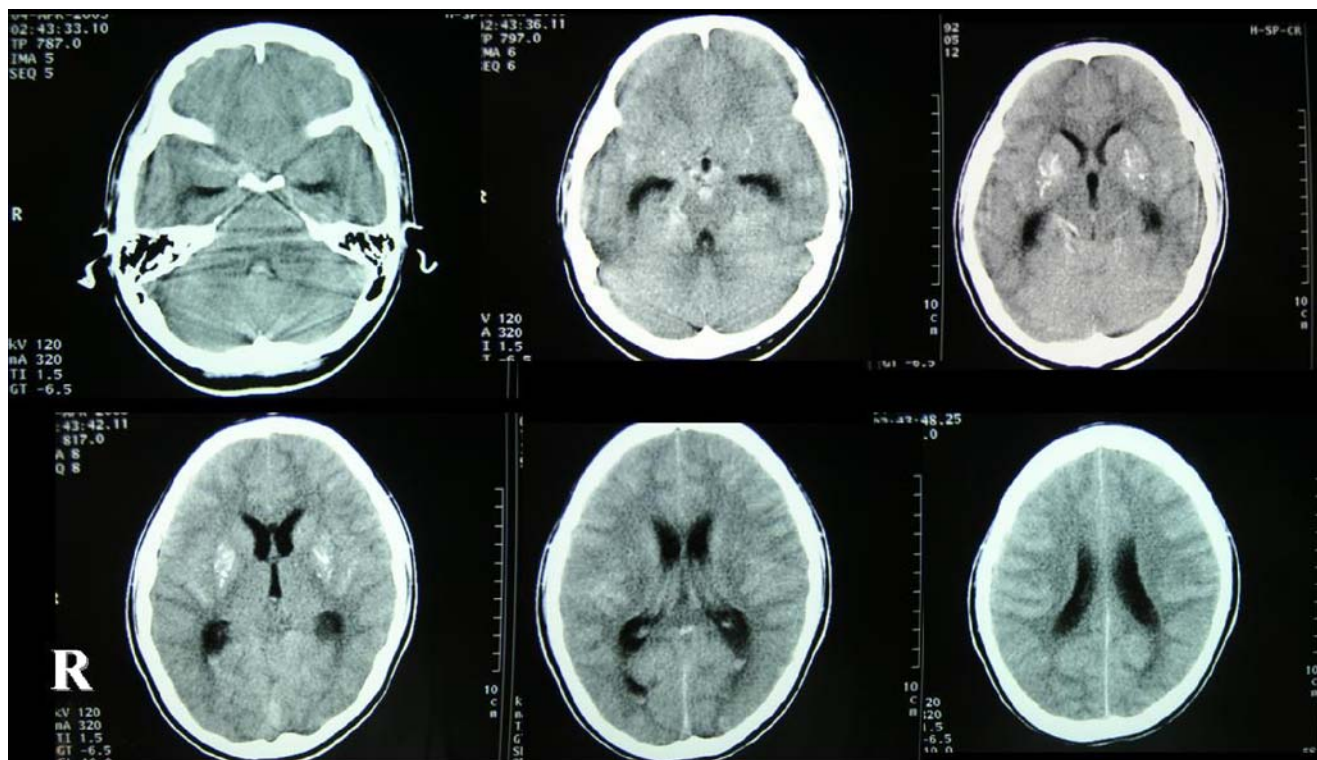
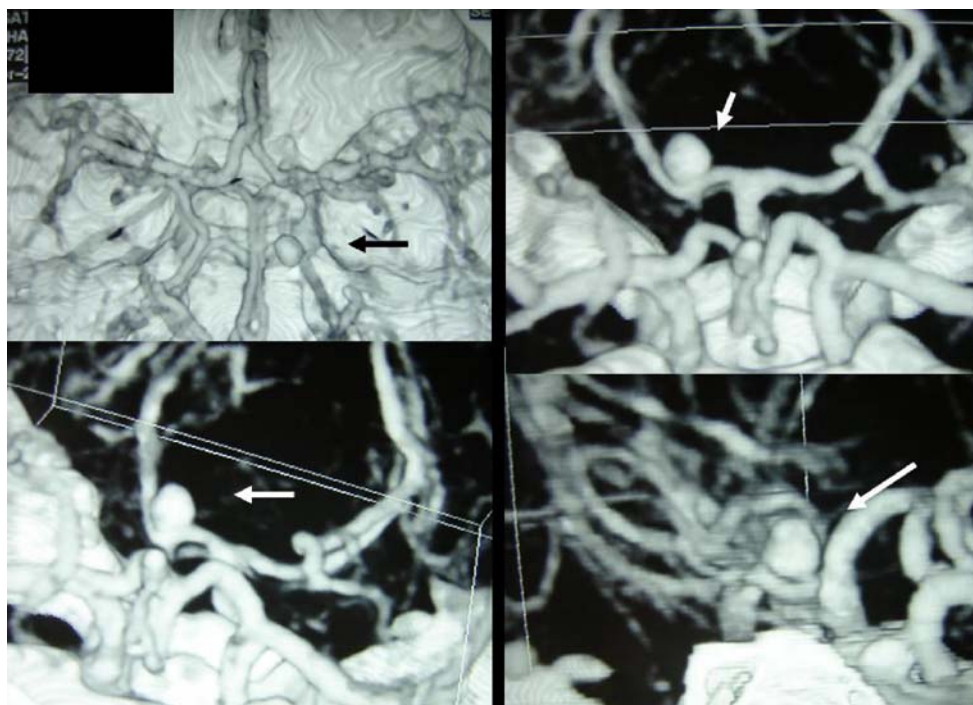


Fig. 2 The computer-assisted tomography of the presented patient on admission showed a thin subarachnoid hemorrhage in the interpeduncular cistern and right ambient cistern

Fig. 3 The three-dimensional computer-assisted tomographic angiography disclosed a saccular cerebral aneurysm (*arrow*) arising from the right posterior cerebral artery and the right posterior communicating artery junction



ambient cistern was widely opened, and the neck clipping was performed under the temporary occlusion of the right PCA. The fenestrated titan clip was applied to avoid the occlusion of thalamo-perforating arteries. Before the final neck clipping, the bleeding happened again near the aneurysmal neck, which had been unexpectedly wide, and then a perforating artery was occluded by the final clip as shown in Fig. 4. After the final clipping, the aneurysmal dome that had been very fragile was partially resected for histopathological examination.

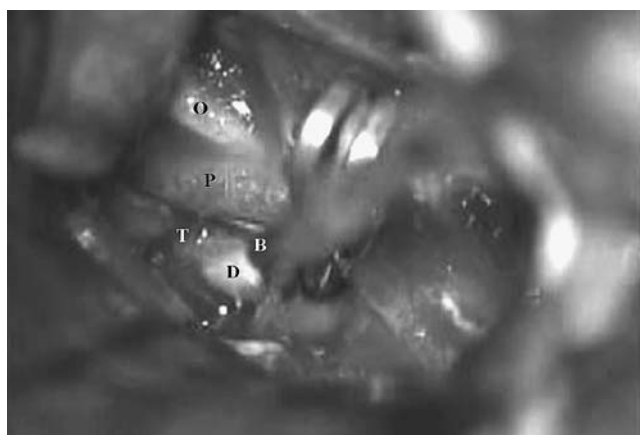


Fig. 4 The photograph of the operation disclosed that a fenestrated clip was applied to the relatively wide neck of the aneurysm and a thalamo-perforating artery was occluded by the clip. *O* right oculomotor nerve; *P* right posterior cerebral artery; *T* the thalamo-perforating artery; *B* blades of the fenestrated clip; *D* dome of the aneurysm

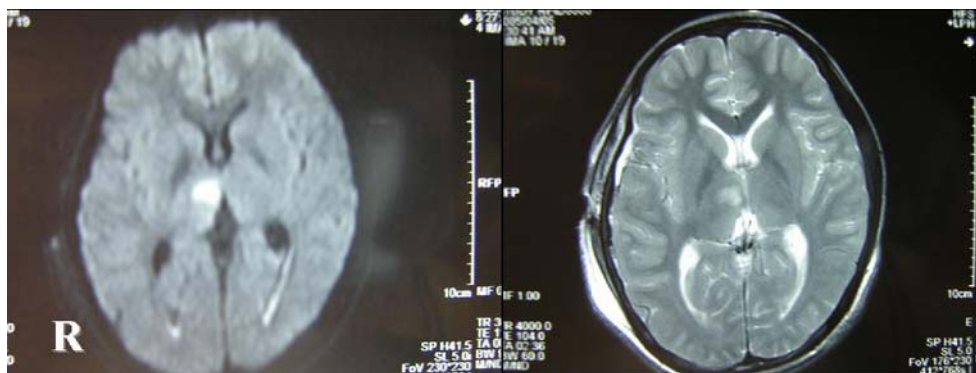
As the two intraoperative ruptures of the aneurysm and blood transfusion introduced danger of instability of a general condition, he received a mild hypothermia therapy for 3 days postoperatively. The right complete oculomotor palsy with ptosis was observed immediately postoperatively in spite of the general anesthesia during a mild hypothermia therapy. The diffusion tensor image and T2-weighted image of MRI 1 day after the operation revealed a cerebral infarction in the right anterior thalamus due to occlusion of the thalamo-perforating artery as shown in Fig. 5. Vertebral angiogram 2 days after the operation showed the complete occlusion of the aneurysm (Fig. 6). After the extubation on day 7 of subarachnoid hemorrhage, he presented left hemiparesis especially in the upper extremity. On May 17, 2005, he was transferred to the prefectural rehabilitation center. After 3 months of intensive rehabilitation, his right oculomotor palsy was completely healed, and he can gait with some assistance now.

The histopathological examination of small specimen of the aneurysmal dome disclosed the thickening of the endothelial inner due to the endothelial hypertrophy and invasion of inflammatory cells including granulocytes in the wall of the dome (Fig. 7).

Discussion

Intracranial aneurysms in children are rare, and their epidemiology is poorly understood. A review of the literature since 1939 were performed by Huang et al. [10].

Fig. 5 The diffusion tensor image (*left*) and T2-weighted image (*right*) of magnetic resonance imaging 1 day after the operation revealed the cerebral infarction in the right anterior thalamus due to occlusion of the thalamo-perforating artery



A total of 706 pediatric intracranial aneurysm cases had been described since 1939. The men/women ratio is 1.8:1. The internal carotid artery bifurcation was the location of 26%, and only 17% were in the posterior circulation. Twenty percent were giant lesions, and 80% presented with subarachnoid hemorrhage. Good- and poor-grade patients comprise 49 and 36% of the total cases, respectively. A favorable outcome was reported in 60%. Seventy-one cases in children under 5 years of age found in the literature, plus one case of their own, were analyzed by Ferrante et al. [7]. In early childhood, these aneurysms seem to have features that distinguish them from the same variety in adults. In children, a congenital pathogenesis appears more convincing because of the higher frequency in the first 2 years of life, the often peripheral site (41.6%), the high frequency of large (50%) or giant (26.8%) aneurysms, and the association with other cerebral and vascular congenital abnormalities. Otherwise, the most pediatric aneurysms are related either to infection/inflammation or trauma and are not congenital. The onset was non-hemorrhagic in 18.1% of cases compared with 2.5% in adults. The surgical outcome seems better in children than in adult patients in terms of both morbidity and mortality.

Kawasaki disease is an acute self-limited vasculitis in childhood that is characterized by fever, bilateral nonexudative conjunctivitis, erythema of the lips and oral mucosa, changes in the extremities, rash, and cervical lymphadenopathy [11]. In the 1960s, some authors had reported this

disease as nodular periarteritis or periarteritis nodosa [3]. A nationwide survey between 1970 and 1980 in Japan by Kawasaki [11] revealed 26,543 cases including 228 deaths. Although the differential diagnosis with infantile periarteritis nodosa was difficult, he emphasized that angitis and aneurysm of coronary arteries accompanied by thrombosis were frequent findings.

Histopathology, distribution, and incidence of vascular changes were studied in tissues obtained at autopsy on 37 children [1]. Elastic and musculoelastic arteries showed a high incidence of arteritic changes; however, the degree of the lesions was, in general, mild to moderate with the exception of the iliac artery, which revealed severe changes and a necrotizing panarteritis which was often accompanied with an aneurysm formation. A high incidence of arteritic changes, of which the main histological feature was necrotizing panarteritis, was seen in extravisceral middle-sized arteries. The coronary artery in particular was involved in each case and mostly had an accompanying aneurysm, some of which had ruptured. Coronary artery aneurysms or ectasia develop in approximately 15 to 25% of untreated children and may lead to ischemic heart disease or sudden death in Kawasaki disease [16].

Central nervous system involvements in Kawasaki disease were reported [2, 6, 8, 18]. Meningitis, facial palsy, and subdural effusion have been reported to be some of the central nervous system complications of Kawasaki disease. Also, cerebro-vascular disease such as cerebral infarction

Fig. 6 The anterior–posterior view (*left*) and lateral view (*right*) of vertebral angiogram on 2 days after the operation showed the complete occlusion of the aneurysm

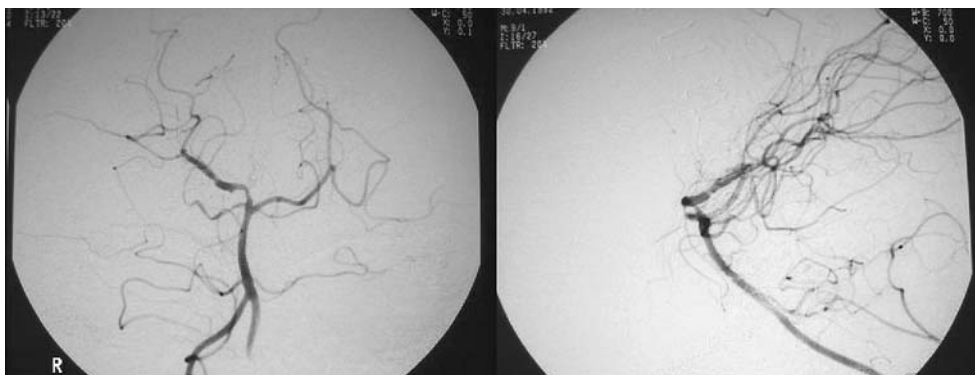
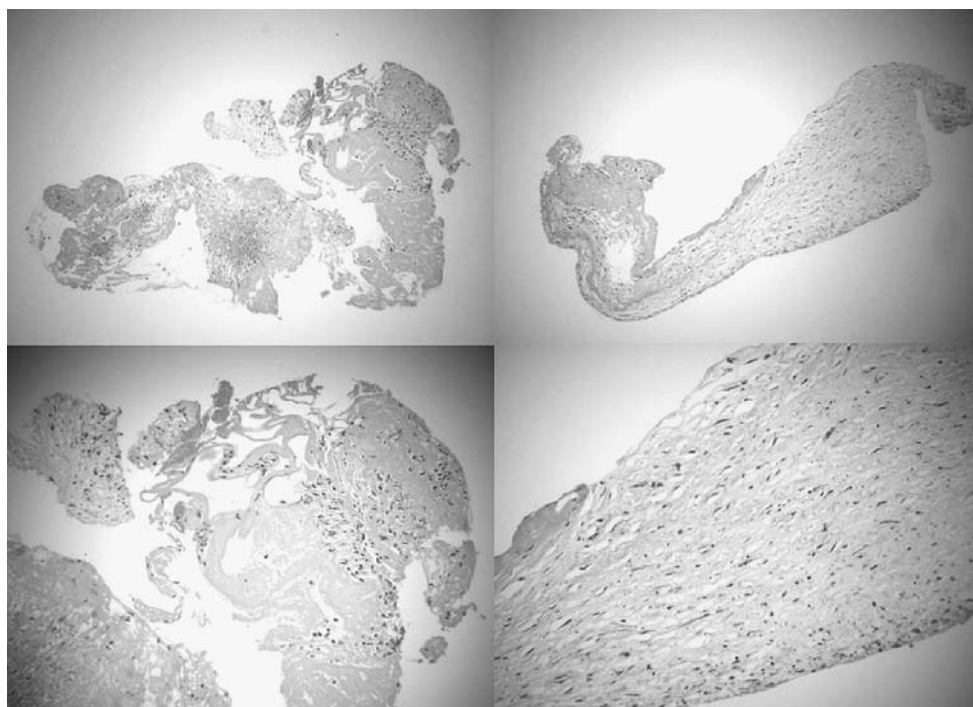


Fig. 7 The histopathological examination of small specimen of the aneurysmal dome disclosed the thickening of the endothelial inner due to endothelial hypertrophy and the invasion of inflammatory cells including granulocytes in the wall of the dome. $\times 100$, upper; $\times 200$, lowerleft; $\times 400$, lowerright



was often reported to happen. Histopathological examination in autopsies of the patients with Kawasaki disease sometimes revealed cerebro-vascular changes, such as endoarteritis, periarteritis, perivascular cuffing, and the focal necrotizing vasculitis [1]. In contrast, the cerebral aneurysm accompanied with Kawasaki disease has never been reported until now.

The presented PCA aneurysm was not a congenital one because the MRI 2 years ago had disclosed no abnormal finding on PCA. The mycotic formation of the aneurysm was also neglected because there was no definitive history of endocarditis and sepsis [4]. There had been no episode of trauma in the patient. It cannot be completely denied the possibility of a dissecting aneurysm. The dissecting aneurysms in vertebro-basilar systems seemed to be very rare in children, and they usually had developed as ischemic stroke [14]. Recently, Lasjaunias et al. [13] documented that dissecting aneurysms were frequent in children of all ages with either associated thrombosis or arterial tear with repeated acute hemorrhage and poor outcome. They reported that 33 children had dissecting aneurysm among 59 consecutive children with cerebral symptomatic aneurysms.

Rare association of coronary artery aneurysms with intracranial aneurysms was reported [9]. Yoneoka et al. [20] retrospectively investigated 12 cases drawn from 483 consecutive cases of aneurysmal subarachnoid hemorrhage at our institute over a period of 22 years, in which a previously undemonstrated (hence de novo) intracranial saccular aneurysm formed and ruptured after successful treatment of a prior aneurysm. The 12 cases constitute 2.5%

of the 483 patients who left the hospital alive. None of these ruptured de novo aneurysms was at the same location as the original lesion.

The histopathological findings of the aneurysmal wall in the presented patient revealed the thickening of the endothelial inner due to the endothelial hypertrophy and the invasion of inflammatory cells including granulocytes in the wall of the dome. These findings seem to be mimicking the acute inflammatory changes of the coronary artery in Kawasaki disease. Chyatte et al. [4] investigated whether inflammation or immunological reaction occurred in cerebral aneurysms. They concluded that extensive inflammatory and immunological reactions are common in unruptured intracranial aneurysms and may be related to aneurysm formation and rupture. Subintimal proliferation and inflammation were also common in dissecting aneurysms [12].

Ultimately, the etiology of the ruptured aneurysm of the presented patient and the relation between aneurysm formation and Kawasaki disease is unclear. Drake et al. [5, 17] had surgically treated posterior-circulation aneurysms mainly through the subtemporal route since 1960s. In contrast, recently, endovascular treatment has been increased especially in vertebro-basilar aneurysm, which was difficult to approach surgically [15]. For this patient, endovascular treatment may have been a better option. The authors had selected craniotomy because resection of the aneurysm was necessary not only for pathological examination but also for the detection of the mycotic aneurysm. The correlation of Kawasaki disease and cerebral aneurysm will still be unclear, as the specimen from aneurysm in children will be difficult to obtain when decreasing direct surgery.

References

- Amano S, Hazama F, Hamashima Y (1979) Pathology of Kawasaki disease: II. Distribution and incidence of the vascular lesions. *Jpn Circ J* 43:741–748
- Amano S, Hazama F (1980) Neuronal involvement in Kawasaki disease. *Acta Pathol Jpn* 30:365–373
- Benyo RB, Perrin EV (1968) Periarthritis nodosa in infancy. *Am J Dis Child* 116:539–544
- Chyatte D, Bruno G, Desai S, Todor DR (1999) Inflammation and intracranial aneurysms. *Neurosurgery* 45:1137–1146
- Drake CG (1961) Bleeding aneurysms of the basilar artery. Direct surgical management in four cases. *J Neurosurg* 18:230–238
- Engel DG, Gospe SM Jr, Tracy KA, Ellis WG, Lie JT (1995) Fatal infantile polyarteritis nodosa with predominant central nervous system involvement. *Stroke* 26:699–701
- Ferrante L, Fortuna A, Celli P, Santoro A, Fraioli B (1988) Intracranial arterial aneurysms in early childhood. *Surg Neurol* 29:39–56
- Fujiwara S, Yamano T, Hattori M, Fujiseki Y, Shimada M (1992) Asymptomatic cerebral infarction in Kawasaki disease. *Pediatr Neurol* 8:235–236
- Harikrishnan S, Stigimon J, Tharakan JM (2005) Intracranial aneurysms, coronary aneurysms and descending aortic coarctation—unreported association. *Int J Cardiol* 18:329–330
- Huang J, McGirt MJ, Gailloud P, Tamargo RJ (2005) Intracranial aneurysms in the pediatric population: case series and literature review. *Surg Neurol* 63:424–432
- Kawasaki T (1982) Kawasaki disease. Mucocutaneous lymph node syndrome or MCLS. *Acta Pathol Jpn* 32:63–72
- Krings T, Piske RL, Lasjaunias PL (2005) Intracranial arterial aneurysm vasculopathies: targeting the outer vessel wall. *Neuroradiology* 47:931–937
- Lasjaunias P, Wuppapapati S, Alvarez H, Rodesch G, Ozanne A (2005) Intracranial aneurysms in children aged under 15 years: review of 59 consecutive children with 75 aneurysms. *Childs Nerv Syst* 21:437–450
- Massimi L, Moret J, Tamburrini G, Di Rocco C (2003) Dissecting giant vertebro-basilar aneurysms. *Childs Nerv Syst* 19:204–210
- Nagashima H, Kobayashi S, Tanaka Y, Hongo K (2004) Endovascular therapy versus surgical clipping for basilar artery bifurcation aneurysm: retrospective analysis of 117 cases. *J Clin Neurosci* 11:475–479
- Newburger JW, Takahashi M, Gerber MA, Gewitz MH, Tani LY, Burns JC, Shulman ST, Bolger AF, Ferrieri P, Baltimore RS, Wilson WR, Baddour LM, Levison ME, Pallasch TJ, Falace DA, Taubert KA (2004) Diagnosis, treatment, and long-term management of Kawasaki disease: a statement for health professionals from the Committee on Rheumatic Fever, Endocarditis, and Kawasaki Disease, Council on Cardiovascular Disease in the Young, American Heart Association. *Pediatrics* 114:1708–1733
- Peerless SJ, Hernesniemi JA, Gutman FB, Drake CG (1994) Early surgery for ruptured vertebrobasilar aneurysms. *J Neurosurg* 80:643–649
- Terasawa K, Ichinose E, Matsuishi T, Kato H (1983) Neurological complications in Kawasaki disease. *Brain Dev* 5:371–374
- Vilela P, Goulao A (2006) Paediatric dissecting posterior cerebral aneurysms: report of two cases and review of the literature. *Neuroradiology* 48:541–548
- Yoneoka Y, Takeda N, Akira I, Ibuchi Y, Kumagai T, Sugai T, Takeda KI, Ueda K (2004) Ruptured de novo intracranial aneurysms. *Acta Neurochir (Wien)* 146:979–981