A 47-year-old male smoker presented with sudden numbness and weakness of the right hand and was immediately admitted to the Kitasato Institute Hospital. Magnetic resonance imaging of the brain revealed multiple fresh infarcts in the left frontal cortex. Transthoracic echocardiography and echography of the carotid arteries did not show any abnormalities, and blood tests did not reveal coagulopathy or collagen diseases. Transesophageal echocardiography showed a mobile mural mass in the aorta attached to the wall from the arch to the descending aorta (Figure 1a). The mass was characterized by an irregular surface and heterogeneous contents. The mass extended to the arch when the probe was pulled up. Figure 1b shows a vertical transesophageal echocardiography view at the level of the aortic arch. A smooth mass had spread over the minor curvature of the arch (Figure 1b, left). Blood flow between the mass and the wall (Figure 1b, right) suggests a space between them. The abnormal mass occupied part of the aortic lumen (Movie in the online Data Supplement). Contrast-enhanced computed tomography showed that an irregularly shaped mass was located in the same region as that revealed by transesophageal echocardiography (Figure 2). The computed tomography scan also indicated left renal infarction. The mass was suspected of being a mural thrombus, and anticoagulant therapy was started. The patient gradually regained grip strength and was referred to our hospital to prevent further embolic complications by surgical removal of the intraluminal mass.

The aortic segment affected by the mass was resected under hypothermic circulatory arrest with antegrade cerebral perfusion through a median plus a lateral thoracotomy. The tumor occupied most of the aortic lumen (Figure 3). The excised aortic segment was replaced by a prosthetic graft with 3 branches that were anastomosed to the brachiocephalic, left carotid, and left subclavian arteries. The patient recovered uneventfully.

Histopathological examination revealed severe atherosclerosis of the resected aorta, left carotid, and subclavian arteries (Figure 4, a and b). Atypical spindle cells with pleomorphic nuclei (Figure 4c) were mainly localized within the atherosclerotic lesion but had penetrated through the tunica intima into the adventitia in some areas (Figure 4, d and e). These cells were immunohistochemically positive for MIB-1 and negative for desmin, S-100, CD34, CD31, Factor VIII, and cytokeratin AE1/AE3 (data not shown). These morphological and immunohistochemical findings indicated a diagnosis of undifferentiated aortic intimal sarcoma.

The patient was given local radiotherapy and adjuvant chemotherapy with ifosfamide and doxorubicin. A more detailed examination revealed multiple metastatic sarcomas in the back and the upper and lower extremities in addition to carcinomatous pericarditis (Figure 5). Although adjuvant therapy reduced some of the peripheral tumors and the patient...
appeared well during the immediate postoperative period, dyspnea on exertion appeared and gradually worsened. The patient died 9 months after surgery as a result of multiple organ failure with disseminated intravascular coagulopathy.

Primary tumors of the aorta are rare and mostly constitute sarcomas that are classified as intimal or mural according to the site of origin, and they are typically poorly and better differentiated, respectively. Highly invasive undifferentiated aortic intimal sarcomas are exceptionally rare and are often diagnosed after death, and fewer than 100 cases have been described. Most sarcomas originate from the descending thoracic or abdominal aorta. The median survival period of patients with an established diagnosis is only 7 months. Earlier diagnosis and treatment constituting en-bloc resection followed by chemotherapy or radiotherapy might prolong survival, but management is frequently palliative because most patients receive the diagnosis when the tumor is at an advanced stage. Identifying such tumors before they cause peripheral emboli or systemic metastasis is difficult. Although intraluminal tumors are difficult to distinguish from atherosclerotic thrombi only by diagnostic imaging, the aorta should be evaluated as soon as tumor embolisms are diagnosed or cardiac/carotid embolic sources are excluded. A thorough diagnostic evaluation should also determine the presence or absence of metastasis. Patients with metastatic tumors are unlikely to survive for long periods despite appropriate treatment.

Disclosures

None.

References

Figure 4. Morphology of atherosclerotic lesion in aorta. Staining of resected aorta with hematoxylin and eosin (a) and elastic Van Gieson (b) shows severe atherosclerosis with atypical spindle cell infiltration, mainly in atherosclerotic lesion. High magnification (c) reveals nuclear pleomorphism of atypical spindle cells (hematoxylin and eosin stain). Staining with hematoxylin and eosin (d) and elastic Van Geison (e) shows that atypical cells have penetrated through tunica media into adventitia.

Figure 5. Computed tomography and magnetic resonance imaging of patient after surgery. Computed tomography 3 months after surgery shows metastatic lesion (arrow) in dorsal surface of left scapula with erosive scapular changes (a). This lesion was preoperatively undetectable (see Figure 2, c through e). Another metastatic lesion (arrow) located lateral to the head of left humerus (b) was outside preoperative computed tomography scanning range. Magnetic resonance image 2 months after surgery shows a well-demarcated, metastatic lesion measuring $8.5 \times 7 \times 4$ cm with internal necrosis inside the soleus muscle (c and d).
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