SUCCESSFUL TREATMENT OF PONTINE CAVERNOUS ANGIOMA IN INDONESIA: CASE REPORT

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ABSTRAK

Penulis melaporkan kasus dari seorang wanita berumur 50 tahun dengan pontin angioma kavernosus yang diterapi di Indonesia dengan mempergunakan fasilitas kesehatan terbatas. Pasien masuk rumah sakit dengan hemiparesis sisi kiri dan paresis saraf kranial V dan VII sisi kanan. Gambaran neuroimajing menampakkan adanya perdarahan batang otak yang disebabkan oleh angioma kavernosus di ventro lateral pons sisi kanan. Angioma kavernosus termasuk perdarahannya berhasil dikeluarkan melalui operasi dengan pendekatan retrosigmoid. Tidak ditemukan adanya defisit neurologi tambahan pasca operasi. Laporan ini mengetengahkan kemungkinan penanganan kasus-kasus dengan mempergunakan peralatan dan tekhnis dasar untuk operasi batang otak di negara-negara berkembang.

Kata kunci: Keberhasilan terapi, operasi batang otak, pontin angioma kavernosus, negara berkembang

ABSTRACT

The authors describe a case of 50 year-old woman with pontine cavernous angioma who was treated in such limited facilities in Indonesia. She was admitted with left sided hemiparesis and right cranial nerves V and VI palsies. Neuroimaging revealed a brain stem hemorrhage due to cavernous angioma at the right ventrolateral pons. The cavernous angioma including hematoma was successfully removed through retrosigmoid approach. There was no additional neurological deficit postoperatively. This report describes the possibility of treating such case with the most basic technology required for brain stem surgery in developing countries.

Keywords: *Succesful treatment, brain stem surgery, pontine cavernous angioma, developing countries*

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INTRODUCTION

Cavernous angioma (cavernoma) is a benign vascular hamartoma, among intracranial vascular malformations, it is one of the rarest. Its necropsy incidence is between 0,39% and 0,53%.[12] Between 10%-23% are located in the posterior fossa with most in the pons.[5,11] Cavernomas in this region have a higher incidence of bleeding and rebleeding than in other locations. Neurological deficits are likely to worsen due to repeated hemorrhages, therefore the current trend is an aggressive management of the lesions reaching the pial or ependymal surface of the brain stem.[7]

In developing countries, cases like brainstem cavernomas are rarely well managed. Lack of diagnosing knowledge and surgical equipments remain to be major problems. In this study we describe a patient treated successfully in dr. Kariadi Hospital, Indonesia for ventrolaterally located brainstem cavernomas.

CASE REPORT

A 50-year-old woman was admitted to a local hospital because of left sided weakness in December 2006, was treated conservatively and returned home because her condition improved. In January 2007 she experienced the same symptoms again, and was referred to our hospital for further treatment.

On admission, she complained of having headache, vomiting and double vision. Neurological examination revealed right cranial nerves V and VI palsies, and left hemiparesis with upper limb strength smaller than lower limb. She had no history of hypertension and no

familial history of brain hemorrhages. Computed Tomography (CT) scan demonstrated a single hyperdense lesion in the pons with enhancement (Fig. 1).

Magnetic Resonance Imaging (MRI) study (0.5 T) revealed a nodular and heterogenous hemorrhage approximately 4x4x4 cm in size with low signal ring in the outer layer of the mass, suggesting cavernoma (Fig.2). The mass occupied primarily the right paramedian area of the pons with bulging of the pial surface of the ventrolateral part. Surgical removal of the lesion was done to prevent further neurological deficits due to repeated hemorrhages.

With the patient in supine position, head was turned to opposite direction and was fixed with tape to the bed. The body was rotated to the left and was given a body pillow so that the patient was comfortable in a hugging-like position. Retrosigmoid approach craniotomy was performed. Skin was incised S shape vertically and bone was drilled just posterior to imaginary line of sigmoid sinus. In order to obtain maximum access to pons and its surrounding area, dura mater was dissected closed to sigmoid sinus, approximately 3 cm in length. Lateral aspect of pons was exposed with minimum cerebellar retraction. Cranial nerves V, VI and VIII were visualized, with cranial nerve V was thinned and appeared yellowish in color. A part of pons was dicolorized, and discolored site was incised. After incision, dark brown hematoma was found. After suctioning the hematoma, angioma was exposed. It was yellowish in color. Angioma was separated from surrounding pons tissue, and was removed completely.

After removal of the angioma, The wound was closed after dural plasty with fascia. Early post operative CT showed residual blood from the surgery (Fig.3). No additional neurological deficits were observed following the surgery. Histopathological study confirmed the diagnosis of cavernoma (Fig.4). It demonstrated widening of blood vessels, which contained red blood cells, thinned endothelial layer and pointed out the presence of hemosiderin staining.

DISCUSSION

Surgery of brain stem cavernomas has two main goals: to achieve complete resection of the lesion and to avoid additional neurological damage to the patient,[1] because remnants of cavernomas may lead to further bleedings. A careful preoperative study is essential step in determining which lesion can be excised safely. Considering several limitations we encounter in Indonesia, in order to treat this lesion, we carefully determine eligibility of surgery and examination to perform based on our tools` availability and evidence based medical sources.

Symptomatic and surgically accessible lesions of cavernoma are recommended for surgery.[15] Our patient presented with left hemiparesis and cranial nerves deficits. She already experienced it twice. In our point of view, she was in a great risk for another rebleeding episode. In the literature, it was said that annual bleeding rate with prior bleeding was 5% and without prior bleeding was 0,6%.[6] In a retrospective analysis of brain stem cavernous malformations, 30 patients were treated conservatively, with a mortality rate of 20%.[4] There is always a surgical risk for the patient, but the risk of bleeding and rebleeding is greater, therefore direct surgery should be one of the options to eliminate the serious complications and symptoms due to rebleeding.

Preoperatively, CT demonstrated atypical hemorrhage in the pons. MRI study was performed, and it accurately revealed the location, the foci of hemorrhage as well as the age of previous hemorrhage.[15] MRI demonstrated a characteristic of cavernous angioma with nodular and heterogenous signal covered with a low signal of hemosiderin ring. Multiple bleedings varying through a patient's life can be differentiated by the appearance of hemogloblin and breakdown products on MRI.[15] Even though we have only 0,5 Tesla MRI machine, as it has been shown in Figure 2, it could clearly demonstrate the characteristic of cavernoma. Therefore diagnosis could be established.

Approach of operation was determined based on MRI appearance. The lesion was located in the ventrolateral part of the pons.. Retrosigmoid approach was chosen because the lesion was extended to the brain stem surface. The lateral side of pons can be easily visualized and the risk of causing hearing loss is very small through this approach. We often use this approach to operate on Cerebello Pontine Angle tumors, it was hoped that familiarity could ease the removal of the angioma. Patient was placed in lateral position, and since there is no head fixation in our hospital, head of patient was turned to the left and was taped to the bed to gain good fixation. Point of entry was determined by accurately observing discolorized pons area caused by hemosiderin staining from the hemorrhage. The darkest area pointed out that the lesion was at its closest point to the surface and was chosen to be the entry point.[2,8,10,13]

Other possible approaches for anterior pontine cavernomas include translabyrinthine, antero-lateral approach (pterional, subtemporal, lateral extreme), and anterior approach (transoral).[1,3,8,9,10,14] Translabyrinthine was not chosen because of its high risk in causing hearing loss to patient postoperatively, whereas subtemporal approach was not chosen because it involves too much retraction of the temporal lobe in order to obtain a wide view of the pons. Last possible approach, transoral, brought great risk of CSF leakage and infection, in addition it only gives narrow view of the pons. Therefore it was not effective for patient's treatment. Ferroli et al reported 51 brain stem cavernomas, 20 of which were in the pons. Retrosigmoid approach was applied to 13 cases with good outcome.[3] Sola et al had experience in treating 17 brainstem cavernoma, 13 of them were in the pons and 9 of which were successfully treated with retrosigmoid approach.[14]

Postoperatively, our patient's condition was generally good. She was alert. 6 months post operation she still experienced facial numbness but no additional neurological deficit was found. She did not experience any rebleeding episode. Early postoperative CT demonstrated complete removal of the lesion (Fig.3). The published outcome of surgically treated patients with brain stem cavernous malformation is good. In larger series, surgical results were unchanged or improved in 69% to 91%.[12,13] Whereas most authors had no surgical death, Porter et al reported a surgical mortality of 3,5% caused by cardiopulmonary arrest and hemorrhagic venous cerebellar infarction.[8]

As in other developing countries, problems like lack of technological requirements, health insurance and access to health services are still encountered in neurosurgical field in our country. First of all, health insurance does not cover for all kind of examinations. If such examination required for establishing diagnosis is not listed then patient has to pay for it. Therefore we only perform basic examinations required to establish diagnosis. In cases like cavernomas, MRI study is a standard tool for diagnosis, and with 0.5 Tesla MRI machine we posses we believe it can provide us the necessary information about the lesion. We are also lack of support from the authorities, who think it is difficult to achieve satisfactory outcomes for patients with neurosurgical diseases. Health system is based on public health priorities, therefore neurosurgery has a very small place in it. We are in the very beginning of process of building such trust for cavernoma surgery through the success of this case.

Other problems we are facing are lack of knowledge and skilled neurosurgeons who are willing to operate on such cases. Since access to health services in our country is divided into 4 levels (primary, secondary, tertiary, quartenary), and that most of neurosurgical facilities are in tertiary or quartenary level, cases like brainstem cavernomas can be easily misdiagnosed by doctors in primary or secondary level due to lack of knowledge. Even amongst neurosurgeons there are still some thoughts that such case is an inoperable one. Perhaps cases of brain stem

cavernomas are actually more than we have encountered and was misdiagnosed as usual brainstem hemorrhages. It was considered as terminal case.

It becomes our responsibility to improve neurosurgical skill ability for brainstem cavernomas in Indonesia and to increase diagnosing capability for doctors in primary and secondary levels, therefore better case findings can be achieved. Another thing we try to develop is intraoperative monitoring. At the moment, we operate brainstem covernoma blindly only by understanding its anatomy-topographic location of surrounding vital structures. Its location in the brainstem area needs careful intraoperative monitoring such as brainstem auditory, motor and sensory evoked potential in order to obtain optimal postoperative results. In the future hopefully such monitoring tools will be available for us to use.

In conclusion, patients that present with hemorrhage in the brainstem or progressive neurological deficits due to growing lesion, should undergo a surgical resection of the lesion, and MR imaging is necessary for diagnostic tool in identifying exact location of cavernoma. Even in developing countries with limited facilities, brain stem cavernoma is no longer a terminal case, and is treatable.

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Figure Legends

Fig.1. Preoperative CT showing hyperdense lesion in the pons, a) pre contrast injection b) post contrast injection

Fig. 2. MRI study showing a characteristic of cavernomas in the right ventrolateral part of the pons, with heterogenous signal intensity and low signal of hemosiderin ring. a) axial view of T1 weighted image, b) axial view of T1 weighted image with contrast agent, c) axial view of T2 weighted image, d) sagittal view

Fig.3. Early post operative CT showing resiual blood from the surgery

Fig.4. Histopathological study with H&E stain. a) widening of blood vessels as compared to surrounding vessels, b) thinned endothelial layer and the presence of hemosiderin staining