CASE REPORT

Intracranial aneurysms in twins: case report and review of the literature

HK Leung 梁浩楷
Y Lam 林　銳
KM Cheng 鄭建明
CM Chan 陳智明
YL Cheung 張毓靈

Intracranial aneurysm in twins is a rare clinical disease entity. Only 15 cases have been described in the literature. We report on a pair of identical twins with intracranial aneurysms. One presented with subarachnoid haemorrhage; digital subtraction angiography showed a left posterior communicating artery aneurysm, which was treated by coiling. The patient's twin sister was called for screening, whereupon digital subtraction angiography revealed a right ophthalmic internal carotid artery aneurysm that was treated conservatively.

Introduction

Subarachnoid haemorrhage (SAH) carries significant morbidity and mortality. In a recent local study, it was found to afflict relatively young subjects (with a mean age of 59 years).[1] Intracranial aneurysm (IA) is the commonest cause of SAH. Both environmental and genetic factors are involved in the development of IA, with familial occurrence having been well documented in the literature.[2] Aneurysms occurring in both identical twins are particularly interesting as this raises the issue of screening for asymptomatic twin. We describe here a pair of monozygotic twins, both with IA but in mirror position, and review the literature on this topic.

Case reports

Case 1

A 57-year-old woman presented with sudden 'collapse' while attending a pop music concert on 19 December 2005. She recovered in 2 minutes and complained of headache, neck stiffness, and dizziness. Upon admission, her Glasgow Coma Scale (GCS) score was 15. Examination revealed no cranial nerve deficit and power in her limbs was full. Computed tomography of brain showed a SAH. Digital subtraction angiography (DSA) revealed a 2-mm posterior communicating artery aneurysm (Fig a). Coiling of the aneurysm was performed. She was discharged on day 25 with a full GCS score and no neurological deficit. She was followed up in our clinic; magnetic resonance angiogram 12 months after the surgery showed no recurrence.

Case 2

The index patient's younger sister, who was entirely asymptomatic and had no abnormal neurological signs, was called for screening. Magnetic resonance angiogram showed an aneurysm at right ophthalmic internal carotid artery (ICA), and subsequent DSA showed a small wide-neck right ophthalmic ICA aneurysm (Fig b). She was treated conservatively and is currently being followed up in our clinic.

Discussion

A literature search of the Medline database was conducted using the key words "twins" and "intracranial aneurysm" or combination of these. The reference lists of the identified reports were also reviewed to search for other relevant articles. In all, 15 instances of twins with IAs were identified.[3-17] In 1942, O'Brien[18] described an instance with a probable diagnosis of aneurysmal SAH in twins from the history, which was never proven since postmortem was not permitted. This case was excluded from our series. Relevant details about the 16 pairs (including our patients) are listed in the Table. Age of presentation with SAH or being screened positive for IA ranged from 30 to 57 years, with a mean of 43 years. The female-to-male ratio was approximately 4:1. The average interval between presentation of both twins in a pair was 2.4 years. The majority (12/16) had aneurysms of the ICA or middle cerebral artery (MCA) or its branches, and in three pairs they were

Key words
Diseases in twins; Intracranial aneurysm; Subarachnoid hemorrhage; Twins, monozygotic

Hong Kong Med J 2011;17:151-4

Queen Elizabeth Hospital, Kowloon, Hong Kong:
Department of Neurosurgery
HK Leung, MB, BS
Y Lam, MB, BS
KM Cheng, FRCS (Edin), FHKAM (Surgery)
Department of Radiology
CM Chan, FRCR, FHKAM (Radiology)
YL Cheung, FRCR, FHKAM (Radiology)

Correspondence to: Dr HK Leung
Email: leungphk@gmail.com

雙胞胎的顱內動脈瘤：病例及文獻回顧

雙胞胎同時患有顱內動脈瘤的病例很罕見，文獻中只記載15宗病例。本文報告一宗雙胞胎顱內動脈瘤的病例，其中一名病發時出現蛛網膜下腔出血，其數碼血管造影顯示左後交通動脈瘤，遂接受彈簧圈治療。病人的孿生姐妹其後接受篩查，數碼血管造影也顯示其右頸內眼動脈段出現動脈瘤，最後採用保守療法治理。

located at the anterior communicating artery and basilar artery (fusion arteries); one pair had multiple aneurysms involving both sites. Seven twin pairs had IAs at the same site, while in three they were at the 'mirrored' position. The remaining six pairs had aneurysms at sites that were not related. In nine pairs, both twins presented with SAH, while in seven pairs one of the twins was screened positive after the other had presented with SAH. On comparing the symptomatic second twins (with SAHs) and the asymptomatic second twins (screened positive), the latter had better outcomes; 67% (6/9) of the former group died during their admission compared with only 14% (1/7) of the latter group did.

Twin IAs exhibit unique characteristics compared to sporadic IAs. Patients are younger at presentation, the proportion of aneurysms in the anterior communicating artery is lower, and many are located at identical or mirror sites (as discussed in the literature).[9,10,12-14] In a recent local study reviewing 266 patients with sporadic IAs, the mean age at presentation was 59 years and nearly half of the aneurysms were located on the anterior or posterior communicating arteries.[1] According to the literature, twin IA patients presented at a mean age of 43 years, with majority of aneurysms located on the ICA and MCA.

The pair of twins in our report illustrated typical characteristics of twin IAs, being located in the ICA and its branching arteries. Female predominance was demonstrated. The age at presentation was 57 years, which was younger than the mean age of onset in patients with sporadic IAs. Time interval between presentation of both twins was significantly shorter than previously reported, which reflects advances in screening test and increased awareness of twin IA, as screening was conducted promptly after diagnosis of the symptomatic twin. Intracranial aneurysm was shown on DSA of the asymptomatic twin and conservative management was adopted.

Twin IAs have played an important role in the understanding of how aneurysms develop. Sharma and Brown[7] reported a pair of twins in 2001. The one with better hypertension control had fewer aneurysms than the poorly controlled twin. As they were likely to share same cerebral vascular architecture, the blood pressure was presumed to have a role in the development of aneurysms. Twin aneurysms, being the first reported cases of familial IAs, have been used as an argument strongly supporting the role of inheritance. Vascular malformation in terms of fusion and branching error have also been discussed by Hager and Steiger,[5] with respect to twin morphogenesis. Currently available

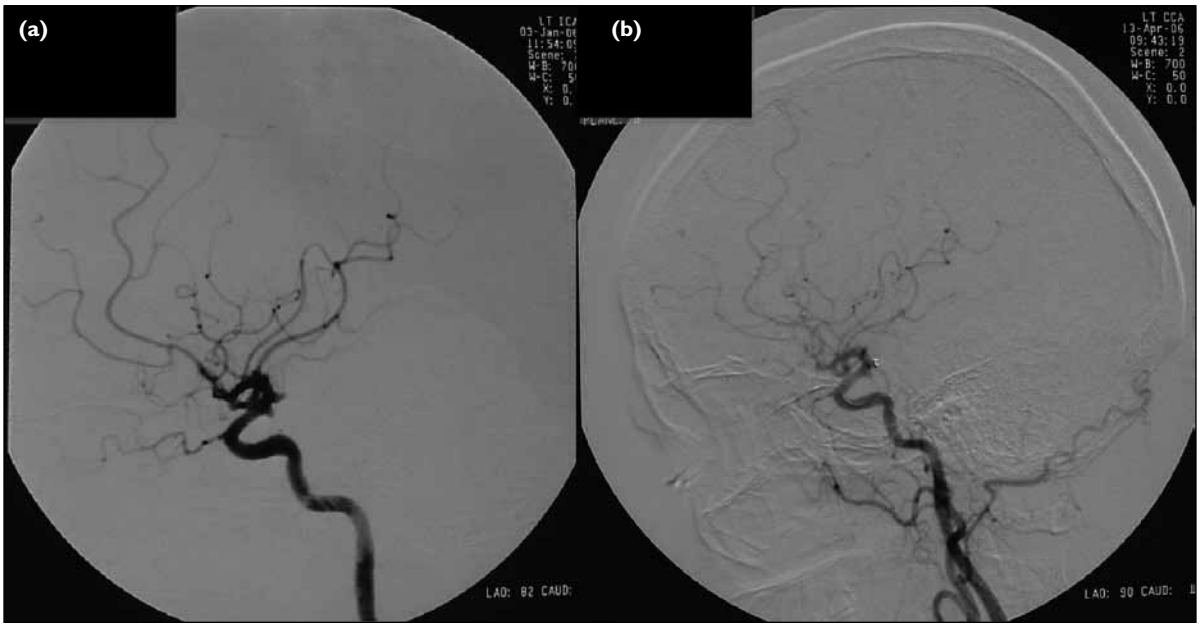

FIG. Digital subtraction angiograms

(a) Lateral view of left internal carotid artery (ICA) of elder sister showed a 2-mm posterior communicating artery aneurysm. (b) Lateral view of left ICA of younger sister showed coiling of the posterior communicating artery aneurysm

TABLE. Aneurysmal subarachnoid haemorrhages of 16 pairs of twins*

No.	Study	Age (years)/ sex	Presentation	Site	Management	Outcome
1	Brisman and Abbassioun,[17] 1971	30/F	SAH	Left and right MCA	Clipping of both aneurysms	–
		35/F	Screening	Right ICA		Death
2	Fairburn,[16] 1973	44/F	SAH	Supraclinoid part of right ICA	Ligation of right ICA	–
		46/F	SAH	Supraclinoid part of left ICA	Conservative	–
3	Wilson and Cast,[15] 1973	42/F	SAH	Left MCA, infundibulum in right PCoA	Conservative	–
		45/F	SAH	Left MCA, infundibulum in right PCoA	Conservative	Death
4	Schon and Marshall,[14] 1984	39/M	SAH	ACoA	Clipping of aneurysm	Death
		43/M	SAH	ACoA (postmortem)		Death
5	Weil et al,[13] 1988	43/F	SAH	6-mm left PCoA, 3-mm left supraclinoid ICA, 3-mm right supraclinoid ICA, fusiform enlargement of upper basilar artery	Clipping of 2 left-sided aneurysm	–
		43/F	Screening	3-mm left PCoA, 3-mm left supraclinoid ICA, 3-mm right supraclinoid ICA, 6-mm basilar artery	Clipping of basilar artery of aneurysm	–
6	Parekh et al,[12] 1992	36/F	Epilepsy	6-7 mm left MCA, left posterior inferior cerebellar artery	Clipping of both aneurysms	–
		37/F	SAH	10-mm right MCA		Death
7	Sharma and Cast,[11] 1993	42/F	SAH	Left MCA, right PCoA	Conservative	–
		45/F	SAH	Left MCA, right PCoA		Death
8	Hagen et al,[10] 1997	39/F	SAH	7-mm right supraclinoidal ICA, 5-mm and 2-mm right MCA	Clipping of all 3 aneurysm	–
		41/F	SAH	Fusiform enlargement of right infraclinoidal ICA, 5-mm right MCA, 4-mm right PCoA, 3-mm left PCoA	Clipping of 2 right-sided aneurysm	Death
9	Nakajima et al,[9] 1998	46/F	SAH	Left MCA	Clipping of aneurysm	–
		36/F	SAH	Right MCA	Clipping of aneurysm	–
10	Sakovich et al,[8] 1998	45/F	SAH	Right MCA	Clipping of aneurysm	–
		46/F	Screening	Left and right ICA	Conservative	–
11	Sharma and Brown,[7] 2001	44/F	SAH	Right ICA, right and left MCA	Clipping of right MCA	–
		44/F	Screening	Right ICA, right cavernous sinus	Conservative	–
12	Porter et al,[6] 2001	41/F	SAH	Basilar artery, superior cerebellar artery	Coiling	–
		41/F	Screening	Basilar artery, right PCoA	Clipping of both aneurysms	–
13	Hager and Steiger,[5] 2004	40/F	SAH	7-mm right MCA	Clipping of aneurysm	–
		40/F	Screening	7-mm right MCA	Clipping of aneurysm	–
14	Ohno et al,[4] 2004	39/M	SAH	Left MCA	Clipping of aneurysm	Death
		42/M	SAH	Left MCA	Clipping of aneurysm	–
15	ter Laan et al,[3] 2009	49/M	SAH	ACoA	Coiling	–
		45/M	SAH	ACoA (MRA)	Clipping of aneurysm	–
16	Present case	57/F	SAH	Left ICA	Coiling	–
		57/F	Screening	Right ICA	Conservative	–

* SAH denotes subarachnoid haemorrhage, MCA middle cerebral artery, ICA internal carotid artery, PCoA posterior communicating artery, ACoA anterior communicating artery, and MRA magnetic resonance angiogram

evidence supports the concept of multifactorial processes in the development of IAs.

Screening of relatives of persons with ruptured aneurysms has always been controversial.[10,17] Twin IA, in particular, with its few case reports, lacks evidence for comprehensive and conclusive advice. In general, authors reporting case of twin IA tend to support screening of the asymptomatic twin, in view of the high mortality associated with ruptured aneurysms.[7,9,12,13] On the other hand, both Astradsson and Astrup[19] and Puchner et al[20] reported cases with presence of IA in one identical twin but upon screening of the other, no aneurysm was found. This poses questions pertaining to unnecessary risks of angiography as well as anxiety in seemingly healthy patients. Nowadays, screening for cerebral aneurysms can be performed by magnetic resonance angiography. Furthermore, whether an identified unruptured aneurysm should be treated is debatable. To settle this issue, a large-scale controlled study which emphasises the importance of reporting of such cases appears necessary.[15] There is also debate about the natural history of aneurysms, and whether a normal angiogram precludes later development of aneurysms. Brisman and Abbassioun[17] reported a case with a negative initial screening angiogram in an asymptomatic twin despite direct injection into the artery, in which an aneurysm was demonstrated later and its progress was monitored by DSA.

In conclusion, current evidence does not provide a definite answer as to whether screening should be carried out. Cases should therefore be judged on individual basis.

References

1. Lap HP, Cheng KM, Yu SC, et al. Size, location, and multiplicity of ruptured intracranial aneurysms in the Hong Kong Chinese population with subarachnoid haemorrhage. Hong Kong Med J 2009;15:262-6.
2. Nahed BV, Bydon M, Ozturk AK, Bilguvar K, Bayrakli F, Gunel M. Genetics of intracranial aneurysms. Neurosurgery 2007;60:213-25.
3. ter Laan M, Kerstjens-Frederikse WS, Metzemaekers JD, van Dijk JM, Groen RJ. Concordant symptomatic intracranial aneurysm in a monozygotic twin: a case report and review of the literature. Twin Res Hum Genet 2009;12:295-300.
4. Ohno S, Ikeda Y, Onitsuka T, et al. Cerebral aneurysms in identical twins [in Japanese]. No Shinkei Geka 2004;32:875-9.
5. Hager P, Steiger HJ. Identical cerebral aneurysms in siblings: report of two families. J Clin Neurosci 2004;11:80-4.
6. Porter PJ, Tymianski M, Muller PJ, Terbrugge KG. What to do when the doctor sees double: identical twins with nearly identical aneurysms. Interv Neuroradiol 2001;7:153-60.
7. Sharma P, Brown MJ. Neurovascular lessons from a pair of identical twins with cerebral aneurysms. Postgrad Med J 2001;77:197-8.
8. Sakovich VP, Lebedeva ER, Nalesnik MV. Heredity in intracranial aneurysms (a study of identical twins) [in Russian]. Zh Vopr Neirokhir Im N N Burdenko 1998;(2):33-5.
9. Nakajima H, Kishi H, Yasui T, et al. Intracranial aneurysms in identical twins. Surg Neurol 1998;49:306-8.
10. Hagen T, Neidl K, Piepgras U. Multiple cerebral aneurysms in identical twins. AJNR Am J Neuroradiol 1997;18:973-6.
11. Sharma RR, Cast IP. Cerebral berry aneurysms in identical twins. Surg Neurol 1993;40:349-50.
12. Parekh HC, Gurusinghe NT, Sharma RR. Cerebral berry aneurysms in identical twins: a case report. Surg Neurol 1992;38:277-9.
13. Weil SM, Olivi A, Greiner AL, Tobler WD. Multiple intracranial aneurysms in identical twins. Acta Neurochir (Wien) 1988;95:121-5.
14. Schon F, Marshall J. Subarachnoid haemorrhage in identical twins. J Neurol Neurosurg Psychiatry 1984;47:81-3.
15. Wilson PJ, Cast IP. "Twin" intracranial aneurysms. Br Med J 1973;1:484.
16. Fairburn B. "Twin" intracranial aneurysms causing subarachnoid haemorrhage in identical twins. Br Med J 1973;1:210-1.
17. Brisman R, Abbassioun K. Familial intracranial aneurysms. J Neurosurg 1971;34:678-82.
18. O'Brien JG. Subarachnoid haemorrhage in identical twins. Br Med J 1942;1:607-9.
19. Astradsson A, Astrup J. An intracranial aneurysm in one identical twin, but no aneurysm in the other. Br J Neurosurg 2001;15:168-71.
20. Puchner MJ, Lohmann F, Valdueza JM, Siepmann G, Freckmann N. Monozygotic twins not identical with respect to the existence of intracranial aneurysms: a case report. Surg Neurol 1994;41:284-9.