Intracerebral Hemorrhage due to Unilateral Moyamoya in a Young Woman during Puerperium

by

ISSN 0975-2366
DOI: https://doi.org/10.31838/ijpr/2020.12.03.311

Research Article

Intracerebral Hemorrhage due to Unilateral Moyamoya in a Young Woman during Puerperium

POPPY KRISTINA SASMITA[1,2*], HERLINA UINARNI[1,3], ROBI IRAWAN[1], TENA DJUARTINA[1], MASHURI[4], HULDANI[5]

[1]Department of Anatomy, School of Medicine and Health Sciences, Atma Jaya Catholic University of Indonesia, Jakarta, Indonesia.
[2]Neurologist at Mitra Keluarga Kelapa Gading Hospital, Jakarta, Indonesia
[3]Radiologist at Pantai Indah Kapuk Hospital, Jakarta, Indonesia
[4]Departement of Radiology, Faculty of Medicine, Lambung Mangkurat University, Banjarmasin, Indonesia
[5]Departement of Physiology, Faculty of Medicine, Lambung Mangkurat University, Banjarmasin, Indonesia
*Corresponding Author
Email ID : poppy.kristina@atmajaya.ac.id. , herlina.uinarni@atmajaya.ac.id.
Received: 16.03.20, Revised: 26.04.20, Accepted: 02.05.20

ABSTRACT

Moyamoya disease is an uncommon cerebral vasculopathy associated with bilateral progressive steno-occlusion of the terminal internal carotid arteries with formation of collateral blood vessels. We present a case of a 26-year-old woman post-partum (1 month) with unilateral moyamoya, came to the hospital with severe headache and vomiting. Ten hours later patient looked loss of consciousness with Glasgow Coma Scale or GCS E2V2M4 and had weakness in the left limbs. Head Computed Tomography (CT) scan showed bleeding at right nucleus caudatus, right anterior crus of internal capsule with intraventricular hemorrhage (entire ventricular system) with minimal midline shift to the left. On examination Digital Subtraction Angiography (DSA) showed stenosis of the right distal internal carotid artery with "puff of smoke" appearance. Patients hospitalized for 17 days and went home with compos mentis and without weakness in the left limbs. After hospitalization, head CT Angiography was performed to evaluate the intracranial and extracranial blood vessels. Patient had cerebral bypass surgery procedure one month later. Six months after surgery, the patient was evaluated with head CT Angiography and showed normal brain parenchyma and brain perfusion. Although a rare case, intracerebral hemorrhage can caused by unilateral Moyamoya disease.

INTRODUCTION

Moyamoya disease was originally described by Suzuki and Kodoma and Suzuki and Takaku in 1957, has an unknown etiology with the characteristics of stenosis or progressive bilateral occlusion of the internal carotid artery with the formation of vascular tissue called 'moyamoya vessels' (1). This is a rare chronic condition with clinical manifestations as infarction or hemorrhage stroke, whereas it affects children and young adults with female predominance. Although this disease often occurs in Japan, several cases have been reported in several places (2,3). We present a case of a young woman having intracerebral hemorrhage due to unilateral moyamoya during puerperium. Angiographic findings, with unilateral moyamoya, are described. We followed the clinical development of patient before and after surgical procedure.

Case Report

A 26-year-old woman was presented in the emergency department with the chief complaints of severe headache, nausea and vomiting 3 hours before admission to the hospital. Patient still awake with blood pressure was 130/90 mmHg, pulse rate was 84x/minute and body temperature was 37°C. There was no neurological deficit on neurology examination.

Electrocardiogram (ECG) and chest X-ray examination were normal. Her laboratory tests showed that haemoglobin 11.5 g/dL, leukocytes 12,000 /mm3, hematocrit 35% and blood sugar 130 mg/dL. She had just given birth with caesarean section 1 month before admission with a history of hypertension in the third trimester of pregnancy and placenta previa. She underwent a cesarean section under general anesthesia at 37 weeks of gestation due to bleeding caused by placenta previa. She did not have any history of connective tissue disorders and risk factors for cerebrovascular disease.

About 10 hours in treatment, the patient showed decreased of consciousness (GCS E2V2M4) with blood pressure 130/80 mmHg, pulse rate 80x /minute and body temperature 37.1ºC. On

neurology examination was obtained pupils were Ø 2 mm with right medial rectus muscle palsy and weakness of the left limb. A head CT scan showed bleeding at right nucleus caudatus, right anterior crus of internal capsule with intraventricular hemorrhage (entire ventricular system) with minimal midline shift to the left (Fig.1). The patient was transferred to the ICU and given 0.5 g/kg BB Manitol IV and Nimodipine 2.5 cc per hour. Three hours later the patient appeared to open eyes spontaneously but fell asleep again with complaint of headache. Patient was given additional analgesic IV therapy and was consulted to the neurosurgery department. Neurosurgeon recommended for Subtraction Digital Angiography (DSA) examination to find abnormal cerebrovascular. The diagnosis of Moyamoya was known after we did DSA that showed stenosis of the right distal internal carotid artery (ICA)with "puff of smoke" appearance (Fig.2).

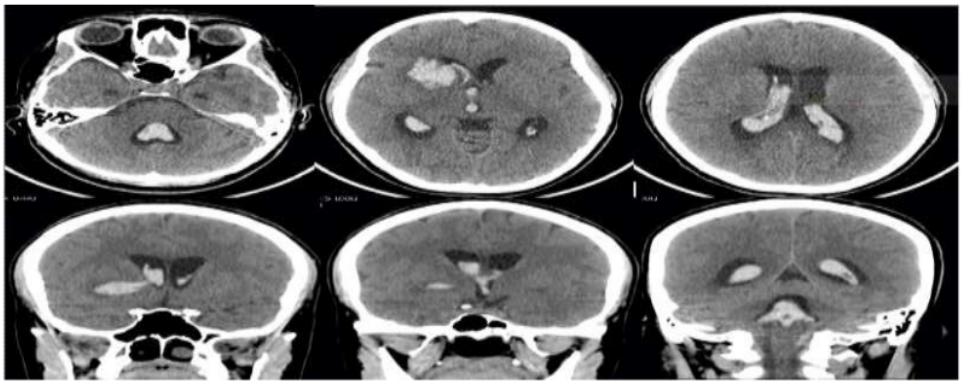

Fig.1: Non-contrast brain CT Scan (upper raw axial wiew and below raw coronal view) demonstates acute hemorrhage as hyperdens signal intensity at right nucleus caudatus, right anterior crus of internal capsule with intraventricular hemorrhage (entire ventricular system) with minimal midline shift to the left.

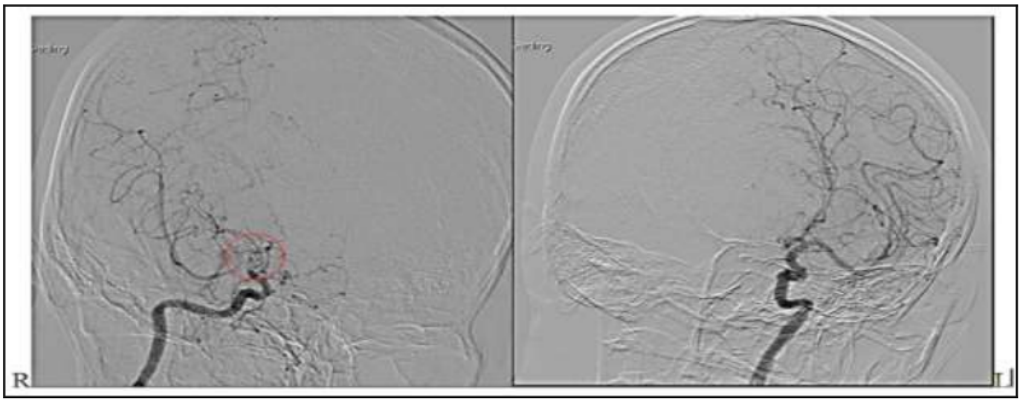

Fig.2: Digital Subtraction Angiography (DSA) bone subtraction, anteroposterior projection, demonstrates stenosis of the right distal internal carotid arterywith "puff of smoke" appearance (red circle).

Patient was hospitalized for 17 days, went home with compos mentis without weakness in the left limbs. After hospitalization, head Computed Tomography Angiography (CT Angiography or CTA) was performed to evaluate the intracranial and extracranial blood vessels (Fig.3).

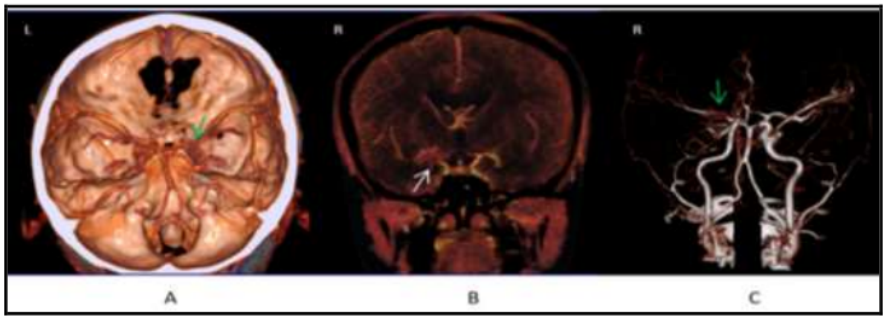

Fig.3 Head CT Angiography. Volume-rendered MDCTA image (A axial view and C coronal vier), B. Multiplanar reformation or reconstruction (MPR), clearly demonstrates stenosis in the right distal internal carotid artery(green arrow) with collateral lenticulostriate artery (white arrow), and right middle cerebral artery got supply from the posterior cerebral artery branches. Extracranial blood vessels are normal.

Patient had cerebral bypass surgery procedure one month later. Six months after surgery, the patient was evaluated with head CTA and showed normal brain parenchyma and brain perfusion (Fig.4 and Fig.5).

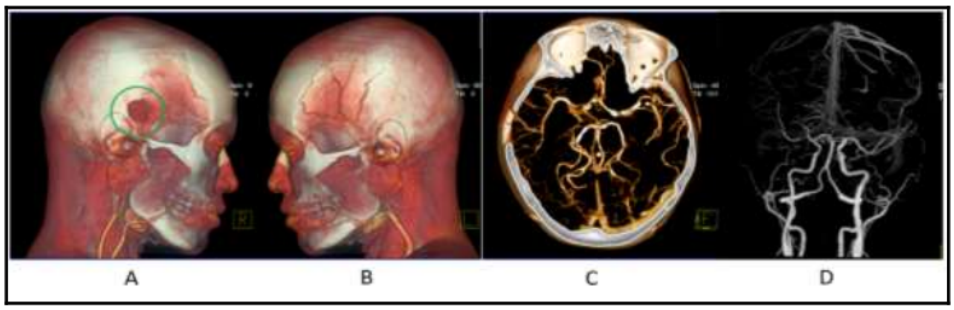

Fig.4: Head CT Angiography after cerebral bypass surgery procedure. Volume-rendered image right lateral view (A) with burr hole after surgery (green circle), left lateral view (B), Axial view (C) dan bone subtraction(D). Intracranial blood vessels are normal.

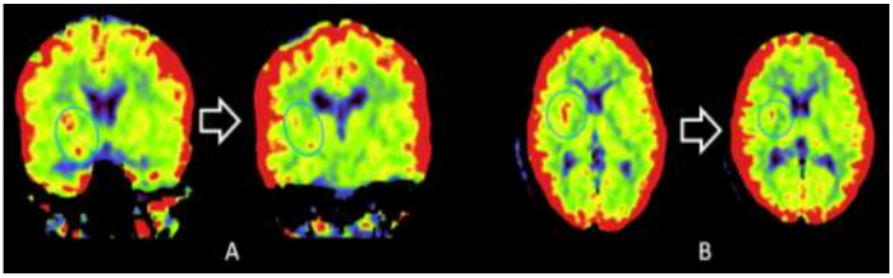

Fig.5: Head CT Angiography before and after cerebral bypass surgery procedure. Image recontruction using application cerebral blood volume, coronal view (A) and axial view (B), showed prominent blood volume in the right basal ganglia before surgery and that prominent blood volume is reduced after surgery.

DISCUSSION

Moyamoya disease is defined as an abnormality of cerebral blood vessels in the presence of chronic progressive stenosis or occlusion in terminal internal carotid artery (ICA) and proximal anterior and middle cerebral arteries (MCA) with development of collateral circulation. If these findings are bilateral and concomitant diseases such as atherosclerosis, trauma, irradiation, etc are refer to as "definite"

moyamoya. According to "The Research Committee on Spontaneous Occlusion of the Circle of Willis of the Ministry of Health and Welfare, Japan", classification of patients with unilateral occlusion or stenosis is included in probable' moyamoya disease(4,5). The occurrence of unilateral moyamoya remains unclear. The incidence rate of moyamoya disease in Japan is 0.35 per 100,000 population with a prevalence of 3.16 per 100,000 population. The exact etiology of moyamoya disease is unknown, some theories said that may be are related to the environment, genetic and viral infections. Janda et al. said that the increased incidence of moyamoya disease in Japan and populations in Asia suggested have relation with genetic predisposition(3). In children, the most clinical presentation is cerebral ischemia, approximately 40% of children under 10 years have clinical symptoms of transient ischemic attack (TIA), while nearly 30% will have symptoms of cerebral infarction. Other symptoms in children are headache and seizures, whereas intracerebral hemorrhage is uncommon. In adults, most of the symptoms are intracerebral hemorrhage reaching approximately 66%, the majority of which are intraventricular and periventricular in location. Only 45% of patients have good neurological status after exposure to intracerebral hemorrhage, and about 7% die at the onset of symptoms (6,7,8,9)

Some imaging modalities has been used for diagnosis moyamoya disease, while DSA is currently considered to be the criterion standard for diagnosis and assessment of moyamoya disease although it cannot really reflect the hemodynamic status of moyamoya disease (10). From angiographic finding, Suzuki and Takaku divided moyamoya disease into 6 stages (Table 1) (6,10). According to that stages, our patient was diagnosed with DSA as moyamoya disease probable stage III.

Table 1. Suzuki Stages of Moyamoya disease (6,10).

Suzuki Stage	Angiographic finding
I	Narrowing of carotid arteries
II	Initial appearance of moyamoya vessels : continued narrowing of the ICA, dilatation of the ACA and MCA
III	Intensification of moyamoya vessels: loss of proximal ACA and MCA, leptomeningeal collateralization from PCA, increase in moyamoya blush
IV	Minimization of moyamoya vessels
V	Reduction of moyamoya vessels
VI	Disappearance of moyamoya vessels

ICA : Internal carotid artery; ACA : anterior cerebral artery; MCA : middle cerebral artery, PCA : posterior cerebral artery

In the literature review by Kusaka et al, involving 173 patients withunilateral Moyamoya disease, identified that unilateral moyamoya disease is an uncommon condition and occurrences in adults are also rare (4). Intracebral hemorrhage in unilateral moyamoya has been reported by Cultrera et al occur in women aged 29 years (10,11).The reason why the present case developed moyamoya disease in puerperium is unclear. Several factors are known to be associated with stroke during pregnancy such as hypercoagulopathy associated with physiological changes and eclampsia, cerebral venous thrombosis, HELPP (High Blood Pressure, elevated liver enzymes, low platelet count) syndrome and post-partum cerebral angiopathy (12,13,14).The pathophysiology of intracerebral hemorrhage in moyamoya disease is not yet fully known, but microaneurysms are formed in dilated arteries, especially those in blood vessels located in the periventricular region and fibrinoid necrosis of arterial walls in the basal ganglia. Pathological studies on collateral blood vessels showed there are signs of stress such as increased blood flow, damage in the internal elastic lamina and thinning of the tunica media which predispose to the formation of microaneurysms. Long-term ischemia plays an important role in the formation of arterial anastomosis and the occurrence of dilatation associated with rupture of blood vessels in the event of an increase in blood pressure (1,8,15). Kakogawa et al reported a combination of increased blood pressure, preeclampsia and the presence of underlying moyamoya disease caused intracerebral hemorrhage stroke (16). Our patient had hypertension history in the third trimester of pregnancy. In our patient, the cause of stenosis of the internal carotid artery underlying moyamoya disease is unknown, there may be aprocess of endothelial and smooth muscle proliferation occurs during gestational hypertension.

Management of moyamoya disease is by cerebral bypass surgery aimed at revascularization, which is differentiate into direct, indirect and

combination techniques. According to Starke et al, the procedure that is often performed in adult patients with moyamoya disease is a combination of indirect and direct techniques because it is more effective and reduces the occurrence of postoperative ischemia (17,18). Our patient had a hybrid (mixed) indirect surgery with excellent results. At this time our patient is doing normal activities.

CONCLUSION

Intracerebral hemorrhage can be caused by unilateral moyamoya disease, although it is rare and as a clinician can think of it as an underlying disease of hemorrhagic stroke during pregnancy or puerperium. Imaging technique such as DSA and CT Angiography required to make diagnosis moyamoya disease and to assess the anatomical and morphological features of the vascular network.

Conflicts of Interest: All of the authors declare that there is no conflict of interest in this article writing.

ACKNOWLEDGMENTS

Thank you to Interventional Radiologists (Dr. Soewandi and Dr. Samuel Tandionugroho) who helped us establish the diagnosis of moyamoya through a good picture of DSA.

REFERENCES

1. Suzuki J, Kodama N. Moyamoya disease-A review. Stroke. 1983; 14:104-109.
2. Shoukat S, Itrat A, Taqui AM, Zaidi M, Kamal AK. Moyamoya disease: A clinical spectrum, literature review and case series from a tertiary care hospital in Pakistan. BMC Neurol. 2009; 9:15.
3. Janda PH, Bellew JG, Veerappan V. Moyamoya Disease: Case Report and Literature Review. Journal of the American Osteopathic Association. 2009;109:545-551.
4. Kusaka N, Tamiya T, Adachi Y, Katayama S, Namba S, Tokunaga K, et al. Adult unilateral moyamoya disease with familial occurrence in two definite cases: A case report and review of the literature. Neurosurgical Review. 2006; 29:82-87.
5. Fukui M. Guidelines for the diagnosis and treatment of spontaneous occlusion of the circle of Willis ("Moyamoya" disease). In: Clinical Neurology and Neurosurgery. 1997;99(Suppl 2):S238-S240.
6. Zipfel GJ, Fox DJ, Rivet DJ. Moyamoya disease in adults: The role of cerebral revascularization. Skull Base. 2005;15(1):27-41.
7. Gosalakkal JA. Moyamoya disease: A review. Neurology India. 2002;50:6-10.
8. Huldani, Mohammad Rudiansyah, Fauzie Rahman, Adelgrit Trisia, Sri Ramadhany, Siti Kaidah, Harun Achmad, Bayu Indra Sukmana, Dhea M Swengly, Shelsy Marippi, Wafa Ahdiya, Muhammad Hasan Ridhoni, Amalia Rahman, Zhasifa Khoirunnisa Suwanto, Gandhi Mahesa Priambodo, Muhammad Rafagih, Abdullah Zuhair. The Influence of Uric Acid Levels on Blood Pressure and Chronic Hypertension towards Hypertension Patient Proteinuria Levels (Overview of the Banjar Ethnic at the Cempaka Banjarmasin Health Center). SRP. 2020; 11(5): 52-56. doi:10.31838/srp.2020.5.09
9. Huldani, Siti Kaidah, Dwi Laksono Adiputro, Harun Achmad, Bayu Indra Sukmana, Deby Kania Tri Putri, Yusrinie Wasiaturrahmah, Renie Kumala Dewi, Didit Aspriyanto, Isnur Hatta, Saka Winias, Ariyati Retno Pratiwi, Ernita sari, Aminuddin Prahatama Putra, Anastasya Deborah M. C Manik, Ketrin Zailin, Ika Kusuma Wardani. Effect of Total Cholesterol Levels and Triglycerides on Blood Pressure Hypertension Patients Overview against Puskesmas Banjar Ethnic Group in Cempaka Banjarmasin. SRP. 2020; 11(4): 384-389. doi:10.31838/srp.2020.4.57
10. Li J, Jin M, Sun X, Li J, Liu Y, Xi Y, et al. Imaging of Moyamoya Disease and Moyamoya Syndrome: Current Status. Journal of Computer Assisted Tomography. 2019;43:257-263.
11. Cultrera F, Giurfrida M, Alberio N, Chiaramonte I. Hemorrhagic unilateral moyamoya: Report of one case. Neurologia. 2004;19(5):277-279.
12. Akamatsu Y, Fujimura M, Uenohara H, Shimizu H, Tominaga T. Development of moyamoya disease in pregnancy and puerperium: Case report. Neurol Med Chir (Tokyo). 2014;54:824-826.
13. Huldani, Harun Achmad, Aryadi Arsyad, Aminuddin Prahatama Putra, Bayu Indra Sukmana, Dwi Laksono Adiputro, Julia Kasab. Differences in VO2 Max Based on Age, Gender, Hemoglobin Levels, and Leukocyte Counts in Hajj Prospective Pilgrims in Hulu Sungai Tengah Regency, South Kalimantan. SRP. 2020; 11(4): 09-14. doi:10.31838/srp.2020.4.03
14. Takagi Y, Kikuta KI, Nozaki K, Hashimoto N. Histological features of middle cerebral arteries from patients treated for moyamoya disease. Neurol Med Chir (Tokyo). 2007;47:1-4.
15. Herlina Uinarni, Conny Tanjung, Huldani, Aminuddin Prahatama Putra, Mashuri, Bayu Indra Sukmana, Heru Wahyudi, Abdullah Zuhair, Aderiel Gabrian Tarius, Wisnu Wiryawan, Renie Kumala Dewi, Yunita Feby Ramadhany, Harun Achmad. The Importance of Ultrasound Findings in Children with Acute Abdominal Pain to Prevent Unnecessary Surgery. SRP. 2020; 11(4): 377-383. doi:10.31838/srp.2020.4.56
16. Kakogawa J, Sadatsuki M, Masuya N, Gomibuchi H, Ohno H, Hara T, et al. Antepartum

intracranial hemorrhage due to unrecognized unilateral moyamoya disease: A case report. Arch Gynecol Obstet. 2011;283(Suppl 1):S19-S22.
17. Starke RM, Komotar RJ, Connolly ES. Optimal surgical treatment for moyamoya disease in adults: direct versus indirect bypass. Neurosurg Focus. 2009;26(4):E8.
18. Herlina Uinarni, Bachtiar Murtala, Muhammad Nasrum Massi, Andi Dwi Bahagia Febriani, Ristaniah Rose Effendy, Mochammad Hatta, St. Maisuri T Challid, Mirna Muis, Ema Alasiry, Burhanuddin Bahar, Huldani, Harun Achmad. Aquaporin-5, Subunit Beta in the Sodium Channel Epithelium, Lung Ultrasonography Examination in Transient Tachypnea of the Newborn. SRP. 2020; 11(6): 466-473. doi:10.31838/srp.2020.6.73
19. Svetlana Dmitrievna Danshina,
20. Alexander Markov, Huldani, Harun Achmad, Causes, symptoms, diagnosis and treatment of melanoma. International Journal of Pharmaceutical Research ISSN: 0975-2366 , 1903-1909, Jul - Sep 2020 | Vol 12 | Issue 3
Herlina Uinarni, Maria Angelina Yashinta G, Liliana Sugiharto, Huldani
,Harun Achmad, Alexander Markov, Dmitry O. Bokov, Left Pulmonary Aplasia: A Case Report, Journal of Pharmaceutical Research ISSN: 0975-2366, 2144-2148, Jul - Sep 2020 | Vol 12 | Issue 3
21. Markov A.A. Problems of the surgical treatment of patients with fractures of theproximal femur on the basis of osteoporosis. Sys. Rev. Pharm. 2019; 10(1): 143-145
22. Markov A.A. The Development of Dental Implant with the Bioactive Covering onthe Basis of Synthetic Complex with Biogenic Elements. Sys. Rev. Pharm. 2020;11(2): 278-283.
23. Markov Alexander. Results of the Experimental Research and Clinical Applicationof Cannulated Screw with Bioactive Coverage on the Basis of NaturalHydroxiapatite during Osteosynthesis of Medial Fractures of Femur's Neck.
24. Journal of Biomimetics, Biomaterials and Biomedical Engineering ISSN: 2296-9845, Vol. 46, pp 52-59© 2020 Trans Tech Publications Ltd, Switzerland

Intracerebral Hemorrhage due to Unilateral Moyamoya in a Young Woman during Puerperium

ORIGINALITY REPORT

12%	13%	2%	4%
SIMILARITY INDEX	INTERNET SOURCES	PUBLICATIONS	STUDENT PAPERS

PRIMARY SOURCES

1	ijpronline.com Internet Source	3%
2	www.ncbi.nlm.nih.gov Internet Source	3%
3	docksci.com Internet Source	2%
4	Submitted to Universitas Hasanuddin Student Paper	2%
5	cyberleninka.org Internet Source	2%

Exclude quotes	On	Exclude matches	< 2%
Exclude bibliography	On		