Multiple External Carotid Artery Aneurysms
with Neurofibromatosis

- Case Report -

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= Abstract =

신경섬유종을 동반한 다발성 외경동맥 동맥류
- 증례보고-

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Multiple external carotid artery aneurysms with neurofibromatosis are rare. Trauma is the primary cause in the
development of aneurysms of the external carotid artery.

A 39-year-old female patient was referred to the emergency room because of a headache and a huge
lump over the left tempo-parieto-occipital region. The physical examination revealed a huge round mass, 5 ×
15 × 18 cm, in the left tempo-parieto-occipital region and low set left ear and multiple café au lait spots in trunk
and extremities.

The external carotid artery angiography demonstrated multiple aneurysms arising from the superficial temporal
artery and occipital artery. A MRI showed a huge hematoma on tempo-parieto-occipital region and postauricular
mass suggested of subcutaneous neurofibroma. Embolization followed by surgical resections of those aneurysms
and neighboring mass were performed and good result was obtained.

We report our case with review of literature.

KEY WORDS: Multiple external carotid artery aneurysms, Neurofibromatosis, Embolization, Hematoma.

Introduction

Neurofibromatosis (NF) is a hereditary disorder of any
organ or system of the body may be primarily or second-
arily involved. But cerebral arterial involvement is unco-
mmon. Variable expression results in two major types of
neurofibromatoses, i.e., bilateral acoustic neurofibromatosis
also known as central neurofibromatosis (neurofibromatosis-2) and von Recklinghausen’s neurofibromatosis(also
known as peripheral neurofibromatosis, neurofibromatosis-
1). Neurofibromatosis-1 is characterized by hyperpigmen-
ted patches of skin (café au lait macules, axillary freckles),
multiple tumors of peripheral nerves, sphenoid wing dys-
plasia, iris Lisch nodules, etc. Bilateral acoustic neurofibromatosis (neurofibromatosis-2) is characterized by neoplasia of cells of neural crest origin.

By 1970 Schecter and Gutstein documented a total of
131 aneurysms of the superficial temporal artery. They
were 0.5-2.5% of operated cerebral aneurysms and the
etiology was mostly trauma. Traumatic aneurysms of the
superficial temporal artery were originally described after
penetrating injuries from dueling or blood letting.

Until now, a review of the literature shows that males
continue to be affected in more than 80% of the cases. The average age reported was 33 years, with almost 60% of the patients between the ages of 20 and 40 years.

The most frequently encountered penetrating injuries associated with traumatic aneurysm development include temporomandibular arthroplasty, hair transplantation, cyst removal, lacerations, gunshot wounds, and chiropractic manipulations.

External carotid artery aneurysm have been attributed to an atherosclerotic or congenital basis, except trauma. There has not been a report of a “true” aneurysm since 1955. An arterial injury may be completely severed, partially severed, or may suffer contusion with subsequent wall necrosis.

We reported on a 38-year-old neurofibromatosis female patient who had multiple external carotid artery aneurysms combined with huge hematoma in the temporo-parieto-occipital area. After embolization of external carotid artery aneurysms, removal of hematoma and aneurysmectomy were performed. The postauricular mass was partially removed.

**Case Report**

A 39-year-old woman complained of huge sized painful mass on left temporo-parieto-occipital area and posterior nuchal area which occurred after minor head trauma about 6 days before transference (Fig. 1). There was no loss of consciousness. The painful mass head enlarged since its discovery. But a thrill or bruit were absent. On admission, she showed multiple caf-au-lait spots and subcutaneous nodules on her trunk and extremities. Peripheral blood studies revealed Hb 8.0 (g/dL), Hct 24 ratio, WBC 5,100 (× 10^9/L), platelet 120,000 (× 10^9/L), bleeding time 2 (min), coagulation time 9 (min), prothrombin time 15 (sec, 84%.

![Fig. 1. A & B] Preoperative view. The huge bulging mass with multiple caf-au-lait spots is seen on the left temporo-parieto-occipital area with low set left ear. C & D] Photography taken 2 months postoperatively. About 150cc of subgaleal hematoma was evacuated and excision of aneurysm was done. The postauricular subcutaneous neurofibroma was partially removed.
1.17 INR) and activated prothrombin time 26 sec. Plain skull films showed skull bone defect on left parieto-occipital area involving foramen magnum shaped gigahraphic pattern and CT scan revealed multiseptated and multistage hematoma in left temporo-parieto-occipital area and skull bone defect in left occipital area (Fig. 2). On MR imaging, mixed high and low density lesions consisted with hematoma were observed at same site, and occipital bone dysplasia and cerebellar cortical dysplasia with secondary cystic formation were noted. Especially a postauricular subcutaneous mass was noted (Fig. 3). After 2 days later, angiography was done which showed multiple external carotid artery aneurysms arising from left superficial temporal artery and left occipital artery. Especially, extravasation of contrast medium was noted from the aneurysm of the left superficial temporal artery. These aneurysms were showed like cirrroid aneurysm. Transarterial embolization with Gelfoam took place completely (Fig. 4). After embolization, dark-brownish subgaleal hematoma 150cc was evacuated and aneurysmectomy was done. A postauricular mass was partially removed (Fig. 5).

Histological examination of the mass and excised aneurysm revealed the fine fibrillary collagenous background punctuated with Wagner-Meissner bodies, these findings defines diffuse neurofibroma, and true vascular elements or schwann cells were not found in the specimen of aneurysmectomy that these findings refered a pseudoaneurysm (Fig. 6). She was discharged 30 days later after surgery and returned to ordinary work.

**Discussion**

Neurofibromatosis is a hereditary disorder. These are two distinct disorders. Bilateral acoustic neurofibromatosis is a chromosome 22 abnormality and von Recklinghausen’s neurofibromatosis is a chromosome 17 abnormality.

Cerebral arterial involvement is an uncommon manifestation of neurofibromatosis and only 43 cases have been reported. In 1951, Bergouignan and Arne classified then into three groups: Group A-only occlusive disorder, Group B-only cerebral aneurysm, Group C-occlusive disorder and cerebral aneurysm.

The average age at onset in Group A was 14 years whereas, that in Group B was 40.2 years and that in Group C was 47.5 years. The onset is delayed when aneurysms are involved. There was a female predominance in all of the groups.

The ischemic type was predominant in the younger population whereas, the hemorrhagic type was predominant in older patients. The affected side was bilateral in 18 cases and unilateral in 19 cases.
The occlusive lesions were almost all limited to the anterior circulation, namely, the frontal half of the circle of Willis.

Reubi\(^7\) studied the vascular lesions associated with neurofibromatosis. He divided them into three categories according to vessel diameter. Small arteries with a diameter ranging from 50 to 400\(\mu\)m developed a concentric growth of the intima and thinning of the media (pure intimal form).

![Fig. 4. A & B] External carotid artery angiogram demonstrating aneurysms arising from left superficial temporal artery and left occipital artery. Extravasation of contrast medium was noted from the aneurysm of the left superficial temporal artery similar to cirsoid aneurysm (arrow). C] Postembolization external carotid arteriogram showing complete occlusion of the aneurysms.
Small arteries with diameter of 100 to 700 μm developed, in the medial or whole sections of the wall, a nodular growth of spindle or epithelioid cells. Small arteries with a diameter of 0.5 to 1mm were characterized by marked, eccentric fibrous intimal thickening and small aggregates of spindle cells in intima were accompanied by a pure intimal form.

A bone defect is often seen with neurofibromatosis, characterized by a defect of ala minor ossis sphenoidalis. The occipital bone is also known as a frequent site of bone defect. The reason for the bone defect is unknown. The defect, however, may be secondary to a mesodermal defect of the periosteum or mass compression. On the other hand, spontaneous bleeding is often associated with neurofibromatosis, with pregnancy being a strong exacerbating factor.

Frontal and preauricular branches of the superficial temporal artery were the most frequent sites of injury. Traumatic aneurysms varied in size from 5mm to 4cm. The aneurysm appeared as a single pulsatile mass in approximately 90% of the cases. Traumatic superficial temporal artery aneurysms resulting from blunt trauma have been associated with various sport injuries, including hockey, rugby, squash and baseball. Superficial temporal artery aneurysms may occur after minor injury or may be associated with a skull fracture.

The latter two mechanisms lead to the development of a false aneurysm, a traumatic aneurysm, or an arteriovenous fistula. A true aneurysm is a localized or diffuse dilation of an artery that involves intima, media, and adventitia. A false aneurysm, or pseudoaneurysm, always implies a break in the arterial wall with subsequent hematoma formation and eventual hematoma organization to become a connective tissue sac.

Atraumatic aneurysm occurs when trauma weakens the arterial wall without a break in the wall.

The development of a false aneurysm caused by trauma or atraumatic aneurysm may be impossible to determine.

The external carotid artery ascends from the level of the third or fourth cervical vertebra medial to the parotid gland. The superficial temporal artery takes its origin at the border of the parotid gland. The temporalis muscle is the only protective tissue present between superficial temporal artery and the skull.

Most patients appeared within 2 to 6 weeks of injury complaining of a single painless mass, headache, pulsations, or ear discomfort. Fewer frequent complaints have
included pain, visual disturbance, dizziness, hemorrhage, cosmetic defect, and neurologic defects attributed to some associated pathologic condition\(^6\).

Approximately 15% to 20% of the patients appeared 6 months to 3 years after the initial injury. The diagnosis is made from a thorough history, physical examination, skull X-ray, Doppler examination, B-mode ultrasound scans, CT scans, angiography, etc. If the angiography was performed within 24 hours of the injury, superficial temporal artery spasm was an early sign of impending traumatic aneurysm development\(^5\). Angiography, MRI and CT scans each had a 100% sensitivity and 100% specificity in the detection of an aneurysm of arteriovenous fistula.

The angiography is required to (1) verify the vascular anomaly\(^2\) 2) delineate the neck of the pseudoaneurysm, and 3) define the involvement of the remainder of the vertebrobasilar system, in neurofibromatosis patient combined with scalp hematoma\(^1\). Several methods of therapy have been described. Nonoperative therapy consisted of maintaining constant pressure over the aneurysm for long periods of time. Winslow et al\(^10\) performed various operations (including ligation of the common carotid artery).

Operative intervention resulted in cure rate of 87%, morbidity rate of less than 5% and no deaths. The patients reported until 1988, aneurysmal clipping and embolectomy in 74%, clipping in 14%, embolization in less than 3%\(^6\). Operative intervention is indicated for relief of symptoms, correction of the cosmetic defect, and to prevent rupture. Ligation and resection is a relatively simple procedure, which may be accomplished with local anesthesia.

Selective catheterization with embolization has been prescribed for a traumatic aneurysm of the proximal superficial temporal artery and the maxillary artery.

We have obtained good results through preoperative embolization of the multiple external carotid artery aneurysms, removal of the hematoma and the postauricular mass, and aneurysmectomy in neurofibromatosis with external carotid artery aneurysm.

**Conclusion**

Neurofibromatosis with external carotid artery aneurysm is rare. Neurofibromatosis patients with superficial temporal and occipital artery aneurysms combined with huge sized hematoma are treated with preoperative embolization, removal of hematoma, partial removal of subcutaneous neurofibroma in postauricular area and aneurysmectomy through surgical intervention. After operation, the symptom was improved and she was discharged without neurological deficit. We report the case and review the literature.

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신경섬유종을 동반한 다발성 외경동맥 동맥류

표세영·김무성·심홍보·이선일·정용태·김수천·심재홍

= 국문초록 =

신경섬유종을 동반한 다발성 외경동맥 동맥류로의 경찰 및 영양동맥의 동맥류는 39세의 남성 환자에서 확인되었다. 환자는 5×15×18cm 규모의 시절로 시절은 39세의 남성 환자에서 확인되었다. 환자는 5×15×18cm 규모의 시절로 시절은 39세의 남성 환자에서 확인되었다. 환자는 5×15×18cm 규모의 시절로 시절은 39세의 남성 환자에서 확인되었다. 환자는 5×15×18cm 규모의 시절로 시절은 39세의 남성 환자에서 확인되었다. 환자는 5×15×18cm 규모의 시절로 시절은 39세의 남성 환자에서 확인되었다. 환자는 5×15×18cm 규모의 시절로 시절은 39세의 남성 환자에서 확인되었다. 환자는 5×15×18cm 규모의 시절로 시절은 39세의 남성 환자에서 확인되었다. 환자는 5×15×18cm 규모의 시절로 시절은 39세의 남성 환자에서 확인되었다. 환자는 5×15×18cm 규모의 시절로 시절은 39세의 남성 환자에서 확인되었다. 환자는 5×15×18cm 규모의 시절로 시절은 39세의 남성 환자에서 확인되었다. 환자는 5×15×18cm 규모의 시절로 시절은 39세의 남성 환자에서 확인되었다. 환자는 5×15×18cm 규모의 시절로 시절은 39세의 남성 환자에서 확인되었다. 환자는 5×15×18cm 규모의 시절로 시절은 39세의 남성 환자에서 확인되었다. 환자는 5×15×18cm 규모의 시절로 시절은 39세의 남성 환자에서 확인되었다. 환자는 5×15×18cm 규모의 시절로 시절은 39세의 남성 환자에서 확인되었다. 환자는 5×15×18cm 규모의 시절로 시절은 39세의 남성 환자에서 확인되었다. 환자는 5×15×18cm 규모의 시절로 시절은 39세의 남성 환자에서 확인되었다. 환자는 5×15×18cm 규모의 시절로 시절은 39세의 남성 환자에서 확인되었다. 환자는 5×15×18cm 규모의 시절로 시절은 39세의 남성 환자에서 확인되었다. 환자는 5×15×18cm 규모의 시절로 시절은 39세의 남성 환자에서 확인되었다. 환자는 5×15×18cm 규모의 시절로 시절은 39세의 남성 환자에서 확인되었다. 환자는 5×15×18cm 규모의 시절로 시절은 39세의 남성 환자에서 확인되었다. 환자는 5×15×18cm 규모의 시절로 시절은 39세의 남성 환자에서 확인되었다. 환자는 5×15×18cm 규모의 시절로 시절은 39세의 남성 환자에서 확인하였다.