A case of malignant lymphoma invading the aorta, which mimicked aortic dissection

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Abstract
In rare cases, malignant lymphoma may invade the aorta and mimic other aortic diseases; thus, it is sometimes difficult to diagnose from image examinations. In such patients, aortic disease may also be caused by degeneration of the aortic wall or necrosis due to chemotherapy. We present the case of a 78-year-old man who was initially diagnosed with an aneurysm or tumor adjacent to the aorta, in which open surgery was required for replacement of the abdominal main vessel. We discuss treatments for malignant lymphoma invading the aorta with reference to previous reports.

Keywords: Malignant lymphoma, Aorta invading, Aortic root replacement

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Introduction

Tumors occurring adjacent to the aorta often invade the aorta. These neoplasms sometimes mimic other aortic diseases and are difficult to diagnose from image examinations. In most cases, these occur in patients with esophageal cancer or lung cancer; reports of aortic invasion in patients with malignant lymphoma are rare. We could find only a small number of case reports describing malignant lymphoma adjacent to the aortic wall; however, those case reports noted that malignant lymphoma sometimes invades the aorta and causes aortic diseases, such as aortic dissection or aneurysm. Some authors reported that such cases may require treatment to avoid aortic rupture, meaning that such lesions should be investigated, despite the fact that there are no guidelines for the treatment of lymphoma with aortic invasion.

We herein present a rare case of malignant lymphoma affecting the abdominal aorta. The patient was treated by open surgery with replacement of the abdominal aorta. The surgical specimen showed evidence of malignant lymphocytes invading the tunica externa of the aorta.

Case report

A 78-year-old man who had been admitted to our hospital and treated for pancreatitis underwent a CT scan following antibacterial drug therapy. The images showed splenomegaly, and several enhanced large lymphocytes in the para-aorta, hepatoduodenal ligament, and mesentery, leading to the suspicion of malignant lymphoma. It also showed a shadow of 27×38 mm in size located adjacent to the abdominal aorta, originating from between the renal arteries and common iliac arteries, suggesting an aortic aneurysm or a tumor invading the aorta. The tumor was enhanced in delayed phase and appeared similar to enlarged lymph nodes (Fig. 1). To obtain diagnostic specimens, we decided to perform open surgery with biopsy of the enlarged abdominal lymphocytes and to simultaneously check whether or not the aortic lesion was malignant lymphoma. The patient had a fever before surgery due to pancreatitis but had not experienced night sweat or weight loss. Routine hematological exams revealed no abnormalities other than HCV positivity. His relevant medical history included STEMI, hypertension, vasospastic angina, reflux esophagitis, prostatic hyperplasia, and inguinal hernia, but he had no history of cancer.

Surgery started with a biopsy, which obtained some lymphocytes for a histological analysis. After the procedure, we separate the retroperitoneum from the abdominal aorta and identified the lesion. There was a mass...
adhering to the aortic wall and the shadow on located around the abdominal aorta was found to be a tumor adjacent to the aorta. The aortic wall was white and edematous. We attempted to remove the mass from the aorta. However, the tumor was found to have invaded the tunica externa; thus, it was impossible to remove the tumor from the aorta without damaging the aorta wall. We therefore decided to complete the surgery by performing replacement of the aorta instead of mass excision.

Microscopic observation of the specimen, which was performed as a pathological examination, revealed that part of the abdominal aortic wall contained a tumor-like mass consisting of follicular lymphatic nodules that were covered by tunica media (Fig. 2). The lymphatic modules were CD10- and BCR-2 positive, indicating that the tumor located adjacent to the aortic wall was follicular lymphoma.

The postoperative course was uneventful. The patient was discharged on postoperative day 13. After the procedure, chemotherapy with obinutuzumab and bendamustine (GB therapy) was initiated for malignant lymphoma. The patient remains alive at approximately one year after the operation.

Fig. 1 A shadow of 27×38 mm, indicating an aortic aneurysm or tumor adjacent to the aorta was observed. We also found splenomegaly, several enhanced large lymphocytes in the para-aorta, hepatoduodenal ligament, and mesentery. Based on these findings, malignant lymphoma was suspected.

Fig. 2 A pathologic examination revealed that the abdominal aortic wall contained a tumor-like mass consisting of follicular lymphatic nodules covered by tunica media. The lymphatic modules were CD10- and BCR-2-positive. This evidence indicated that the lesion adjacent to the aortic wall was follicular lymphoma.
Discussion

Although neoplasms such as esophageal cancers or lung cancers often invade the aortic wall, there are few reported cases involving adhesion of malignant lymphoma to the aorta\(^1,4\). Malignant lymphoma is a cancer that can start adjacent to any lymphatic organ and spread to anywhere in the body. Sometimes lymphoma occur in the para-aortic lymph nodes surrounding the aorta and mimic aortic diseases, such as aortic aneurysm or aortic dissection; however, invasion of the aorta is rare. In some reported cases involving adhesion of malignant lymphoma to the aorta, surgical treatment was required. These reports note that invasion of the aorta by lymphoma may cause aortic disease\(^2,5-8\). Alexander et al. and Matthew et al. reported that ectopic lymphoma cells in the aortic wall may cause degeneration of the vessel wall and promote the development of aortic aneurysm\(^6,9\). In addition, Oda et al. reported that tumor cells may have affinity to arteriosclerotic lesions because they appeared not only in the aortic aneurysm but also in the arteriosclerotic parts without a visible mass\(^7\). Some other studies have reported that the malignant lymphoma sometimes causes aortic disorder.

Malignant lymphoma is usually treated by chemotherapy. However, the aortic walls which are invaded by lymphoma cells are weakened, therefore aortic rupture after chemotherapy due to tumor necrosis was a concern. Yiu et al. and Estrera et al. noted that those cases require subsequent investigations before the chemotherapy\(^2,4\). Repair of the aorta before chemotherapy is necessary in order to avoid such potentially fatal events.

It may be difficult to decide how to investigate lesions located adjacent to the abdominal aorta. Certainly, it is possible to treat patients with aortic invasion by open surgery; however, endovascular stent grafting also works effectively. Our search of the PubMed database revealed only 15 reports describing aortic disease caused by invasion of the aorta by malignant lymphoma. Table 1 shows the aortic disorder, type of lymphoma, and treatment in each case. Six of these patients were treated by open surgery and seven were treated by endovascular surgery; none of the patients died due to the investigations\(^2,9\). This may mean that both open surgery and endovascular procedures are both sufficiently effective and indicate that it is appropriate for surgeons to choose the approach on an individual basis. For example, Sumbul et al. described that they treated a ruptured aorta with invasion by malignant lymphoma by endovascular surgery\(^3\). In addition, Masuda reported that they treated the aorta adjacent to lymphoma in the same way\(^5\). Sumbul et al. reported that endovascular treatment of aortic rupture reduced procedure-related and early mortality and morbidity, while open surgery was associated with disadvantages in relation to bleeding and infection; thus, considering the condition of the patient, they chose to perform an endovascular procedure\(^3\). It is necessary for surgeons to consider the approach that best suits each patient.

In our case, when we found the abnormality on the CT scan, we suspected that the lesion was an aortic aneurysm or a tumor invading the aorta; however, we could not diagnose the lesion. We performed open surgery with the aim of obtaining specimens and diagnosing the lesion. Actually, we decided to perform the investigation because the tumor had invaded the tunica externa, which meant that we could not remove the tumor without resecting part of the abdominal aorta. Open surgery is

| Year | Author | Aortic Disease | Type of Tumor | Investigation |
|------|--------|----------------|---------------|---------------|
| 2002 | Williamson AE | Aortic aneurysm | Aggresive low grade lymphoma | Endovascular repair |
| 2005 | Estrera AL | Aortic aneurysm | Follicular lymphoma | Open surgery |
| 2007 | Lotto AA | Aortic dissection | Follicular lymphoma | Endovascular repair |
| 2012 | Sumbul AT | Aortic rupture | Mantle cell lymphoma | Endovascular repair |
| 2013 | Yiu WK | Aortic aneurysm | DLBCL | Endovascular repair |
| 2017 | Margaux Pontailler | Aortic rupture | Lymphocytic lymphoma | Open surgery |
| 2018 | Oda Y. | Aortic tumor | IVL | - |
| 2018 | Hiraoka T | Aortic aneurysm | DLBCL | Open surgery |
| 2018 | Jan Raupach | Aortic rupture | B cell lymphoma | Endovascular repair |
| 2019 | Paula C | Aortic rupture | DLBCL | Endovascular repair |
| 2019 | K. Masada | Aortic rupture | - | Endovascular repair |
| 2019 | Tanaka H | Aortic rupture | CMML | - |
| 2020 | Alexander M | Aortic rupture | SLL | Open surgery |
| 2020 | Matthew L. Inra | Aortic dissection | SLL/CLL | Open surgery |
| 2020 | Ogikubo M | Aortic aneurysm | Follicular lymphoma | Open surgery |
more invasive in comparison to endovascular surgery; however, we are of the opinion that open surgery is suitable for undiagnosed cases.

Conclusion

We reported a case of malignant lymphoma with aortic invasion. Such tumors can cause aortic diseases, such as aortic dissection or aneurysm, because they cause degeneration of the aortic wall. We believe that such aortic lesions must be treated in order to avoid aortic rupture and that open surgery and endovascular surgery are both appropriate approaches for the investigation of aortic lesions.

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