Acute Ischemic Stroke due to Undifferentiated Sarcoma: A Case Report and Literature Review

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Abstract:
Tumor emboli due to a sarcoma are usually confirmed by an autopsy or operative findings. A sarcoma embolus in an acute stroke patient is rare. We herein report a 37-year-old man with acute stroke caused by internal carotid artery occlusion who underwent embolectomy. A histopathological analysis of an embolus obtained with a mechanical retriever device was diagnosed as undifferentiated sarcoma. This is the first case of extracardiac sarcoma extraction via mechanical retrieval performed during intervention for acute ischemic stroke. A histopathologic evaluation with embolectomy is important for diagnosing tumor emboli.

Key words: mechanical embolectomy, tumor embolus, sarcoma

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Introduction
Stroke caused by large-vessel occlusion due to tumor emboli is uncommon. Tumor emboli are usually confirmed by an autopsy or operative findings (1). A few reported patients with stroke due to tumor emboli were diagnosed by a histological examination after mechanical endovascular embolectomy. Verifying an extracardiac tumor embolus, especially sarcoma, is very rare.

Case Report
A 37-year-old right-handed man experiencing migraine presented with sudden-onset left-sided limb weakness and was immediately admitted to our hospital. He had left hemiplegia with neglect, mild dysarthria, and a National Institute of Health Stroke Scale (NIHSS) score of 13. Magnetic resonance imaging of the brain revealed acute infarcts in the right middle cerebral artery area. Subsequent magnetic resonance angiography and carotid duplex scanning showed an occluded right internal carotid artery. Intravenous recombinant tissue plasminogen activator (0.6 mg/kg) was administered 120 minutes after symptom onset, without neurological improvement. Endovascular treatment was chosen because his symptoms suggested a clinical radiological mismatch (Figure A and B). Cerebral angiography showed right internal carotid artery proximal occlusion. Mechanical endovascular embolectomy with 3 passes of a Trevo ProVue Retriever 4×20 mm (Stryker, Kalamazoo, USA) was successful, achieving partial recanalization with thrombolysis in cerebral infarction (TICI) grade 2b (Figure C-E).

A pathological examination of the embolus revealed atypical cells with pleomorphic nuclei (Figure F) that were immunohistochemically positive only for vimentin and desmin (Figure G), suggesting undifferentiated sarcoma. Transesophageal echocardiography showed no cardiac tumor. Positron emission tomography-computed tomography revealed the uptake at the aortic root, indicating that the embolus originated from an occult aortic sarcoma (Figure H). One month after the stroke, his NIHSS score was 4, and his modified Rankin Scale score was 2. He was discharged on day 54 and later prescribed proton therapy for the sarcoma.

Discussion
To our knowledge, this is the first case of extracardiac undifferentiated sarcoma extraction via mechanical retrieval performed during acute ischemic stroke intervention. Undif-
Differentiated sarcomas are rare and are often diagnosed by an autopsy. In our case, a histopathological analysis of the embolus obtained from the mechanical retriever device was diagnostic of an undifferentiated sarcoma. Thus, mechanical embolectomy may be useful for evaluating tumor-induced acute large-vessel occlusions in order to obtain a histological diagnosis.

Several trials have demonstrated the benefit of endovascular therapy for acute stroke patients with large-vessel occlusion (2), although the embolectomy-retrieved emboli were not sufficiently investigated histopathologically. As the appearance of the embolus in our case could not be macroscopically distinguished from that of a common thrombus, it was difficult to diagnose it as a tumor embolus without a routine pathological examination. As reports of tumor emboli extraction due to mechanical retrieval in acute ischemic stroke are important, we explored recently reported stroke patients with tumor emboli treated with endovascular embolectomy (Table). Of the 14 cases, 10 (71%) were of cardiac origin, 6 of which (60%) were cardiac myxomas. Cardiac origin is among the most frequent origins of intracranial tumor emboli (3). The occlusion sites were treated with clot retrieval (n=10) and aspiration embolectomy (n=5) devices. Altogether, 12 (85%) cases achieved successful recanalization (TICI 2b-3). The extracardiac tumors included breast cancer, melanoma, and lung cancer (4-6). In these cases, the primary lesion had already been confirmed before embolectomy, whereas in our case, it was diagnosed after embolectomy.

In conclusion, a histopathological evaluation following embolectomy is important for diagnosing tumor emboli. Although embolectomy for tumor emboli appears to be effective and relatively safe, further studies are needed to verify our results.

Ethics approval was provided by the Ethics Committee of Ichinomiya Nishi Hospital, Japan.

The authors state that they have no Conflict of Interest (COI).
Table. Summary of Reported Cases with Histological Confirmation of Cardiac and Non-cardiac Tumor Causing Stroke with Endovascular Embolectomy.

| Case | Tumor types                       | Primary origin | Age/sex | Initial NIHSS | Site of occlusion | Treatment modalities                                      | Recanalization | Clinical outcome | Reference |
|------|-----------------------------------|----------------|---------|---------------|------------------|---------------------------------------------------------|----------------|-----------------|-----------|
| 1    | Papillary fibroelastoma           | Unknown        | 64/M    | 16            | Right M1         | Solitaire®                                             | TICI 3         | NIHSS 3         | [7]       |
| 2    | Myxoma                            | Unknown        | 45/NA   | 22            | Left M1          | TPA + Solitaire® + Trevo® + intracranial stent Solitaire® | TICI 0         | NIHSS 19, mRS 4 | [8]       |
| 3    | Myxoma                            | Unknown        | 34/NA   | 26            | Left M1, A2      | TPA+ Forced suction thrombectomy (Optimo® + Penumbra®)  | TICI 3         | NIHSS 3, mRS 2  |           |
| 4    | Myxoma                            | Unknown        | 46/M    | NA            | Left carotid T   | TPA+ Forced suction thrombectomy (Optimo® + Penumbra®)  | TICI 2b        | Left eye blindness | [9]       |
| 5    | Myxoma                            | Unknown        | 34/M    | 9             | Right M1 proximal | TPA + Merci®                                            | TICI 3         | mRS 2           | [10]      |
| 6    | Papillary fibroelastoma           | Unknown        | 62/M    | 24            | Left M1          | TPA + Solitaire®                                      | TICI 2b        | NIHSS 10, mRS 3 | [11]      |
| 7    | Unclassified sarcoma              | Known          | 55/M    | 22            | Right distal ICA | Penumbra®                                             | TICI 3         | Improved (details unknown) | [12]      |
| 8    | Myxoma                            | Unknown        | 4/M     | 16            | Left M1          | TPA+Solitaire®                                         | TICI 3         | Only mild weakness |           |
| 9    | Myxoma                            | Unknown        | 70/M    | 11            | Left carotid T, M1,M2 | TPA + Penumbra® + Trevo®                              | TICI 2b        | NIHSS 1, mRS 2  | [14]      |
| 10   | Papillary fibroelastoma           | Unknown        | 75/M    | 18            | Left M1          | Penumbra®                                             | TICI 3         | Improved (details unknown) | [15]      |
| 11   | Breast tumor                      | Known          | 62/F    | 19            | Left PCoA, M1, A1 | Merci®                                                | No flow through the MCA | Motor aphasia, right hemiplegia, NIHSS 1 | [4]       |
| 12   | Melanoma                          | Known          | 22/F    | 4             | Left MCA         | Forced suction thrombectomy                             | TICI 3         |                 | [5]       |
| 13   | Lung adenocarcinoma               | Known          | 69/F    | 16            | Right M1         | Mechanical clot retrieval                                | TICI 2b or 3   | Death           | [6]       |
| 14   | Undifferentiated sarcoma          | Unknown        | 37/M    | 13            | Right proximal ICA | TPA + Trevo®                                           | TICI 2b        | NIHSS 4, mRS 2  | Present case |           |

F: female, ICA: internal carotid artery, NA: not available, NIHSS: National Institute of Health Stroke Scale, M: male, MCA: middle cerebral artery, mRS: modified Rankin Scale, PCoA: posterior communicating artery, TICI: Thrombolysis in Cerebral Infarction, TPA: tissue plasminogen activator

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