An Autopsy Case of Intimal Sarcoma of the Abdominal Aorta with Bone Metastasis and Lymph Node Metastasis: A Case Report and Review of the Japanese Literature

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Abstract

A 73-year-old man complained of sternoclavicular joint pain; blood tests revealed elevated C-reactive protein. The patient developed delirium; magnetic resonance imaging showed metastatic bone tumors. He died two weeks after admission. Autopsy revealed abdominal aortic intimal sarcoma with metastasis to the peritracheal lymph nodes and sternum. Peripheral arterial embolism and bone metastasis are common symptoms of aortic intimal sarcoma, which implies a place for aortic intimal sarcoma in differential diagnoses of embolism or bone tumors of unknown origin.

Key words: abdominal aortic intimal sarcoma, lymph node metastasis, bone metastasis, delirium, elevated C-reactive protein

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Introduction

Primary aortic intimal sarcoma is a rare disease. The first case of aortic intimal sarcoma in Japan was reported in 1989 by Miyakoshi; to date, there have been 14 reported cases in Japan (1-14). Most patients develop tumor embolisms, but because early diagnosis is difficult, the disease has a poor prognosis and is often first diagnosed in autopsy (1, 2, 4, 6, 13-18).

We report a 73-year-old man who was diagnosed on autopsy with abdominal aortic intimal sarcoma. He presented with pain in the left sternoclavicular joint, an elevated serum C-reactive protein (CRP) level, and delirium as initial symptoms. We report this unique case, in which the patient developed pain consequent to bone metastasis and had lymph node metastasis.

Case Report

A 73-year-old man with delirium was admitted to our hospital complaining of left sternoclavicular joint pain. He had a history of diabetes mellitus, dyslipidemia, and stent placement for right renal arterial stenosis in 2011. In early August 2013, he experienced pain around the left sternoclavicular joint, and approximately the same time, he was unable to use either the television remote control or his car seatbelt. Although he visited his local physician several times, his symptoms did not improve and blood tests revealed no significant abnormalities except for an elevated CRP level. In mid-August, he developed delirium, and came to our hospital for detailed examination.

Physical examination findings on admission included: height, 160 cm; weight, 63 kg; body temperature, 36.8°C; blood pressure, 150/89 mmHg; and heart rate, 100 beats/min. Pressure pain was observed in his chest at the left and right sternoclavicular joints, sternum, and near the right fourth and fifth ribs. Examination of the head, abdomen, back, and limbs revealed no abnormalities, and no lymph node swelling was noted. Neurologically, the patient presented with slightly reduced consciousness; his Glasgow Coma Scale score was 14 (E4V4M6); however he scored 9 of a possible 30 on Hasegawa’s Revised Dementia Scale. No neck stiffness was seen, and no cranial nerve, motor system,
Laboratory testing performed on admission revealed the following: white blood cell count, 7.4×10^3/μL (reference range: 4.5-7.7×10^3/μL); CRP, 20.04 mg/dL (reference range: 0.0-0.3 mg/dL); red blood cell count, 3.13×10^6/μL (reference range: 4.32-5.07×10^6/μL); hemoglobin, 8.3 g/dL (reference range: 13.5-15.7 g/dL); serum iron, 33 μg/dL (reference range: 100-150 μg/dL); ferritin, 1,276.6 ng/mL (reference range: 18.6-261 ng/mL); blood glucose, 254 mg/dL (reference range: >one half of blood sugar); and hemoglobin A1c, 8.6% (reference range: 4.6-6.2%). Electrolytes, vitamins, renal function, and hepatic function were normal. Blood coagulation testing revealed fibrin degradation products, 5.5 μg/mL (reference range: 0-10 μg/mL), and D-dimer, 1.2 μg/mL (reference range: 0.0-1.0 μg/mL). Serum immunological testing revealed no specific abnormalities. Levels of tumor markers revealed that soluble interleukin-2 receptor (sIL-2R) level was 1,350 IU/mL (reference range: 124-466 IU/mL), and no other abnormalities. As indicated, blood test results showed elevated levels of CRP, ferritin, sIL-2R, fibrin degradation products, and D-dimer. Examination of cerebrospinal fluid revealed a nucleated cell count of 0/μL (reference range: 0-5/μL); protein, 83 mg/dL (reference range: 10-40 mg/dL); glucose, 102 mg/dL (reference range: >one half of blood sugar); anti-N-methyl-D-aspartate antibody was negative. Results of polymerase chain reaction testing for herpes simplex virus deoxyribonucleic acid (DNA) and cytodiagnosis were negative.

Following admission, the patient often developed severe delirium at night and complained of pain in the left and right sternoclavicular joints, sternum, and right fourth and fifth ribs. MRI of the mediastinum revealed a low-intensity signal in the sternum on T1-weighted imaging (T1WI) and a high-intensity signal on T2-weighted imaging (T2WI) and fat-suppressed T2WI (Fig. 1). A low-intensity signal was observed in the right pedicle of the upper thoracic spine on T1WI, and a tumor shadow with a high-intensity signal was seen on T2WI. Gallium scintigraphy revealed abnormal uptake in the sternum (Fig. 2). Further blood test results showed an elevated CRP level; however, there were no signs of infection, such as fever. Results of blood, urine, and cerebrospinal fluid cultures were also negative. The primary tumor could not be identified by MRI or scintigraphy findings. However, a metastatic or multifocal bone tumor was suspected. After two weeks of testing to identify a primary tumor lesion, the patient developed vomiting and abdominal pain and subsequently died.

An autopsy was performed approximately 2 hours after the patient’s death. Macroscopic pathological findings revealed necrosis extending from the duodenum to the ileum. No macroscopic lesions were observed in the ribs or vertebrae, and no macroscopic masses were observed in the lungs, esophagus, stomach, intestinal tract, liver, spleen, pancreas, bladder, prostate, testicles, or kidneys. Calcification or sensory system abnormalities were observed. No symptoms of Parkinsonism, such as muscle rigidity and tremors, were noted.
was seen in the coronary arteries, but no valve vegetation was observed. No macroscopic masses were observed in the aorta from the chest to the abdomen. Spindle tumor cells were observed microscopically in the abdominal aortic intima. We could not find tumor cells in lymphatic vessels. Immunohistochemical staining of tumor cells were positive for vimentin (Fig. 3), which suggests that the tumor cells were derived from mesenchymal cells. Among other tested markers, AE1/AE3, S-100 protein, CD56, and desmin were negative; αSMA (smooth muscle actin) and HHF-35 (muscle-specific actin) were almost negative; and Ki-67 was positive for 16% cells. Spindle tumor cells were observed in the superior mesenteric artery (Fig. 4). Proliferation of spindle tumor cells was observed in the ribs, thoracic vertebrae,
Aortic intimal sarcoma is a malignant mesenchymal tumor that arises in the aorta, in which the defining feature is intraluminal growth that obstructs the lumen of the vessel of origin and seeding of emboli to peripheral organs (17, 18). Most patients with aortic intimal sarcoma are middle-aged and have symptoms of embolism (17, 18). Tumor emboli can cause intestinal ischemia and metastasis in bone, adrenal gland, spleen, brain, heart, kidney, liver, lung, and skin (18). The prognosis for patients with aortic intimal sarcoma is poor, with survival of only a few months (17, 18).

In our case, the pathological findings led to a diagnosis of abdominal aortic intimal sarcoma, revealing peritracheal lymph node metastasis and bone metastasis to ribs, vertebrae, and sternum near the sternoclavicular joint. Autopsy and immunohistochemical findings showed that tumor cells were mainly in the vessel and originated from mesenchyme. Tumor embolism of the superior mesenteric artery resulted in intestinal necrosis extending from the duodenum to the ileum, which was considered to be the direct cause of death. In the neuropathological examination, the patient exhibited no invasive tumor cells into the central nervous system. Recent study showed that acute-onset dementia might be relevant to paraneoplastic neurological syndrome, especially paraneoplastic encephalomyelitis (19). Most paraneoplastic neurological syndrome cases were accompanied by carcinoma such as small cell lung cancer, breast cancer, and gynecologic tumor (20). Only a few cases with paraneoplastic neurological syndrome accompanied by sarcoma have been reported. We could not clearly diagnose his psychiatric symptoms as Lewy body disease, bad general condition, or paraneoplastic neurological syndrome. Although his delirium might have been caused by Lewy body disease, it was intensified by the general deterioration.

A literature search revealed only 14 cases of primary aortic intimal sarcoma in Japan (1-14) (Table). Although the age at onset of this disease ranges from the 20s to the 70s, it seems to be more common among older people, with seven of the 15 reported patients (including ours) being in their 70s. Initial symptoms commonly include symptoms related to embolism, such as pain in the abdomen, back, and extremities. The CRP levels were elevated in five of the 7 patients in whom CRP values were available, which suggests that many patients such as our case have elevated CRP. Of the three patients with psychiatric symptoms, delirium was noted only in our case, indicating that delirium is a rare symptom of this disease. Metastasis to multiple organs was reported in seven patients. Bone metastasis was the most common sign, followed by adrenal metastasis, renal metastasis, and splenic metastasis. Burke and Virmani reported a review of cases of aortic intimal sarcoma with metastasis to lymph nodes (21), but lymph node metastasis in this disease is rare. Although tumor cells may metastasize by blood flow, the mechanism and frequency of the metastasis to the lymph nodes are unclear and warrant further study.

Most cases of this disease present with symptoms of em-
The authors state that they have no Conflict of Interest (COI).

### Table. Comparison of Our Patient’s Characteristics with 14 Previous Japanese Case Reports of Primary Aortic Intimal Sarcoma.

| Case | Year | Age/ gender | Primary symptom | PL | CRP | Metastatic lesion | Cause of death |
|------|------|-------------|----------------|----|-----|------------------|----------------|
| 1    | 1989 | 50/M        | back pain      | TA | elevated | Lu, S, K, Bo | MOF |
| 2    | 1992 | 76/M        | back pain      | T  | normal   | Ag            | MI  |
| 3    | 1996 | 54/F        | fever          | T  | elevated | no            | MOF |
| 4    | 2000 | 73/M        | dyspnea        | A  | n.d.     | P, S, K, Ag | MOF |
| 5    | 2002 | 75/M        | abdominal pain | T  | n.d.     | Li, K, Ag, SI | MOF |
| 6    | 2002 | 78/M        | dyspnea        | T  | -        | no            | MOF |
| 7    | 2005 | 63/M        | lower limbs pain | TA | n.d.     | no            | MOF |
| 8    | 2008 | 78/M        | left upper limb pain | T  | n.d.     | no            | survive |
| 9    | 2008 | 49/M        | trachyphonia   | T  | n.d.     | no            | survive |
| 10   | 2009 | 69/M        | back pain      | T  | elevated | no            | survive |
| 11   | 2010 | 69/M        | back pain      | T  | n.d.     | no            | RT  |
| 12   | 2011 | 70s/M       | abdominal pain | T  | n.d.     | no            | survive |
| 13   | 2013 | 70/M        | fever          | T  | elevated | Br, Li, S, Bo | pneumonia |
| 14   | 2014 | 60s/M       | dyspnea        | TA | n.d.     | Bo, Ag        | heart failure |
| our case | 2016 | 73/M        | ostealgia      | A  | elevated | Bo, Ly        | necrosis of SI |

A: abdominal aorta, Ag: adrenal glands, Bo: bone, Br: brain, F: female, K: kidney, Li: liver, Lu: lung, Ly: lymph node, M: male, MI: myocardial infarction, MOF: multi-organ failure, n.d.: not documented, P: pancreas, PL: primary lesion, RT: recurrence of tumor, S: spleen, SI: small intestine, T: thoracic aorta, TA: thoracoabdominal aorta

bolism, and in some cases, the diagnosis was made by surgery (3, 5, 7, 9-12). However, antemortem diagnosis is difficult, and most cases are diagnosed by autopsy (1, 2, 4, 13-16). In all cases, prognosis is poor, with death generally occurring within several months of onset (15-18). Our patient presented with pain resulting from a metastatic tumor and elevated CRP prior to exhibiting symptoms of embolism. Of the patients listed in Table, only the patient reported by Yoshikawa et al. (13) and our patient are thought to have developed symptoms of this disease because of metastatic tumors. Although this disease is difficult to diagnose early, it must be considered in differential diagnoses of patients with elevated CRP levels who also show symptoms of embolisms of unknown origin or symptoms of metastatic tumors.

The authors state that they have no Conflict of Interest (COI).

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