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**Multimodal treatments of Rt. gastroepiploic arterial leiomyosarcoma with hepatic metastasis: A case report**

Seo HI *et al.* Case of intra-abdominal arterial leiomyosarcoma with hepatic metastasis

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**Abstract**

Leiomyosarcoma of an artery is very rare, and cases with hepatic metastasis are even rarer. We describe a case of 70-year-old man who presented intra-abdominal hyper-vascular mass and hepatic mass on follow up computed tomography due to rectal cancer. It was diagnosed as a leiomyosarcoma of the right gastroepiploic artery with hepatic metastasis by surgical resection. Multiple metastases had recurred at the liver. He is alive more than 53 mo through multimodal treatments (three times of surgical resections, radiofrequency ablation, transarterial chemoembolization, chemotherapies and target therapy). Multimodal treatments, including active surgical resection, may be helpful in the treatment of aggressive diseases such as arterial leiomyosarcoma with metastasis.

**Key words:** Arterial leiomyosarcoma; Multimodal treatments; Hepatic metastasis; Intra-abdominal arterial leiomyosarcoma; Surgical resection

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**Core tip:** An arterial leiomyosarcoma (aLMS) is a very rare and aggressive disease. The prognosis is also very poor. We experienced a 70-years-old man who had an intra-abdominal (aLMS) with hepatic metastasis. He was treated multimodal treatments that consisted of 3 times of surgery, radiofrequency ablation, transarterial chemoembolization and target therapy. He was still alive after these treatments for 53 mo. Multimodal treatments could be helpful this kind of disease.

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**INTRODUCTION**

The <leiomyosarcoma> (LMS) is very rare malignant tumor. They usually originate in the smooth muscle of the soft tissues and uterus[1]. About 2% of LMS cases occur in the smooth muscle of the vessel wall and many of which (60%) occur in the inferior vena cava; the occurrence of LMS involving the veins is about 5 times higher than that of the arteries[1]. The most common site of arterial LMS (aLMS) is the peripheral artery, and the intra-abdominal artery is rare location to occur the aLMS[1]. To the best of our knowledge, this is the first presentation of intra-abdominal aLMS with distant single liver metastasis. We report the clinical course of aLMS that originated from the right gastroepiploic artery with hepatic metastasis during multimodal treatments [three times of surgical resections, radiofrequency ablation (RFA), transarterial chemoembolization (TACE), chemotherapies (CTx) and target therapy] and review the literature about aLMS.

**CASE REPORT**

A 70-year-old man had previous operation history owing to renal cell carcinoma and rectal cancer (pT2N0M0, stage IIA), 10 years and 6 months ago. Abdominal computed tomography (CT) and magnetic resonance imaging (MRI) performed 6 mo after the low anterior resection revealed new 47 mm sized hypodense hepatic mass and 23 mm sized hypervascular mass at great curvature side of stomach (Figure 1A, B). It was highly suspected to be a malignant gastrointestinal stromal tumor (GIST) with hepatic metastasis. Fluorine-18 fluorodeoxyglucose positron emission tomography/computed tomography (FDG PET/CT) demonstrated hypermetabolic low-density lesion in S8 of the liver and the greater curvature of the stomach with a maximum standardized uptake value (SUVmax) of 3.4 and 2.3, respectively (Figure 1C). For confirming the diagnosis, we planned to perform ultrasonography-guided core needle biopsy. The core needle biopsy specimen of the liver mass showed malignant spindle cell with increased mitosis (7/10 HPFs). Immunohistochemistry results were positive for desmin and smooth muscle actin (SMA) and negative for CD34, c-kit, DOG-1, S-100, and HMB45. The aLMS were more suspected than GIST in these findings. The mass in the greater curvature of the stomach showed high vascularity on endoscopic ultrasonography; therefore, fine needle aspiration biopsy was not performed because of a bleeding risk. Laparotomy was performed with a diagnosis of the omental GIST and primary hepatic LMS. The omental mass resection and S8 segmentectomy were performed. The omental mass was originated from Rt. gastroepiploic artery on surgical and microscopic field. This mass was a 3.0 cm × 2.7 cm sized aLMS (Figure 2A, 2B). Histopathology showed moderate cellular atypia, high mitotic rate (10/10 HPFs), and 2/3 histologic grade according to the FNCLCC grading system. Ki-67 proliferation index was 4.1% (Figure 2C). Immunohistochemistry results were positive for CD34, CD31, desmin and SMA and negative for c-kit, DOG-1 and S-100. The liver mass was a 5.0 cm × 3.0 cm × 1.5 cm sized metastatic aLMS with a clear resection margin (free margin: 0.3 cm). Ki-67 proliferation index was 9.3%. The patient was discharged on the 9th day after the operation without any complication. It was planned that adriamycin mono-CTx as an adjuvant treatment (4 cycles), but after the 3rd CTx, it was stopped because of neutropenic fever.

Abdominal CT was performed every 3 mo after the operation to check for recurrence. At 14 mo after the 1st operation, CT and MRI revealed 2.7 cm, 5 mm, and 8 mm sized metastatic masses on of the liver. No extrahepatic metastasis was noted on FDG PET/CT. Right anterior sectionectomy was performed 15 mo after the first operation. There were 3.0 cm × 2.7 cm and 1.0 cm × 1.0 cm sized metastatic vascular LMSs in pathology. Histopathology and immunohistochemistry showed a high mitotic rate (22/10 HPFs) and a Ki-67 proliferation index of 4.1%. The resection margin was very close to the mass (<1 mm). Ifosfamide mono CTx was administered after the surgery by 4 cycles.

At 8 mo after the 2nd operation, CT, MRI and FDG PET/CT revealed 1.6 cm and 1.4 cm sized seeding metastatic nodules on diaphragm and the liver. Diaphragm partial resection and intra-operative RFA was performed 9 mo after the 2nd operation (24 mo after the 1st operation). Diaphragm mass was also diagnosed as metastatic vascular LMSs. Etoposide-cisplatin CTx was administered after the surgery by 6 cycles.

At the 11th mo after the third operation, there were multiple recurrences of remnant liver in MRI and TACE (lipiodol 2 mL and Adriamycin 10 mg) was performed.

And palliative pazopanib was also started. The patient followed up during 28 mo after the 3rd operation (53 mo after the 1st operation). He was alive with stable hepatic metastasis controlled by CTx. Treatment modalities summarized at Figure 3.

**DISCUSSION**

Over half of aLMS cases originate from the pulmonary artery, followed by the extra-abdominal peripheral artery, and including this case, only 9 cases of aLMS originating from the intra-abdominal arteries, except the aorta, have been reported (Table 1)[2-9].

Compared to other origin LMS, the case of vascular LMS have worse prognosis. In case of LMS with metastasis, metastatic vascular LMS shows similar results as metastatic LMS of other origin[1]. However, the prognosis is not well-known owing to the low number of cases in aLMS. It is presumed that aLMS may be more aggressive than LMS, and hence, the prognosis is expected to be about the same or worse[1]. Because aLMS is more aggressive than LMS, and directly seed to artery; therefore, it has a higher possibility of metastasis[10].

Clinical signs of aLMS are diverse depending on the area of origin and most of them are due to mass effect[9,10]. In this case, the primary mass sized 3 cm was located in the intra-abdominal area. The patient had no symptoms owing to aLMS. Although, the size of the primary cancer of the right gastroepiploic artery was 3 cm, which was small, it was accompanied by distant metastasis at the time of diagnosis.

For a diagnostic confirmation, biopsy is needed, and radiological assisted core needle biopsy is preferred over open biopsy because there is low risk of complications[10]. However, like this case, if the mass is located in the intra-peritoneal region and shows hyper-vascular findings, bleeding could occur after core needle biopsy; moreover, bleeding control could be difficult in this location. PET-CT showed SUVmax values of 3.4 and 2.3 each, which had relatively low uptake at first, and uptake was not noted at lesion recurrence. More precise imaging studies needed to overcome this limitation.

There has been reported LMS cases treated with surgical resection, radiotherapy, and CTx[1,4,6,9].

CTx or chemoembolization has been the main treatment of LMS with hepatic metastasis[10]. Recently, RFA also shows good result for metastatic LMS[10]. However, recently, just like in other cases of metastatic cancer, liver resection shows better results[11]. If a resection of metastasis is possible, surgical treatment and additional treatment including CTx can lead to a good response.

Although aLMS showed aggressive clinical features, multimodal treatment (resection, CTx, RFA, chemoembolization and target therapy) might be helpful to manage this kind of disease.

**ARTICLE HIGHLIGHTS**

***Case characteristics***

A 70-years-old man visited due to an intra-abdominal mass and hepatic mass in follow up abdominal computed tomography (CT) scan after rectal cancer surgery.

***Clinical diagnosis***

In the CT and magnetic resonance imaging (MRI), it was diagnosed a malignant gastrointestinal stromal tumor (GIST) with hepatic metastasis.

***Differential diagnosis***

At the core needle biopsy of the liver, it was more suspected as a leiomyosarcoma (LMS). Before the surgery, these were omental GIST and hepatic LMS.

***Imaging diagnosis***

At first, it was diagnosed a omental GIST and hepatic metastasis in CT and MRI.

***Pathological diagnosis***

The surgical specimen diagnosed as aLMS with hepatic metastasis.

***Treatment***

Multimodal treatments were done (3 times of surgery, chemotherapy, transarterial chemoembolization, radiofrequency and target therapy).

***Related reports***

There were only 9 reports about intra-abdominal arterial leiomyosarcoma (aLMS). This is the first report of intra-abdominal aLMS with hepatic metastasis.

***Term explanation***

aLMS is very rare and aggressive disease. The prognosis is very poor. There were few reports of this disease. So the treatment is also not established.

***Experiences and lessons***

Active treatments using multiple modalities may be helpful for these kinds of patients.

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**Table 1 Literatures about intra-abdominal arterial leiomyosarcoma**

|  |  |  |  |  |  |  |  |
| --- | --- | --- | --- | --- | --- | --- | --- |
| **Reference** | **Sex** | **Age (yr)** | **Site** | **Symptom** | **Treatment** | **Metastasis** | **Follow-up** |
| Hopkins (1968)[2] | M | 55 | Common iliac | Claudication | Surgery | - | Died 3 wk |
| Stringer (1977)[4] | M | 49 | IMA | Pain, palpable mass | Surgery, RTx, CTx | Lung | Died 7 yr |
| Birkenstock *et al* (1976)[3] | M | 55 | Common iliac | Pain, palpable mass | Surgery | - | No evidence of disease |
| Gutman *et al* (1968)[7] | F | 55 | Common iliac | Claudication | Surgery | - | No evidence of disease |
| Delin *et al* (1990)[6] | F | 72 | Common iliac | Claudication, pain | Surgery | - | Died 7 mo |
| Gill *et al* (2000)[7] | F | 76 | Renal | Pain | Surgery | - | f/u 9 mo |
| Rohde *et al* (2001)[8] | F | 51 | Splenic | Pain, weight loss | Surgery + CTx (AD+ifosphamide) | - | f/u 1 yr |
| Blansfield *et al* (2003)[9] | F | 42 | Common iliac | Pain | Surgery | - | No evidence of disease |
| Current case | M | 70 | Rt. gastroepiploic | None | Multimodal | Liver | f/u 36 mo |

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**Figure 1 Diagnosis imaging of the patient.** A: Computed tomography showed 43 mm sized hypodense mass at S8 of the liver (thin black arrow) and 23 mm sized hypervacular mass at great curvature side of stomach (white arrow); B: Magnetic resonance image showed a well-defined encapsulated lesion (thin black arrow) in S8 of the liver, which showed a strong enhancement during the arterial dominant phase, with wash out during the delayed phase. The mass (thin black arrow) in the greater curvature of the stomach was accompanied by engorgement of the gastroepiploic vein: C: Fluorine-18 fluorodeoxyglucose positron emission tomography/computed tomography images showed a hyper-vascular mass at great curvature side of stomach (white arrow) and a hyper-vascular metastatic mass at S8 of liver (thin black arrow)



**Figure 2 The gross find of arterial leiomyosarcoma (aLMS) and pathological diagnosis.**The omental mass and A: Gross finding of aLMS. A white colored expending nodular mass was present on soft tissue. A medium sized artery was present on adjacent connective tissue; B: Tumor consisted with spindle cells and adjacent to medium sized vessels (H&E staining, ×40); C: Tumor cells showed spindle shaped nucleus with rounded end and eosinophilic cytoplasm (H&E staining, ×200). They formed fascicular pattern and made stage horn shaped vascular spaces, frequently.



**Figure 3 Timeline of multimodal treatments.**