Intracardiac leiomyomatosis: a comprehensive analysis of 194 cases

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Abstract

Intracardiac leiomyomatosis is rare but has been increasingly reported in recent years. Owing to its rarity, intracardiac leiomyomatosis has been reported only as isolated case reports and case series. This disorder is thought to be underestimated and easily overlooked in the clinic, while it is dangerous owing to the risk of sudden death caused by total outflow tract obstruction. We performed an electronic literature search for intracardiac leiomyomatosis and identified 194 cases that were reported in English from 1974 (the first reported case) to September 2012. Our aim is to provide a detailed and comprehensive review of the clinical presentation, diagnosis, histopathological characterization, treatment and prognosis of this disorder. According to our analysis, intracardiac leiomyomatosis is most common in the fifth decade, and the mean age of detection is ~50 years. Most patients had undergone previous hysterectomy/myomectomy or had a coexisting uterine leiomyoma when admitted. The most common clinical presentations were dyspnoea, syncope, oedema of the lower extremities and palpitation. Transoesophageal echocardiography, computed tomography and magnetic resonance imaging are helpful in the preoperative diagnosis and to guide the surgical management. Complete removal guarantees an excellent outcome, with no recurrence or postoperative death, while incomplete removal leads to recurrence in one-third of patients. Anti-oestrogen therapy is not imperative after incomplete removal owing to its inability to prevent recurrence.

Keywords: Intracardiac leiomyomatosis • Secondary cardiac tumours • Metastasectomy

INTRODUCTION

Metastatic cardiac tumours are relatively common; however, intravenous tumour extension into the heart is very infrequent. Intracardiac leiomyomatosis (ICLM), a condition rarely seen, is characterized by histologically benign smooth muscle tumours of extracardiac origin reaching the right heart by direct extension into the venous channel. Owing to its rarity, the accumulated knowledge of ICLM has derived mainly from isolated case reports and case series. This tumour has been suggested to originate either from a uterine leiomyoma with vascular invasion or from the venous smooth muscle wall itself. When extending beyond the inferior vena cava (IVC), this intravenous tumour can reach as far as the right atrium (RA), the right ventricle (RV) or even the pulmonary arteries (PAs). Most patients are middle-aged women. Similar to other intracardiac tumours, the clinical presentation of ICLM is non-specific. The advent of echocardiography, computed tomography (CT) and magnetic resonance imaging (MRI) has allowed early and accurate evaluation of this lesion, while the final diagnosis must be made by postoperative pathological examination. Although histologically benign, ICLM is thought to be clinically aggressive owing to the risk of sudden death caused by total outflow tract obstruction.

MATERIALS AND METHODS

We performed an electronic literature search for ICLM in MEDLINE/PubMed, Web of Science and Cochrane Library. The search terms ‘intravenous leiomyoma’, ‘intravenous leiomyomatosis’, ‘intracardiac leiomyoma’, ‘intracardiac leiomyomatosis’, ‘intravascular leiomyoma’, ‘intravascular leiomyomatosis’ ‘cardiac extension’, ‘heart extension’ and ‘pulmonary extension’ were used to identify case reports and case series published in English. Cases with intravascular mass extension into the right heart and confirmed by histopathological examination as leiomyoma were included. All the references cited in each publication were evaluated for additional eligible cases.

We found 163 articles with cases that met the characterization of ICLM (see Supplementary material). Of these, any patient reported more than once in different articles was treated as one case, and the medical information was integrated into the analysis. Finally, we identified 194 cases of ICLM that were reported in English up to September 2012.

Adhesion was defined as the attachment of tumour to the vascular wall, and it may lead to blood vessel rupture or even fatal haemorrhage when the tumour is removed. Complete removal was confirmed by the surgeon during the operation or by postoperative imaging examination. Unless otherwise specified, incomplete removal mentioned here includes partial excision of the tumour and non-surgical treatment (in patients who refused surgery), because both of these left residual tumour in the patient. Recurrence was defined as the postoperative regrowth of the tumour detected by imaging examination. The recurrent and survival data were analysed with the standard Kaplan–Meier actuarial technique. A two-tailed probability value of <0.01 was considered to indicate statistical significance. All the data in our
analysis, including demographics, clinical presentation, histopathological characterization, treatment and prognosis, were extracted from these 194 cases using Excel 2007 (Microsoft Corp., Redmond, WA, USA) and Prism 5.0 (Graphpad Software, La Jolla, CA, USA).

RESULTS

According to our collection of 194 patients, ICLM is seen exclusively in women. Recently, this disorder has been reported with increasing frequency, but it is certain that cases still go unreported. Half of the cases were reported within the last 10 years. Age prevalence was accurately available in 187 patients (Fig. 1), giving an average 47.6 years (range: 20–81 years). Reproductive history was not mentioned in 154 patients (79.4%), and of the remainder, 10 of 40 patients (25.0%) were nulliparous and 30 of 40 (75.0%) were multiparous. One hundred and four patients (53.6%) had undergone a previous hysterectomy/myomectomy, and 58 (29.9%) had a coexisting uterine leiomyoma on admission. Only four patients (2.1%) were reported with a normal uterus. The time between the hysterectomy/myomectomy and detection of ICLM was available for 84 patients, giving an average of 87.8 months (range: 3 months to 47 years).

The clinical presentation of ICLM varied (n = 177 patients) and is shown in Table 1. On auscultation, a heart murmur was heard in 65 patients (33.5%; 54 systolic, six diastolic and five unidentified). Tumour plop was noted in 11 patients (5.7%) and pericardial rub in only two (1.0%).

Chest roentgenogram was mentioned for 51 patients, and the results were normal in 30 (58.8%) and showed heart enlargement in 21 (41.2%). Electrocardiographic results, which were available in 54 patients, were normal in 22 (40.7%), and showed a low voltage in 13 (24.1%), non-specific ST-T change in 11 (20.4%), sinus tachycardia in seven (13.0%) and arrhythmia in four (7.4%).

In 148 cases, the route of tumour extension was described and is shown in Table 1. Distant locations were recorded in 172 patients, with extension into the RA, RV and PAs (Table 1). The adhesion situation was available in 76 patients; 21 (27.6%) did not have any adhesions, while adhesion at the lower diaphragm (1.3%) had a small area of attachment at the level of the diaphragm.

Of 105 patients for whom a preoperative diagnosis was made, ICLM was correctly diagnosed in 57 (54.3%), while 31 (29.5%) were misdiagnosed as RA myxomas, followed by thrombus-in-transit in four (3.8%), pulmonary embolism in three (2.9%) and IVC embolism in two (1.9%).

Treatment of 11 (5.7%) patients was not provided in detail. Apart for four patients (2.1%) who died before surgery and four (2.1%) who refused surgery, surgical excision was performed in 175 patients (90.2%). The surgical procedures are summarized in Table 2. In addition to surgical excision of the tumour, tricuspid valve replacement was performed in seven patients (4.0%) and repair in six (3.4%). The duration of postoperative hospital stay was available for 55 patients, with an average of 10.6 days (range: 4–33 days).

The follow-up time was provided for 123 patients, with an average of 27.3 months (range: 2 months to 12 years). Follow-up of complete removal was recorded for 79 patients (75 of them with follow-up time provided), and no recurrence was reported. The longest postoperative follow-up was 12 years. Follow-up of incomplete removal was recorded for 36 patients and, of these, recurrence occurred in 12 (33.3%; Fig. 2; log-rank P value < 0.0001) and death in four (11.1%; including one patient who died 3 days after surgery, Fig. 3; log-rank P value = 0.0022). The intracardiac location of recurrence was recorded for five patients (two re-extension into the RA, two into the RV and one into the PAs). Two of them underwent reoperation. Among the 36 patients with incomplete removal, out of those who had anti-oestrogen therapy (13), four had a reported recurrence, whereas out of those without anti-oestrogen therapy (23), eight had a recurrence (Fig. 4; log rank P value = 0.8259).

Table 1: Summary of clinical details of intracardiac leiomyomatosis

<table>
<thead>
<tr>
<th>Clinical data</th>
<th>No. of patients (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Presenting symptoms (n = 177 patients)</td>
<td></td>
</tr>
<tr>
<td>Dyspnoea</td>
<td>65 (36.7)</td>
</tr>
<tr>
<td>Syncope</td>
<td>47 (26.6)</td>
</tr>
<tr>
<td>Oedema of the lower extremities</td>
<td>46 (26.0)</td>
</tr>
<tr>
<td>Palpitation</td>
<td>36 (20.3)</td>
</tr>
<tr>
<td>Fatigue</td>
<td>19 (10.7)</td>
</tr>
<tr>
<td>Ascites</td>
<td>16 (9.0)</td>
</tr>
<tr>
<td>Jugular vein distension</td>
<td>13 (7.3)</td>
</tr>
<tr>
<td>Chest pain</td>
<td>13 (7.3)</td>
</tr>
<tr>
<td>Abdominal pain</td>
<td>12 (6.8)</td>
</tr>
<tr>
<td>Asymptomatic</td>
<td>23 (13.0)</td>
</tr>
<tr>
<td>Route of extension (n = 148 patients)</td>
<td></td>
</tr>
<tr>
<td>Iliac vein</td>
<td>98 (66.2)</td>
</tr>
<tr>
<td>Ovarian vein</td>
<td>26 (17.6)</td>
</tr>
<tr>
<td>Renal vein</td>
<td>21 (14.2)</td>
</tr>
<tr>
<td>Both iliac and ovarian veins</td>
<td>10 (6.8)</td>
</tr>
<tr>
<td>Gonadal vein</td>
<td>6 (4.1)</td>
</tr>
<tr>
<td>Hypogastric vein</td>
<td>5 (3.4)</td>
</tr>
<tr>
<td>Uterine vein</td>
<td>5 (3.4)</td>
</tr>
<tr>
<td>Hypohepatic or hepatic vein</td>
<td>5 (3.4)</td>
</tr>
<tr>
<td>Pelvic vein</td>
<td>2 (1.4)</td>
</tr>
<tr>
<td>Extent of involvement (n = 172 patients)</td>
<td></td>
</tr>
<tr>
<td>Right atrium</td>
<td>69 (40.1)</td>
</tr>
<tr>
<td>Right ventricle</td>
<td>81 (47.1)</td>
</tr>
<tr>
<td>Pulmonary artery</td>
<td>22 (12.8)</td>
</tr>
</tbody>
</table>
DISCUSSION

Historical perspective

In 1907, the initial cases of ICLM were described in German by Dürrck and Hörmann, respectively [1, 2]. The first report of ICLM in English was by Mandelbaum et al. in 1974 and originated from the IVC [3]. In 1980, Timmis et al. noted the first case of ICLM originating from the uterus [4] and, in the same year, anti-oestrogen therapy was first used by Tierney et al. to prevent possible recurrence due to incomplete excision [5]. In 1982, Maurer and Nanda used the term ‘intracardiac leiomyomatosis’ and diagnosed this disease on the basis of transthoracic echocardiography (TTE) for the first time [6]. The first complete removal of an ICLM using a one-stage procedure was reported by Iverson et al. in 1983 [7] and using a two-stage procedure by Basso et al. in 1984 [8]. In 1989, Torres et al. were the first to use transoesophageal echocardiography (TOE) for preoperative evaluation of the intracardiac portion of an ICLM [9]. In 1998, Marom et al. reported this lesion in a pregnant woman [10]. Kullo et al., in 1999, provided the first description of recurrent ICLM due to incomplete excision [11]. In 2002, Quade et al. performed cytogenetic and molecular studies of ICLM [12]. The longest ICLM (75 cm) was reported by Lam et al. in 2004 [13]. In 2006, Jeanmart et al. reported the first successful complete excision via thoracoscopy alone [14]. More recently, Okada et al. reported an older patient (81 years old) who tolerated the one-stage complete removal procedure well in 2012 [15].

Epidemiology and characterization

Although ICLM occurs at ages ranging from 20 to 81 years, it is most common in the fifth decade (Fig. 1). The mean age of detection is ~50 years. In our collection of cases, this disorder is seen exclusively in women. Ten patients were nulliparous, while 30 were multiparous. Given that the reproductive history was not mentioned in most cases (79.4%), its relation to this disorder cannot be specified. It is postulated that ICLM arises from an invasive uterine myoma or from the vascular wall [8, 16, 17].
Approximately 83.5% of the patients had undergone a previous hysterectomy/myomectomy or had a coexisting uterine leiomyoma, suggesting uterine leiomyoma with vascular invasion as the origin of this lesion. A few patients were found with a normal uterus [3, 18–20], which may support the theory of ICLM originating from the venous smooth muscle wall itself. The interval between hysterectomy/myomectomy and the detection of ICLM varied from merely 3 months [21] to as long as more than 40 years [22, 23]. Furthermore, two cases of pregnant women with ICLM have been described [10, 24].

Clinical manifestations

Even with intravenous tumour extension as far as the IVC, symptoms may be absent until cardiac insufficiency develops due to intracardiac extension [4, 25–27]. The clinical presentation of ICLM usually depends on the extension and size of the tumour [3, 28–30]. Right ventricular extension may cause complete outflow tract obstruction [31, 32], and patients with extensions as far as the PAs may develop severe pulmonary embolism [28, 33, 34] or even sudden death [35]. Asymptomatic manifestations were reported in 13.0% of ICLM patients because of little or no compromise of venous return. Symptomatic patients presented with various symptoms, the most common being dyspnoea, syncope, oedema of the lower extremities and palpitations, followed by fatigue, ascites, jugular vein distension, chest pain, abdominal pain and hepatomegaly. In a few cases, other manifestations, such as dizziness [19, 33, 36–40], cyanosis [31, 41–43], cough [42, 44], vomiting [24, 43, 45], chest tightness [27, 33, 46], menstrual disorder [32, 46–48], orthopnoea [39, 49, 50], lethargy [51–53], headache [11, 54, 55], sweating [24, 43, 55], postmenopausal bleeding [56–58], weight loss [56, 59], shock [42, 60], nausea [43, 61], hypotension [24, 42], skin petechiae [31, 62], haemoptysis [63, 64], scleral icterus [62], facial oedema [65] and urinary incontinence [53], have been reported.

Although seldom seen, concomitant embolic phenomena in the distal vasculature occurred in a few patients. As an unusual clinical presentation, acute Budd–Chiari syndrome due to thrombotic occlusion of large hepatic veins has been reported [22, 56]. Another patient with embolism in an artery of the left parietal lobe was admitted with sudden onset of right hemiparesis and slurring of speech [66]. In addition, a case of ICLM with severe hypoxia culminating in generalized seizure activity in a pregnant woman has been described [24].

Depending on the size and mobility of the tumour, ICLM with RV extension (including intermittent prolapse into the RV) through the tricuspid orifice may damage the valve apparatus, leading to tricuspid insufficiency or stenosis. A systolic or diastolic murmur was noted in 33.5% of patients. In 5.7% of the reported patients, tumour plop was heard when the tumour was usually mobile in the flow of blood. Electrocardiographic findings showed normal results in 40.7% of patients, while low-voltage, non-specific ST–T change, sinus tachycardia and arrhythmia have also been reported.

Imaging examination

Imaging examination is helpful to the preoperative diagnosis and detection of recurrence of ICLM [37, 66, 67]. The chest roentgenogram, which was mentioned for 51 patients, presented non-specific cardiomegaly in 58.8% of these and was not helpful for the detection of the tumour unless there was extensive calcification [6, 68].

Echocardiography is a more convenient method and can provide real-time information on an intracardiac tumour [69]. Currently, TTE is widely used, but the visualization of an IVC tumour may be unsuccessful owing to a pattern of echo-free space caused by blood in a tubular tumour [8, 53]. Transoesophageal echocardiography is superior to TTE in evaluation of a right atrial mass by providing more accurate images [9, 11, 21, 26, 55]. Transoesophageal echocardiography has been used to show the fine details of the IVC, interatrial septum, tricuspid valve apparatus and attachments of the tumour [9, 26, 36]. To avoid vein injury during tumour removal, intraoperative TOE monitoring was suggested to guide the surgeons on localization and adherence [36, 39, 58].

Enhanced CT and MRI are usually used to evaluate the size, location, origin and extent of an ICLM, because they can provide excellent anatomical information [28, 47, 49, 66]. Magnetic resonance imaging usually shows a mass of isosignal intensity on T1 weighting, and a heterogeneous and high-intensity signal in some regions on T2 weighting, due to cystic or hyaline degeneration [27, 47]. However, a patient with an ICLM that showed a mildly enhanced signal in both T1 and T2 weighting has been described [65]. Motion artifacts may be the main disadvantage of MRI. Cardiac catheterization is not usually required, because most of the time adequate information is provided by non-invasive imaging. Whole-body scans with radioactive elements can be used to exclude residual tumours in other locations.

Generally speaking, it is a challenge to make a correct diagnosis of ICLM before surgery, and its occurrence is considered to be underestimated [59], but with advances in imaging technologies, this situation has substantially improved.

Differential diagnosis

Differential diagnosis primarily encompasses right atrial myxoma, thrombus-in-transit, renal cell carcinoma, Wilms’ tumour, intracardiac leiomyosarcomatosis, benign metastasizing leiomyoma and other metastatic cardiac tumours. Almost 30% of the patients were misdiagnosed preoperatively as having RA myxomas. In contrast to the right-sided characterization of ICLM, myxoma has a left-sided predominance, with an incidence between 87.3 and 88.5% [70–72]. Although most left atrial masses are myxomas, they constitute only 43% of right atrial masses [18]. Most myxomas originate from the interatrial septum with a stalk and may attach to the walls of the cardiac chambers [36, 65, 69]. Renal cell carcinoma, which should be distinguished from ICLM by postoperative pathological examination, is the most common tumour that can extend into the IVC and right heart [49, 69]. A history of previous hysterectomy/myomectomy or a coexisting uterine leiomyoma may be helpful. Although, in rare instances, Wilms’ tumour with intracardiac extension in adult has been reported, it is commonly seen in children [21]. Renal cell carcinoma and Wilms’ tumour can also be distinguished by detection of renal tumour on imaging examination. Thrombus-in-transit, which typically occurs postoperatively and frequently leads to sudden death, usually gives a ‘popcorn’ appearance within the cardiac chambers [11, 42, 67].
Intracardiac leiomyosarcomatosis, a malignant type in the smooth muscle tumour spectrum, often shares clinical presentations with ICLM, depending on the size and site [49], and should be differentiated by histopathological examination and immunohistochemical staining [52, 62, 65]. Benign metastasizing leiomyoma generally originates from the uterus and metastasizes to other organs, such as the lung, lymph nodes, bone, skeletal muscle, subcutaneous tissue or retroperitoneal space. Although ICLM is rarely coincident with benign metastasizing leiomyoma in the lung [11, 16, 57], the latter alone usually shows no occupation of vascular channels. Other metastatic cardiac tumours commonly involve the pericardium/myocardium, and the primary nodule can be detected by whole-body positron emission computed tomography.

**Treatment**

Considering the risk of sudden death caused by total outflow tract obstruction, early treatment is recommended when ICLM is highly suspected [75]. Surgical removal of the ICLM is curative, and many surgical techniques have been applied in the treatment of this disease. A quaternary classification system has been described to choose the best surgical strategy for different patients [63].

Complete removal is strongly recommended, because no recurrence has been reported. Blind removal of the tumour via sternotomy or laparotomy alone is dangerous, because this may lead to incomplete retrieval or fatal retroperitoneal haemorrhage [31, 56].

A double approach, using a one- or two-stage procedure, is performed more commonly and thought to be safer. The two-stage procedure involves separate removal of the intracardiac and intra-abdominal components at intervals varying from 7 days to 2 years. Although the patient reported by Altinok and intra-abdominal components at intervals varying from 7 days to 2 years. Although the patient reported by Altinok [47] was followed up every 3–6 months, an interval as long as 2 years should be avoided. The advantages of this procedure are as follows [39, 46]: (i) suitability for patients in poor condition who cannot tolerate a one-stage procedure; and (ii) it is recommended by some authors when the tumour is extensive or adheres tightly to the vascular wall. The interval can be decided according to the recovery status of the patient [46]; however, rapid growth leading to re-extension into the right cardiac chamber during the waiting interval has been reported [37, 47]. When the patient presents as an emergency [27, 39], a separate surgery is mandatory, with sternotomy first.

More recently, a one-stage operation, in which the sternotomy and laparotomy are performed simultaneously, is being increasingly reported. This approach needs the co-operation of a combined surgical team. The one-stage procedure has several advantages, as follows [40, 73]: (i) avoidance of haemodynamic complications during the interval between two stages; (ii) avoidance of the risk of a second general anesthetic; (iii) reduction of the risk of tumour embolism caused by incomplete resection in the first stage; and (iv) better economic benefits. Whether the risk of bleeding increases is still disputed, because the operative time is not excessively prolonged. Most one-stage operations are performed with cardiopulmonary bypass and total circulatory arrest; in only three cases was the on-pump beating-heart technique used when removing the intracardiac mass [74–76].

In 2011, a woman with an ICLM originating from the uterus was admitted to our department. The tumour was completely removed in a one-stage procedure by co-operation of cardiac surgeons, vascular surgeons and gynaecologists. The patient was asymptomatic, with no evidence of recurrence on 13 month follow-up. Our experience also confirmed that, if there is no adhesion to the IVC or heart at any point, a one-stage complete resection without cardiac arrest is feasible and safe, and leads to a satisfactory outcome [77].

Intraoperative TOE monitoring is used to guide the surgeons about the localization and adherence, to avoid vein injury [36, 39, 58]. Surgery with endovascular techniques has been successful in preventing distal embolization [37]. Furthermore, with the advent of minimally invasive techniques, laparoscopic excision of an abdominal tumour [40] and successful complete excision via thoracoscope alone [14] have been reported.

When the tricuspid valve apparatus is injured by the tumour, valve replacement or repair may be required. A total hysterectomy is recommended if ICLM is confirmed as originating from a coexisting uterine leiomyoma. Bilateral salpingo-oophorectomy should also be performed because of the oestrogen-dependent characteristic of the tumour. To prevent recurrence, ligation of the originating vein is imperative [52].

Whatever the surgical approach chosen, the best choice of treatment is complete removal. Neither medication (including anti-oestrogen therapy using tamoxifen or letrozole) nor radiotherapy alone has been reported to provide a satisfactory outcome. Long-term follow-up should be arranged in case of recurrence.

Incomplete removal is not recommended, because recurrence occurred in 12 patients (33.3%). When the resection is incomplete because of technical difficulty or the patient refuses further surgery, anti-oestrogen therapy (using tamoxifen or letrozole) has been suggested by some authors [21, 27, 33, 50]. Although the oestrogen-dependent characteristic of ICLM is widely recognized, our analysis showed that postoperative anti-oestrogen therapy does not help to prevent recurrence. The possible explanation seems to be that some ICLMs are not oestrogen dependent, but this supposition cannot be confirmed because the oestrogen level in the tumour was not available for most patients with recurrence (10 of 12). The side-effects of this medical treatment include associated menopause, which leads to osteoporosis and heart disease [24]. We conclude that anti-oestrogen therapy is not imperative after incomplete removal. We acknowledge that when dealing with case reports as the source of data, publication bias evidently favours cases with unique features. Moreover, follow-up of incomplete removal was provided in only 36 patients in our analysis, and no randomized controlled data are available, so this recommendation for postoperative anti-oestrogen therapy is still controversial and needs to be assessed further.

**Pathology and pathogenesis**

Macroscopically, ICLM originating from the uterus usually has a serpentine appearance, with iliac or ovarian vein extension into the IVC and RA. In contrast, a few patients with ICLM originating from the venous smooth muscle wall itself have been reported [3, 17–20, 28]. The tumour is generally smooth and rubbery, with a greishy-white or rusty colour, with or without an intumescent intracardiac head [21]. In patients with a long course of this disease, the tumour may be calcified [6, 19, 28, 32, 68] and extend more than 50 cm [13].

Intracardiac leiomyomatosis is histologically benign. Microscopic examination of atrial tumours often shows a bundled arrangement of spindle-shaped cells and fibromuscular tissues with
therapy is not imperative after incomplete removal, owing to its leads to recurrence in one-third of patients. Anti-oestrogen occurrence or postoperative death, while incomplete removal Complete removal guarantees an excellent outcome, with no recurrence or postoperative death. Recurrence after incomplete removal was found in 33.3%, and four patients died during follow-up.

Prognosis

Four patients died before surgery due to heart failure [32, 56], septic shock [22] and pulmonary embolism [35]. Intraoperative death occurred in three patients [31, 47, 79], two of whom died from an IVC tear [31, 79]. Whether a one- or two-stage procedure is chosen, the short- and long-term outcome of complete removal is excellent, with no recurrence or postoperative death reported. Recurrence after incomplete removal was found in 33.3%, and four patients died during follow-up [20, 63, 80].

CONCLUSIONS

Although ICLM is rare and always occurs sporadically, it is more common than previously thought. It is most common in the fifth decade, and most patients had undergone a previous hysterectomy/myomectomy or had a coexisting uterine leiomyoma. When a right atrial mass is identified in middle-aged women with previous hysterectomy/myomectomy or uterine leiomyoma, a high degree of suspicion should be maintained, and an abdominal CT should be arranged to confirm the origin. Complete removal guarantees an excellent outcome, with no recurrence or postoperative death, while incomplete removal leads to recurrence in one-third of patients. Anti-oestrogen therapy is not imperative after incomplete removal, owing to its inability to prevent recurrence.

SUPPLEMENTARY MATERIAL

Supplementary material is available at ICVTS online.

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REFERENCES

Intravenous leiomyomatosis (IVL) is a rare benign neoplasm characterized by leiomyomatous proliferation extending from the uterus through the uterine veins into the inferior vena cava (IVC) and right heart chambers. It is predominantly seen in women of reproductive age, with a mean age of diagnosis around 40 years. IVL often presents with nonspecific symptoms such as dyspnea, chest pain, and right heart failure. The disease can be classified into intravenous (IV) and intracardiac (IC) forms. 

IVL is typically diagnosed with imaging modalities such as magnetic resonance imaging (MRI) and computed tomography (CT). MRI is preferred due to its high soft tissue contrast, enabling better visualization of the tumor's extent and involvement of the right heart chambers. Cardiac MRI is particularly useful in assessing the extent of the lesion and its impact on cardiac function. 

Intracardiac IVL can present with various clinical features, including arrhythmias, heart failure, and sudden death. The diagnosis is often made during surgical exploration or at autopsy. The treatment approach varies depending on the extent and clinical presentation. In some cases, IVL is managed conservatively with close monitoring and follow-up imaging. For larger lesions or symptomatic patients, surgical resection may be considered. 

Surgical resection is the definitive treatment for IVL. In some cases, a two-stage approach is necessary to achieve complete resection without compromising cardiac function. The surgical strategy should be individualized based on the tumor's anatomical location, size, and involvement of the right heart chambers. Postoperative management includes close cardiac monitoring and follow-up imaging to assess for tumor recurrence. 

Overall, IVL is a rare condition with a favorable prognosis. With advances in imaging and surgical techniques, the outcomes for patients with IVL have improved significantly. Further research is needed to better understand the pathophysiology and optimize management strategies for this uncommon disease.