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A Case of Huge Metastatic Diaphragmatic Leiomyoma with Right Atrial Thrombosis

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A benign metastasizing leiomyoma is a very rare presentation. It is mainly found in female who have a surgical history of uterine leiomyoma. We present a rare case of a huge diaphragmatic metastasizing leiomyoma with right atrial thrombosis. A 47-year-old female visited the cardiothoracic surgery outpatient department because of recurring neck swelling and pain. Contrast-enhanced computed tomography of the neck revealed acute thrombus in the right brachiocephalic vein and acute thrombophlebitis in the superior vena cava, left brachiocephalic vein, and internal jugular vein. Incidental right pleural effusion and large hepatic mass were detected. Low-molecular-weight heparin was used to improve thrombosis before surgery, and a chest tube was inserted to improve right pleural effusion. A large subphrenic tumor originating from the right diaphragm was removed and no liver invasiveness was detected. After surgery she treated using anticoagulant and thrombosis was decreased.

Keywords: Anticoagulants, Benign metastasizing leiomyoma, Diaphragm, Subphrenic tumor, Thrombosis

Introduction

A benign metastasizing leiomyoma (BML) is a very rare presentation. It is mainly found in female who have a surgical history of uterine leiomyoma. In most cases, metastasizing leiomyoma occurs in the pelvic cavity or lungs, but it rarely occurs in the ribs, vertebra, diaphragm, or skull base. Here, we report a huge diaphragmatic metastasizing leiomyoma with right atrial (RA) thrombosis.

Case report

A 47-year-old female visited the cardiothoracic surgery outpatient department one year ago because of recurring neck swelling and pain. The patient had no traumatic history but had a medical history that included hepatitis B virus treatment, iron replacement therapy. She underwent uterine myomectomy nine years ago, but it did not involve morcellation.

She was diagnosed and treated for acute thrombophlebitis in the right internal jugular vein (IJV) 3 years prior. The right neck pain and swelling occurred suddenly and were similar to the previous experience.

Contrast-enhanced computed tomography (CT) of the neck revealed acute thrombus in the right brachiocephalic vein and acute thrombophlebitis in the superior vena cava (SVC), left brachiocephalic vein, and IJV (Fig. 1). Incidental right pleural effusion was detected. CT of the chest revealed a large hepatic mass with obliteration of the inferior vena cava (IVC) (Fig. 2).

The patient consulted with physicians in hepatology and internal medicine who evaluated her via dynamic CT and magnetic resonance imaging (MRI) of

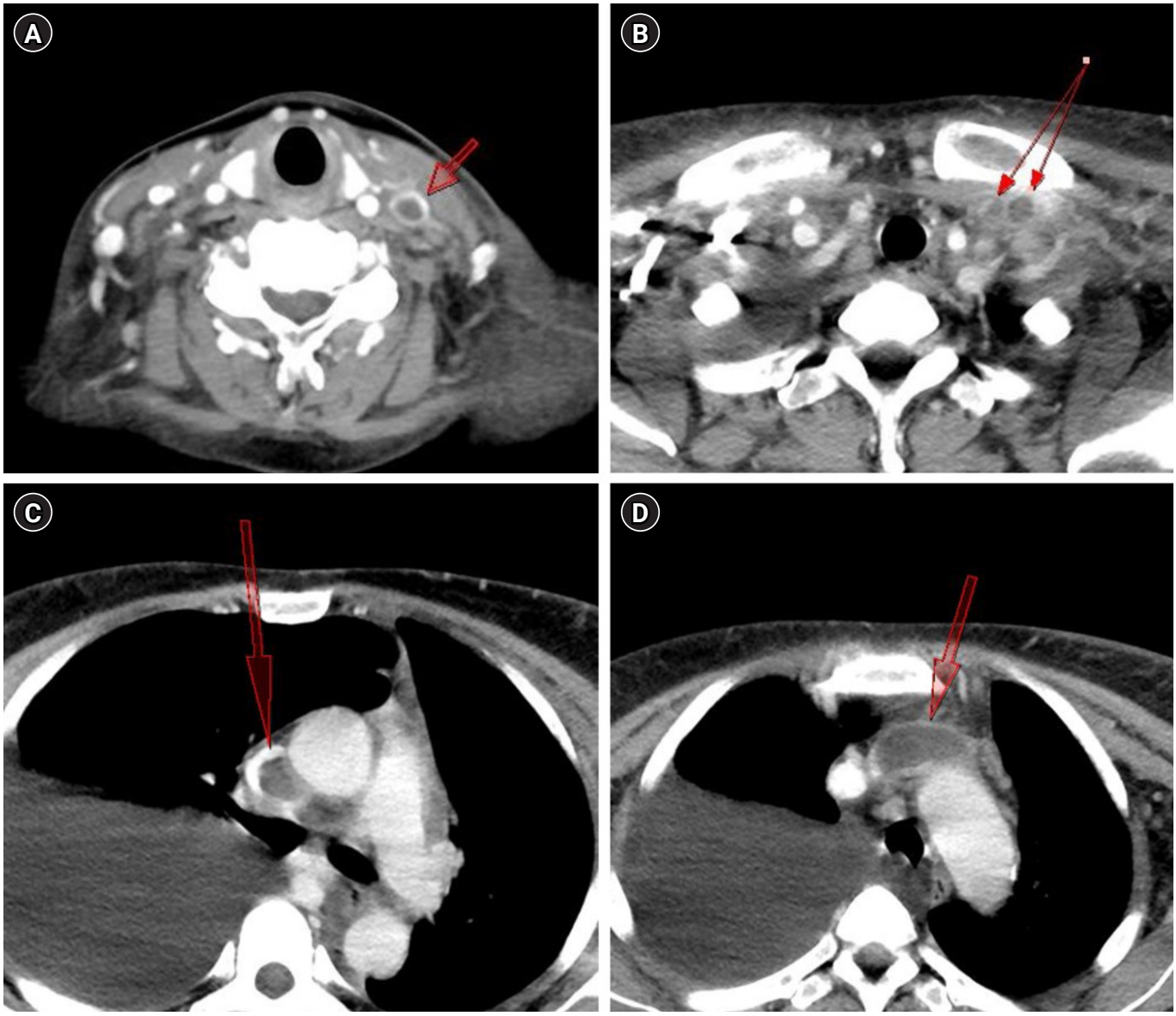

Fig. 1. A neck computed tomography (CT) image revealing acute thrombophlebitis (arrow) in (A) internal jugular vein, (B, C) left brachiocephalic vein, (D) superior vena cava.

the liver. These revealed a 12.5 cm × 18 cm × 22 cm heter ogeneous enhancing mass in the right lobe (Fig. 3). The huge mass origin was not clearly discriminated on CT and MRI, raising the possibility of hepatic malignancy. Therefore, a biopsy was performed to accurately determine its origin and make a diagnosis. The huge mass was biopsied with ultrasound and was confirmed to be leiomyoma.

The hepatobiliary surgeon and cardiothoracic surgeon contacted her for the operation. Low-molecular-weight heparin (LMWH) was used to improve RA thrombosis before surgery, and a chest tube was inserted to improve right pleural effusion. A large subphrenic tumor originating from the right diaphragm was removed and no liver invasiveness was detected. The diaphragm defect size was 4 cm × 4 cm and the cardiothoracic surgeon performed a simple closure (Fig. 4).

The pathological results indicated a BML (Fig. 5). LMWH was re-used on postoperative day 1 (POD1), and a non-vitamin K antagonist oral anticoagulant (NOAC) was used on POD7. Follow-up CT angiography was performed on POD38. The RA thrombosis was decreased (Fig. 6).

Two months after BML operation, she experienced recurring episodes of significant vaginal bleeding. Subsequently, a total hysterectomy with bilateral salpingo-oophorectomy was conducted. Currently, she has been on NOAC, and a recent

follow-up CT angiography revealed an improvement in thrombosis (Figs. 7, 8).

Discussion

Leiomyomas are generally benign smooth muscle tumors with a very low malignancy potential of about 0.1% [1]. BML was first described by Steiner in 1939 [2]. It is a rare tumor, with < 200 cases documented in the literature, and it occurs in the pelvic cavity, lungs, and lymph nodes, but rarely in the vertebrae or skull base [3]. Many cases are identified as metastatic uterine leiomyoma, and estrogen and progesterone receptors are often positive. BML is histologically benign but often includes multiple metastases accompanied by other hematogenous metastases.

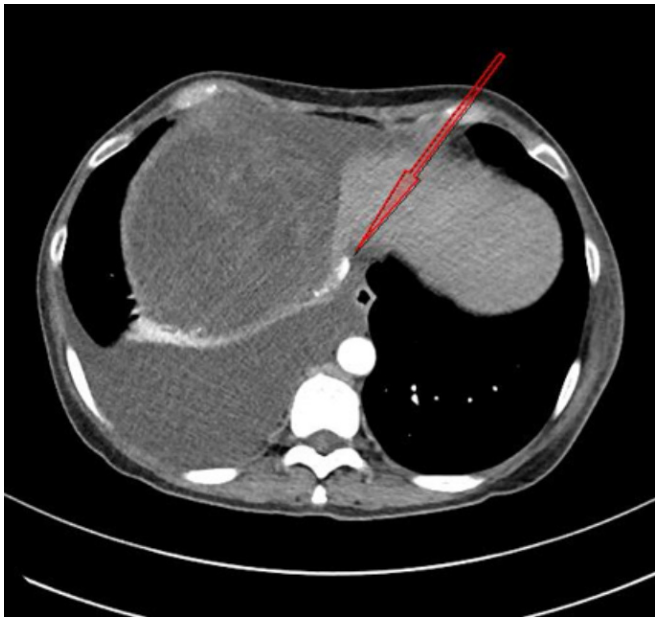

Fig. 2. A chest computed tomography image revealing huge mass in subphrenic area with obliteration of inferior vena cava (arrow).

Some cases of BML occur due to peritoneal seeding after myomectomy or hysterectomy for uterine leiomyoma. BML has been diagnosed in female who have not undergone a previous uterine myoma surgery [4-7]. Metaplastic transformation of the coelomic epithelium explains BML in almost any location where mesothelial mesenchyme exists [8].

Our patient had a history of uterine leiomyoma, but it was not detected using the morcellation method, so the cause was unclear. The patient did not have multiple metastases but had severe thrombosis (SVC-RA) due to the large tumor.

The huge mass pressed IVC, reducing the venous return to RA. So venous congestion has increased not only RA but also SVC-IJV, then the patient developed large thrombosis (SVC-RA).

In order to improve the patient RA function, consider the use of a preoperative antithrombotic agent. After 1 week using LMWH, the patient's thrombosis was improved. After surgery, mass effect was disappeared and IVC obliteration was improved. The need for NOACs for thrombosis decreased after the operation.

A standard treatment for BML has not been firmly established. Due to the hormone-sensitive nature of BML, treatments involve hormonal manipulation through either surgical or medical oophorectomy. Furthermore, regression of metastatic lesions has been observed when estrogen levels significantly decrease, particularly after pregnancy termination and menopause [9,10].

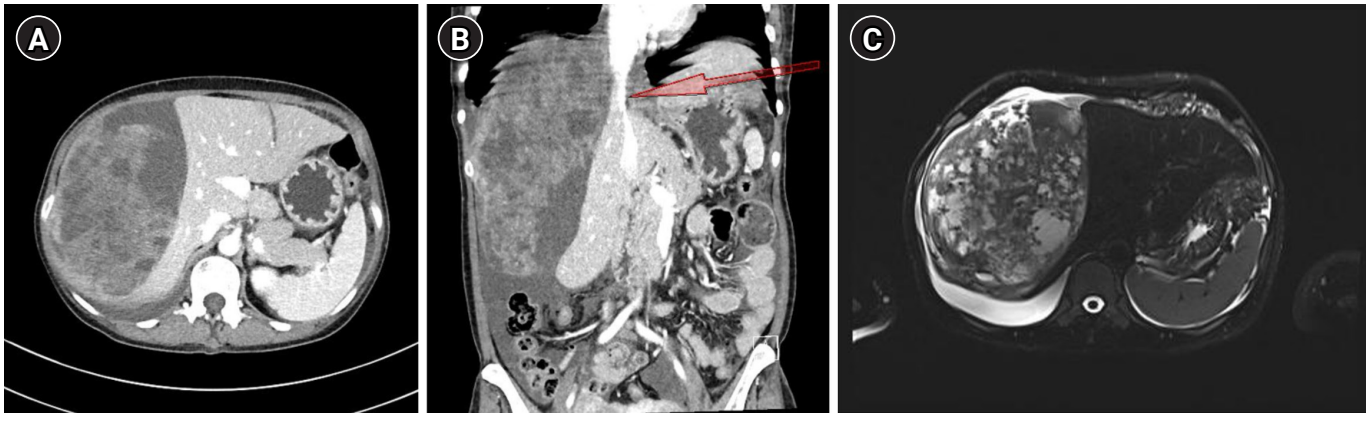

Fig. 3. A liver computed tomography (CT) and liver magnetic resonance (MR) image revealing huge mass in subphrenic area with obliteration of inferior vena cava (IVC). (A) Liver CT image. (B) Liver CT axial image show obliteration of IVC (arrow). (C) Liver MR T2 image show heterogeneous bulky mass in perihepatic space.

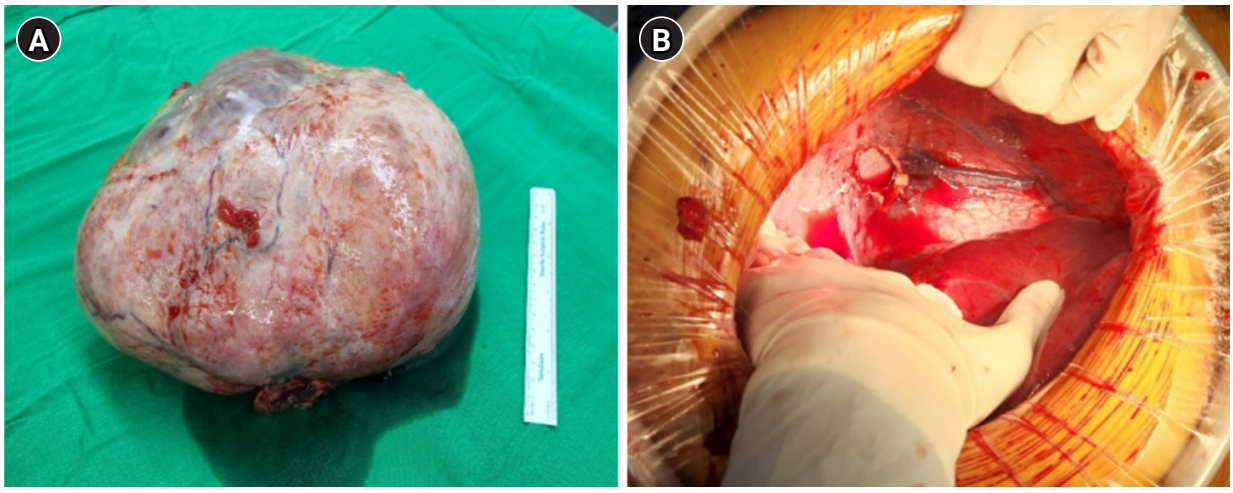

Fig. 4. (A) A 23 cm ×19 cm × 8 cm size subphrenic tumor. (B) A 4 cm × 4 cm size diaphragm defect.

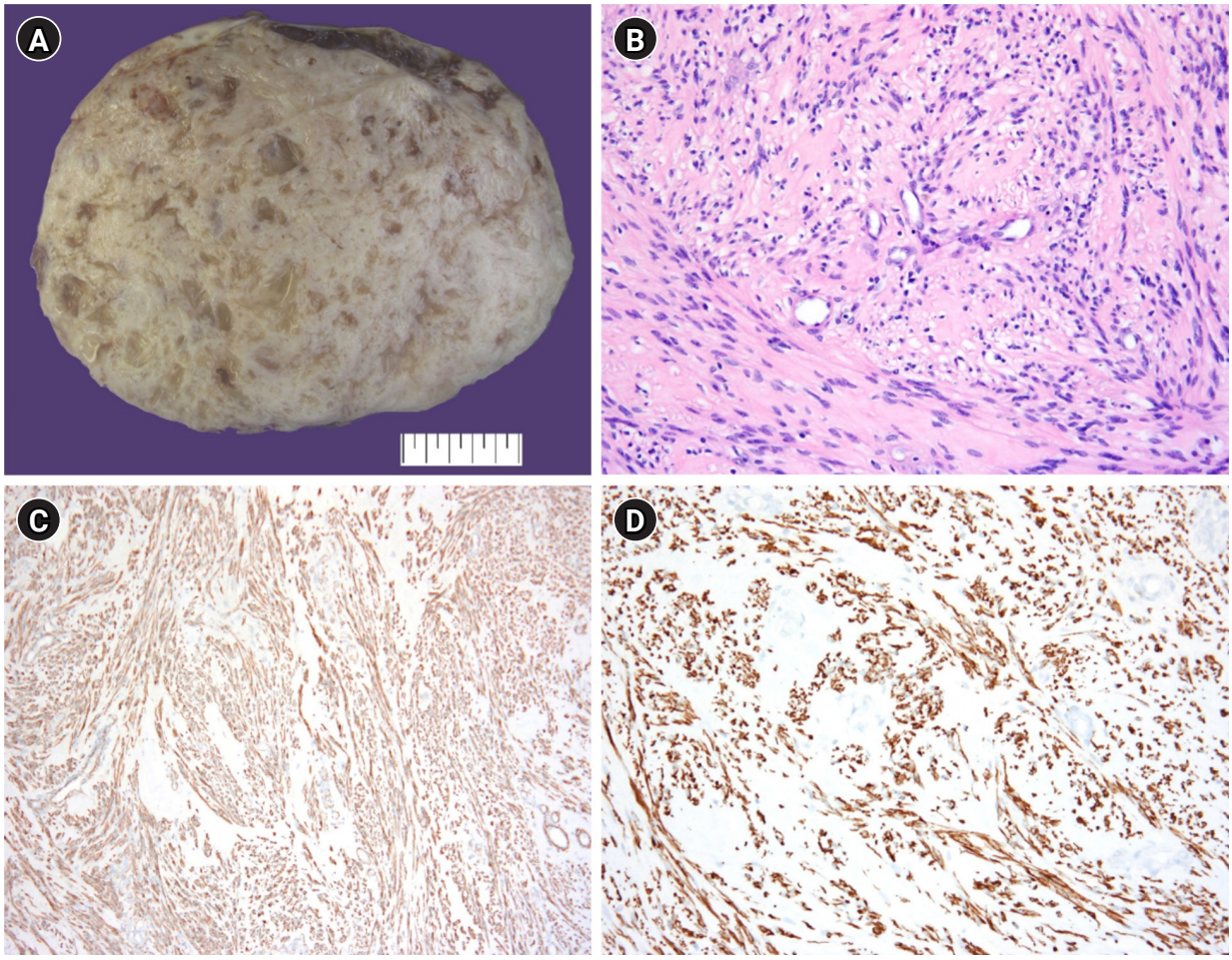

Fig. 5. (A) The tumor was well circumscribed and showed gray-white cut surface with myxoid change in gross examination. (B) The tumor was composed of short spindle cells without atypia (H&E, 200×). (C) Positive for smooth muscle actin (200×). (D) Positive for desmin (200×).

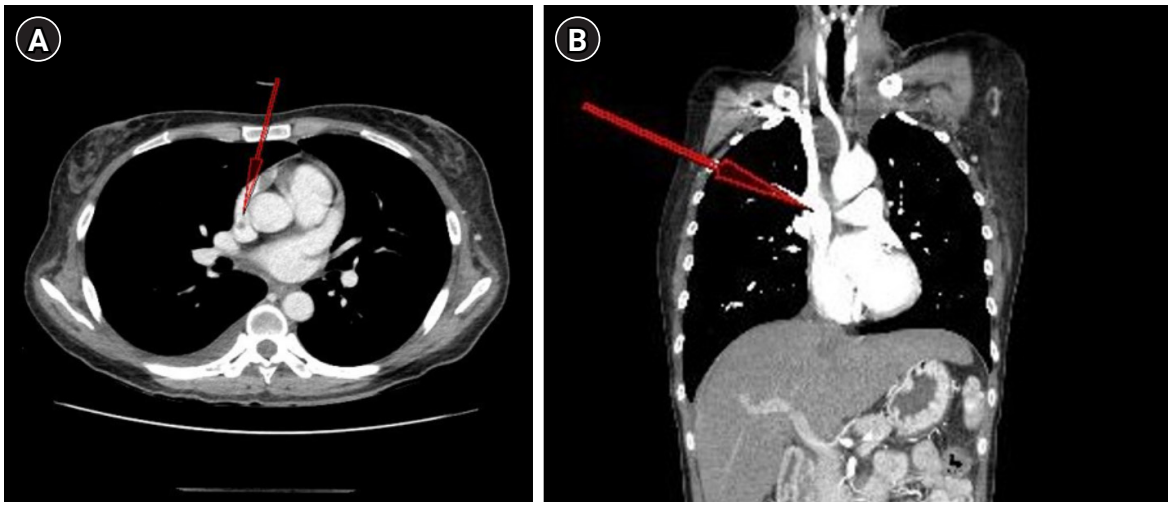

Fig. 6. A Angio computed tomography image revealing decreased volume of extensive venous thrombus in left brachiocephalic vein to right Atrium. (A) Decreased right atrial thrombosis, (B) improved luminal narrowing of superior vena cava.

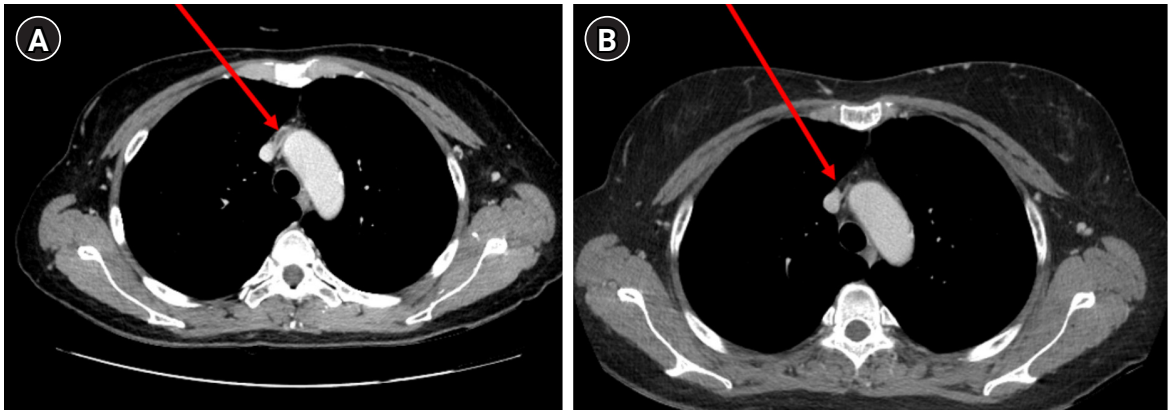

Fig. 7. A Angio computed tomography image revealing decreased volume of extensive venous thrombus in superior vena cava (SVC). (A) Decreased SVC thrombosis (after 4 months), (B) Absence SVC thrombosis (after 1 year).

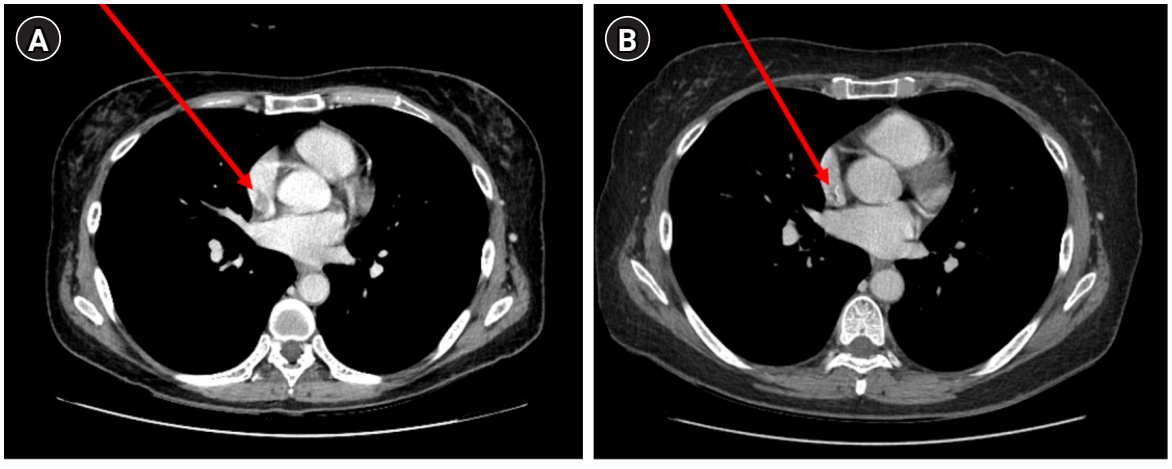

Fig. 8. A Angio computed tomography image revealing decreased volume of extensive venous thrombus in right Atrium. (A) Decreased right atrial (RA) thrombosis (after 4 months), (B) more decreased RA thrombosis (after 1 year).

Reversible medical castration using GnRH agonists, which suppress endogenous gonadotropin secretions required for gonadal steroid production, has shown good therapeutic outcomes in several reports [11,12]. However, the choice of treatment may vary based on the metastatic site of BML, and complications can also influence the prognosis, playing a crucial role in treatment decisions. Complete resection is the priority to treat complication combined BML, and additional hormonal therapy or surgical oophorectomy must be considered. It is also necessary to treat the combined disease.

There were some cases about BML. Our case report very huge BML, so mass effect such as IVC obliteration and severe complication was combined. However, our case is meaningful in the removal of a large BML, and with additional conservative treatment, clinical progress has improved, and complications have also decreased.

Although the pathophysiology of BML is unclear, it can be accompanied by severe complications, which require additional research.

In conclusion, BML is a very rare condition that primarily affects premenopausal female. The metastatic sites are diverse, resulting in various clinical features and timing of discovery, and treatments vary. However, hormone therapy and surgical resection are the most effective treatments and concomitant complications are confirmed to improve survival after removing the tumor.

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Ethics approval

This study was approved by the IRB of Busan Paik Hospital (IRB No. 2023-09-067). Patient consent: waived due to retrospective study.

Conflict of interest

The author has nothing to disclose.

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