SHORT REPORT

Epithelioid Hemangioendothelioma Causing Acute Abdominal Aortic Obstruction and Bilateral Limb Ischemia

A. Doi,1 A. Ishida,2 M. Imamaki,2 H. Shimura,2 Y. Niitsuma2 and M. Miyazaki2

1Department of Cardiovascular Surgery, Narita Red Cross Hospital, 90-1, Iidacho, Narita, Chiba 286-8523, Japan
2Department of General Surgery, Graduate School of Medicine, Chiba University, 1-8-1, Inohana, Chuo-ku, Chiba City, Chiba 260-8677, Japan

A 32-year-old man was admitted with sudden onset of bilateral leg pain. Acute abdominal aortic occlusion was revealed by means of examinations and the patient underwent an emergency operation. Both legs were recanalized following thrombectomy from bilateral femoral arteries and the aorta. Because of the unusual appearance of the emboli, it was sent for pathological examination and was diagnosed epithelioid hemangioendothelioma. Epithelioid hemangioendothelioma is a rare vascular tumor of intermediate malignancy. It usually originates from veins and rarely presents with acute ischemic symptoms.
© 2007 European Society for Vascular Surgery. Published by Elsevier Ltd. Open access under CC BY-NC-ND license.

Keywords: Epithelioid hemangioendothelioma; Vascular tumor; Acute aortic obstruction; Limb ischemia.

Introduction

Tumors of vascular origin are rare, and tumors of aortic origin, even less frequent. We encountered a case of an acute aortic occlusion which was surgically treated. Postoperatively, the histological diagnosis was epithelioid hemangioendothelioma.

Epithelioid hemangioendothelioma is a rare vascular tumor of intermediate malignancy, usually venous in origin.1,2 It generally occurs within soft tissues as a solitary lesion. It has a low but definite possibility to metastasize, and the current treatment of choice is complete excision of the tumor. We herein describe the dilemmas of a rare tumor of malignancy causing an emergent situation.

Report

A 32-year-old male was referred for cardiovascular consultation because of delirium, and bilateral leg pain that had continued for more than 7 hours. On physical examination, he was found to have paralysis, and paresthesia dominantly observed in the left leg and bilateral absent femoral pulses.

Significant laboratory findings included an elevated aspartate aminotransferase of 111 IU/l, lactate dehydrogenase of 668 IU/l, creatine phosphokinase of 8550 IU/l, C-reactive protein of 11.6 mg/dl. Electrocardiogram showed normal sinus rhythm. Transthoracic echocardiogram did not reveal any thrombus or tumor inside the heart. Enhanced computed tomography (CT) of the whole aorta and pelvis showed occlusion of the aorta from just above the aortic saddle to bilateral common iliac arteries. Aortogram showed that the aorta was occluded below the origin of the inferior mesenteric artery. The right common iliac artery was enhanced by collateral arteries, but the left iliac artery was not visualised (Fig. 1).

Under the clinical diagnosis of aortic thrombembolism, we first attempted thrombectomy from bilateral femoral arteries, but improvement of blood flow could not be obtained. Therefore, we decided to perform transperitoneal aortotomy, and successfully took out the emboli from the abdominal aorta to bilateral common iliac arteries using balloon catheters.

1533–3167/00017 + 03/0 © 2007 European Society for Vascular Surgery. Published by Elsevier Ltd. Open access under CC BY-NC-ND license.
Intraoperatively, macroscopic change of the adventitia of the aorta was not seen and the specimen was extracted without much stress. Flow was reestablished to both legs. As we expected that the aortic obstruction was due to thromboembolism, and did not strongly suspect tumors of malignancy to be the cause, we did not excise the aorta. The longitudinal aortic incision was closed with a prosthetic patch graft.

The emboli macroscopically consisted of red, bloody and grayish-white specimen. As the emboli seemed slightly atypical, the specimen was sent for pathological examination. Reperfusion of the lower limbs was filtered from the femoral vein and the internal jugular vein with 2 sets of continuous hemodiafiltrations concomitantly to prevent myonephropathic metabolic syndrome. Postoperatively, compartment syndrome was observed in both legs. Bilateral fasciotomies were performed immediately, and both limbs were salvaged.

Interestingly, microscopical findings of the emboli revealed that there were round-like to fusiform tumor cells that showed epithelioid, funicular, vesicle proliferation in a myxoid background. Its dysplasia was mild, cell division was rare, and less than 5 mitoses/10 high-power fields were seen. Single cell vacuolization was seen among the tumor cells. Immunohistochemical studies demonstrated the tumor cells CD31 and CD34 positive. As a consequence, the emboli were proved epithelioid hemangioendothelioma (Fig. 2a, b).

Postoperative transthoracic echocardiography and CT did not reveal any abnormalities in the heart or proximal aorta. 18F-fluorodeoxyglucose positron emission tomography (FDG-PET) showed increased FDG uptake only in the patch graft. We assumed that the tumor arose from the abdominal aortic wall where the aorta was occluded. Adjuvant therapy was not applied. Four years after the operation, the patient is able to walk with no evidence of recurrence or metastasis.

Discussion

Epithelioid hemangioendothelioma (EHE) is a rare vascular tumor of intermediate malignancy, usually venous in origin.1,2 It can occur within soft tissues...
or parenchyma of organs such as liver, lung, and bone. Affecting the sexes about equally, the age of onset varies greatly.

EHE usually develops as a solitary, slightly painful mass in either superficial or deep soft tissue. In some cases, symptoms such as edema or thrombophlebitis due to occlusion of the vessels are seen. But because EHE of arterial origin is quite rare, ischemic symptoms as in this case are not often seen. The death rates from the disease in the lung and liver vary from 40% to 65%, respectively, compared with a 13% death rate in soft tissues.

An important feature in EHE is the presence of intracytoplasmic vacuoles occasionally containing erythrocytes, which reflects the primitive vasoformative character of EHE. This character is also called single cell vacuolization and was seen in the present case. It has been reported that high mitotic rate (>6 mitoses/10 high-power fields) correlated clearly with bad prognosis, which was not the case here. Immunohistologic studies are mandatory for the diagnosis of EHE. CD31, CD34, factor VIII, and vimentin are the markers that are commonly used. In the present case, CD31 and CD34 were used as markers and were positive.

Although EHE is considered a tumor of intermediate malignancy, it is reported that it has a potential to recur in 12–13% and metastasize in 21–61% of the cases. The most common site of metastasis is the lung and the liver, and it can occur after quite a period of time. In the present case, it is doubtful that complete resection of the tumor was achieved. Due to the emergent situation and the unexpected cause of the aortic occlusion, we were not able to excise the aorta where the primary site of the tumor is suspected. Fortunately, no signs of regional or distant metastasis have been observed. Resection of the aorta may become necessary in the future. Careful follow-up utilizing imaging such as CT is necessary.

References


Accepted 13 September 2007