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The Classification and Surgical Strategy of Intracardiac Leiomyomatosis

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BACKGROUND: There is a great deal of heterogeneity in the surgical strategy to treat intracardiac leiomyomatosis (ICL), leading to a need to create a theoretical tool to clarify this situation.
METHODS: The data of 14 cases of ICL surgically treated in Anzhen Hospital from February 1995 to February 2009 were retrospectively reviewed. A system for classifying ICL was proposed based on four features of the lesion: size of intracardiac component; extent of inferior vena cava (IVC) involvement; venous pathway from uterus to IVC; and laterality of the lesion in the pelvis. The 14 cases of ICL were treated through multiple surgical strategies.
RESULTS: There were no operative deaths. The follow-up was 73.1 ± 59.2 months and one patient died from recurrence due to incomplete excision 5 months after the primary procedure. The 5-year survival rate calculated through Kaplan-Meier survival curve was 93.16 ± 4.98%. Of the surviving patients, 13 had ICL, 10 were in the New York Heart Association (NYHA) class I, and three were in NYHA class II.
CONCLUSION: The surgical treatment of ICL can obtain a good mid- to long-term survival rate and satisfactory heart function, and the proposed classification system for ICL may be helpful to guide the selection of the surgical strategy for ICL, and may serve as the future basis for standardising the reporting of ICL management. [*Asian J Surg* 2009;32(3):129–36]

Key Words: intracardiac leiomyomatosis (ICL)

Introduction

Intravenous leiomyomatosis (IVL) is an uncommon non-malignant tumour, which originates from the smooth muscle cells and is usually confined to the pelvic venous system.[1] When IVL extends into the right side of the heart, it becomes intracardiac leiomyomatosis (ICL). Only about 200 cases of ICL have been described in the literature in the forms of sporadic cases reports. With its cardiac involvement, which may reach deep into the pulmonary artery branches,[2–4] ICL is a rare but severe clinical situation needing to be dealt with and requiring much attention, and the clinical situation is also very complicated.[5] We report a cohort of 14 cases of ICL which were successfully treated through multiple surgical approaches and strategies in the time frame from February 1995 to February 2009. Through analysing the data of these 14 ICLs we developed a classification system, in order to clarify the complicated situation and thus help the pre-operative assessment and the choosing of the surgical strategy according to the anatomy of the tumour. This classification system may act as the basis for standardising the reporting of future ICL management.

Patients and Methods

The Ethics Committee approved this retrospective study and waived the need to obtain patient consent for the

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study. Written informed consent was obtained from each patient for the operation.

Patients characteristics

From February 1995 to February 2009, 14 cases of ICL were operated on with multiple surgical strategies. All the patients were female with an age range from 20 to 62 years (mean, 43.5 ± 9.8). The disease course was 3 months to 2 years. Three patients were in the New York Heart Association (NYHA) functional class IV, 10 patients were in NYHA functional class III, and one was in NYHA functional class II. A computed tomography/magnetic resonance imaging (CT/MRI) scan, preprocedure transthorax echocardiography (TTE) and abdominal type B ultrasonography were adopted to identify the anatomical pathological features of each ICL patient. According to the clinical and anatomic pathological features of ICL, we developed a classification system, which we named "The Anzhen quaternary classification scheme for ICL" (Table 1). Of the 14 ICLs, there were four ICLs with intracardiac and intravenous parts less than the minimum diameter of the inferior vena cava (IVC), and which originated from the left internal iliac vein (NAIL type ICL). Two ICLs had intracardiac and intravenous parts less than the minimum diameter of the IVC, and originated from the right ovarian vein (NAOR type ICL). One ICL had intracardiac parts less than the minimum diameter of the IVC, and originated from the right ovarian vein but where the supra hepatic IVC was blocked by the ICL (NBOL type ICL). One ICL had an intracardiac part which was less than the minimum diameter of the IVC, and originated from the left ovarian vein, but the retro hepatic IVC were blocked by the ICL (NCOL type ICL). One ICL had its intracardiac part less than the minimum diameter of the IVC, and originated from left internal iliac vein, but the infra hepatic IVC were blocked by the ICL (NDIL type ICL). Two ICLs had intracardiac parts greater than the minimum diameter of the IVC, and originated from left internal iliac vein, but there was no blockage in IVC (YAIL type ICL). Two ICL had their intracardiac parts greater than the minimum diameter of the IVC, and originated from right ovarian vein, and the supra hepatic IVC was blocked by the ICL (YBOR type ICL). The final ICL had its intracardiac part greater than the minimum diameter of the IVC, and originated from the left internal iliac vein, and the retro hepatic IVC was blocked by the ICL (YCIL). The patient characteristics are shown in Table 2. The cardio thorax ratio (CTR) ranged from 0.45 to 0.67.

Operative procedure

As shown in Table 3, 14 ICL patients were all placed under general anaesthesia. A cardiopulmonary bypass (CPB) was then established, and crystalloid cardioplegia or cold blood cardioplegia was applied as needed. Of those

Table 1. The Anzhen concepts of quaternary classification for ICL

Grade	Type	Concept	Meaning
1st grade	N	Intracardiac part of ICL has no intumescences	The intracardiac part is less than the minimum diameter of IVC
	Y	Intracardiac part of ICL has intumescences	Yes, the intracardiac part is bigger than the minimum diameter of the IVC
2nd grade	A	ICL have no adherence and blockage to IVC	ICL floats in IVC, and there is no blockage of IVC
	B	Adheres to and blocks supra hepatic IVC	ICL blocks the venous returns in supra hepatic IVC
	C	Adheres to and blocks retro hepatic IVC	ICL blocks the venous return in retro hepatic IVC
	D	Adheres to and blocks infra hepatic IVC	ICL blocks the venous return in the infra hepatic IVC
3rd grade	I	From internal iliac vein	ICL originated from internal iliac vein
	O	From vena ovarica	ICL originated from vena ovarica
4th grade	L	On body left	ICL located on the left side of the body
	R	On body right	ICL located on the right side of the body

Putting the four characters from the 4 grade classification together in sequence, each of which represents one of four-grade classification, we can constitute a quaternary classification scheme for ICL. For example, NAIL specifies that the intracardiac part of the ICL has no intumescences, and the caval part of the ICL does not adhere to or block to the IVC, and the ICL originates from the left internal iliac vein.

Table 2. Patient characteristics of the 14 cases of ICL

S.no	Type	Age	Main symptoms	Preprocedure hysterectomy
1	NAIL	20	Palpitation	
2	NAIL	42	Palpitation	
3	NAIL	39	Palpitation	
4	NAIL	62	None	
5	NAOR	40	Palpitation	
6	NAOR	56	Palpitation	Sub hysterectomy, 3 yrs ago
7	NBOL	44	Ascites, oedema, syncope	
8	NCOL	37	Ascites, oedema, syncope	Hysterectomy, 5 yrs ago
9	NDIL	52	Ascites, oedema, syncope	Hysterectomy, 1 yr ago
10	YAIL	48	Palpitation, haemoptysis	Hysterectomy, 3 yrs ago
11	YAIL	45	Palpitation	Hysterectomy, 2 yrs ago
12	YBOR	39	Palpitation, haemoptysis	
13	YBOR	41	Palpitation, ascites, syncope	Sub hysterectomy, 3 yrs ago
14	YCIL	44	Palpitation, ascites	Hysterectomy, 4 yrs ago

*Oedema = oedema at lower extremities; sub hysterectomy = subtotal hysterectomy. NAIL: ICL with its intracardiac part and the intravenous part less than the minimum diameter of the IVC, and originated from left internal iliac vein; NAOR: ICL with its intracardiac part and the intravenous part less than the minimum diameter of the IVC, and originated from right ovarian vein; NBOL: ICL with its intracardiac part less than the minimum diameter of the IVC, and originated from right ovarian vein, but the supra hepatic IVC were blocked by the ICL; NCOL: ICL with its intracardiac part less than the minimum diameter of the IVC, and originated from left ovarian vein, but the retro hepatic IVC were blocked by the ICL; NDIL: ICL with its intracardiac part less than the minimum diameter of the IVC, and originated from left internal iliac vein, but the infra hepatic IVC were blocked by the ICL; YAIL: ICL with its intracardiac part greater than the minimum diameter of the IVC, and originated from left internal iliac vein, but there is no blockage in IVC; YBOR: ICL with its intracardiac part greater than the minimum diameter of the IVC, and originated from right ovarian vein, and the supra hepatic IVC was blocked by the ICL; YCIL: ICL with its intracardiac part greater than the minimum diameter of the IVC, and originated from left internal iliac vein, the retro hepatic IVC was blocked by the ICL.

Table 3. Fourteen ICL distribution in quaternary classification and choosing of the surgical strategy

S.no	Type	Approach	CPB style	CPB cannulation	Pedicles
1	NAIL	Sternotomy	Moderate hypothermic CPB	SVC & IVC	Pulled
2	NAIL	Sternotomy	Moderate hypothermic CPB	SVC & IVC	Pulled
3	NAIL	Sternotomy	Moderate hypothermic CPB	SVC & IVC	Pulled
4	NAIL	Sternotomy	Moderate hypothermic CPB	SVC & IVC	Pulled
5	NAOR	Sternotomy	Moderate hypothermic CPB	SVC & IVC	Pulled
6	NAOR	Laparotomy	Normothermic CPB	Femoral & Jugu V	Excised
7	NBOL	Sternotomy	DHLF	SVC & IVC	Pulled
8	NCOL	SLtomy	DHLF	SVC & Femoral V	Excised
9	NDIL	SLtomy	DHLF	SVC & Femoral V	Excised
10	YAIL	SLtomy	DHCA	SVC & Femoral V	Excised
11	YAIL	SLtomy	DHLF	SVC & IVC	Excised
12	YBOR	Sternotomy	Moderate hypothermic CPB	SVC & IVC	Pulled
13	YBOR	SLtomy	DHCA	SVC & Femoral V	Excised
14	YCIL	SLtomy	DHLF	SVC & Femoral V	Excised

*SLtomy = sternolaparotomy; CPB = cardiopulmonary bypass; Jugu V = jugular vein; femoral V = femoral vein; DHLF = deep hypothermia and low flow; DHCA = deep hypothermia and circulatory arrest; SVC = superior vena cava; IVC = inferior vena cava.

six patients with the NA type ICL, the whole length of the ICL was pulled directly out from the IVC through the right atrium opening in three patients, and the ICL pedicle was firstly excised and then the whole length of the ICL was taken out through the right atrium incision in two patients. Only the internal iliac vein was dissected through laparotomy, and then the pedicle of the ICL was excised and pulled out through the internal iliac vein

incision in one patient. For the one NBOL type ICL, the IVC could not be cannulated due to the blockage of the ICL intumescences, so deep hypothermia low flow (DHLF) was launched through a single cannula in the superior vena cava (SVC), and the ICL intumescence was just partially excised. For the one NCOL type ICL, a deep hypothermia circulatory arrest (DHCA) was launched after dissecting the retro hepatic IVC fully, and then the whole body of the ICL intumescences and their two ends were dissected out through the incision in the retro hepatic part of IVC. For the one NDIL type ICL, DHLF was launched and the infra hepatic IVC was dissected and incised, and the intumescences of ICL were dissected out with both ends. For the two YAIL type ICLs, the whole body of the ICL was pulled out through right atrium incision. Three patients, with either YBOR or YCIL type ICL, were all treated under the support of DHLF or DHCA, and the intumescences were dissected and pulled out through both incisions in the right atrium and IVC. The internal iliac veins were ligated in two patients. A combined operation was taken to remove the pelvic and intravascular lesions in the same procedure.

Statistical analysis

Categorical data are given as total numbers and relative frequencies. Continuous data are given as mean ± standard deviation (SD). A p value of less than 0.05 was considered statistically significant. A 5-year survival rate is calculated through the Kaplan-Meier survival curve and is expressed by mean ± SD. Statistical analyses were performed using statistical software SPSS 13.0 (SPSS Inc, NUIT, and Evanston, Illinois, USA).

Results

For the whole group, the CPB duration extended from 25 to 146 minutes (mean, 78.2 ± 43.7 minutes), the clamping time extended from 0 to 78 minutes (41.6 ± 37.5 minutes), the DHCA time for the two ICLs were 26 and 31 minutes, and the nasopharyngeal temperature during the CPB was 28.7°C ± 6.9°C. All cardiopulmonary bypasses were discontinued without incident and the retro-operative course was not complicated in any of the patients. All patients recovered well from the surgery. In 14 patients, the pathological findings manifested extensive hard rubbery lesions, most of them being yellowish, polymorphic shaped tumours. Histology demonstrated these to be leiomyoma, and some with grossly dilated IVC suggested a protracted clinical course. The intravascular lesion measured 30 to 48 cm in length and 0.3 to 7.2 cm in width at the broadest point. Positive oestrogen receptors were detected in four cases. There were no operative deaths. One patient had to undergo the procedure a second time because of bleeding. The blood losses were 480.8 ± 211.5 mL and the transfusion requirements were 370.4 ± 184.8 mL. The ICU stay was 22 ± 8.5 hours, and the post-operative length of hospital stay was 11.2 ± 3.6 days.

The patient follow-up was 100% complete. The mean follow-up was 73.1 ± 59.2 months (cumulative, 75.3 patient years; range, 1 to 130 months). All of the follow-ups were completed at the out-patients department using a CT scan and echocardiography. One patient with a YBOR type ICL suffered a recurrence with a full blockage of the IVC in the third month after the primary procedure due to the incomplete excision of the lesion. The patient refused a second procedure and died from a pulmonary embolisation 2 months later. The 5-year survival rate calculated through the Kaplan-Meier survival curve was 93.16% ± 4.98%. Of the surviving 13 patients, 10 were in NYHA functional class I, and three were in NYHA functional class II. There was no recurrence of sub diaphragmatic neoplasm or intracardiac neoplasm during the follow-up.

One patient (Patient No. 2) underwent a salpingo-oophorectomy during the primary procedure, and four patients (Patients No. 1, 4, 5 and 12) underwent second stage salpingo-oophorectomy 2 to 4 weeks after the primary procedure. Another two patients (Patients No. 3 and 7) underwent resection of the pelvic fibroids 4 weeks after the primary procedure. Out of these seven patients, the internal iliac veins or the ovarian veins from which the ICL originated were all ligated, and other than Patient No. 7 who had an incomplete excision of the intumescences at the orifice of the IVC, there were no recurrences of ICL or IVL.

Discussion

Intravenous leiomyomatosis most commonly enters through the lumen of the iliac vein, and grows into the IVC, sometimes reaching the right atrium, ventricle, and pulmonary artery.[5,6] Occasionally, the ovarian vein provides an alternative route to the subphrenic segment of

the IVC. The diagnosis of leiomyoma extending to the right side of the heart may be made when facing a middle-aged woman with a right-sided heart tumour, especially in the case of prior hysterectomy or subtotal hysterectomy.[7] A CT scan provides the best diagnostic information, since it can demonstrate an intravenous tumour with an ovarian or internal iliac vein as well as IVC and right atrium involvement.[8,9] Added to the information obtained from echocardiography and type B ultrasonography, we can obtain the needed information for classification using the quaternary classification scheme for ICL.

The necessity of classifying the ICL

Although leiomyoma is histologically benign and characterised by slow growth, ICL can lead to severe complications such as syncope, right heart failure, or ascites and oedema in the lower extremities, and should be radically excised. But the leiomyomatosis extending to the right chambers of the heart is a very rare condition and there are no more than 200 cases reported in the literature. To date only sporadic reports can be reviewed regarding pathological findings. Academic circles have an exiguous understanding of the subject, and the surgical strategy, prognosis and follow-up of ICL are now somewhat uncertain to medical world. There is discord in many aspects of ICL, such as classification and preprocedure evaluation, and the surgical strategy to be chosen. For example, when facing an ICL, a cardiac surgeon can choose sternolaparotomy, sternotomy or just a laparotomy as the surgical approach. He or she also can choose to conduct the operation with or without a CPB, and can also choose normothermic CPB without clamping the aorta or moderate hypothermia CPB with cardiac arrest, or DHLF or DHCA as the supporting measure for the procedure. The surgeon also can choose to drain the vena cava through cannulating both the SVC and IVC, or cannulating the SVC and femoral vein on the healthy side, or cannulating the jugular vein and femoral vein on the healthy side. A large number of techniques have been utilised for tumour excision: pull the tumour out through the iliac vein or the opened right atrium, or directly excise the pedicle of the ICL. The procedure can be performed in a one- or two-stage procedure, but there are no existing report standards for ICL management.

Several factors may explain the heterogeneity during the procedure being chosen and adopted. Firstly, because of poor documentation on these tumours, such as insufficient knowledge of tumour anatomy or erroneous preoperative diagnosis of myxoma, thrombus in the right sided chambers of the heart or IVC, sometimes the correct diagnosis can only be made intraoperatively.[10] These situations have led surgeons to improvise surgical strategy. Secondly, surgical strategy may be guided by "analogous thinking" from more experienced surgical situations. Removal of an extension to the IVC of a kidney neoplasm for instance, often requires a complete circulatory arrest. Similarly, this strategy may be chosen for ICL. In some cases in our series, only a few minutes of CPB were required to remove the tumour, suggesting that circulatory arrest and associated risks can be avoided on most occasions. To date, there has been no effort to classify ICL in any of the global literature. Hence we put forward this new classification system for ICL, in order to provide a manageable tool and a basis for standardising the reporting of future ICL management.

The demographic characteristics of ICL and the relationship between clinical manifestations and the quaternary classification scheme for ICL

A paper on intravenous leiomyomatosis with cardiac extension was first published in 1907 from an autopsy analysis.[11] According to our literature research using MEDLINE, from 1980 to 2007, there were 87 cases of ICL with detailed anatomic pathological descriptions reported in English and Chinese literature. All reported patients were women, ranging in age from 26 to 72 with a mean age of 47 years, and a history of uterine leiomyoma was observed in each case. Most patients were in the fifth and early sixth decades of life and were sometimes asymptomatic.[12,13] The patients in our series were relatively younger (mean, 43.5 ± 9.8 years old) than in these earlier studies.

The quaternary classification scheme for ICL can be used to stratify and understand the symptoms of different types of ICL. The occlusion of the IVC leads to typical signs and symptoms of right heart failure.[14] Total obstruction of the tricuspid valve will result in sudden death.[15,16] This kind of emergency mostly appeared in Y type ICL, which has large intumescences in its intracardiac part. For the NA type ICL however, there are no signs of lower extremity oedema or ascites, or incarceration in the right side chambers of the heart. B, C or D types of ICL would have signs of IVC blockage, leading to collateral circulation established around the blockage so much

more haemorrhage would happen when dissecting the IVC during the resection procedure. ICL grows along different routes of the venous system into the IVC and the right atrium. The different route of extension makes a difference in the ease of excision of tumour masses. These patients are sometimes symptomatic.[12]

Choosing the surgical strategy through the quaternary classification scheme for ICL

The first grade classification refers to the relative size of the ICL intracardiac part to the diameter of IVC (N or Y type). It can decide if the ICL body needs to be taken out through a right atrium incision. Y type ICL must be taken out through the opening in the right atrium. However for N type ICL, it is not necessary to take ICL out through right heart opening, and ICL could be taken out through IVC or the internal iliac vein or vena ovarica incision. If we decide to take out ICL through IVC or the internal iliac vein or vena ovarica incision, we could cannulate the jugular vein and femoral vein in the healthy side, and the femoral artery to establish CPB. Through this method of cannulation, sternotomy can be avoided, and only laparotomy is needed. For NA type ICL, the CPB support can also be spared.[17–19] For cases of YB or YC or YD type ICL, an abdominal plus thoracic approach is mandatory; the tumour dissected from the IVC must be taken out through the open atrium under CPB with SVC and IVC drainage. Some authors recommend that since this is a "quick and easy" procedure a circulatory arrest is not necessary; and a right mini-thoracotomy may be preferred to a sternotomy.[18,20]

The 2nd grade classification refers to the relative size of the intravenous part of ICL to the diameter of IVC (A, B, C, D type). It can decide the surgical approach. A type (NA or YA type) ICL can be approached just through sternotomy. The intracaval part and intracardiac part of ICL body could be taken out through the right atrium opening under the support of DHLF or moderate hypothermia with cardiac arrest or even normothermia CPB without clamping the aorta. NA type ICL can also be approached only through laparotomy. Some literature highlighted concerns that blind traction of the tumour seemed dangerous, and caution must be taken during removal of the tumour. Simply pulling out the tumour from the right atrium is not feasible. The site of the attachment is located in the pelvic veins, and attempted removal just from the thoracic approach results in either the failure of the complete retrieval of the tumour or tearing of the vein at the point of attachment. Intraoperative deaths have occurred from massive retro peritoneal haemorrhage after avulsion of the tumour from the cardiac end. But seven cases in our series which involved pulling out the ICL pedicles did not have this kind of complication and the recurrence of the IVL and ICL could be prevented through second stage hysterectomy and ligation of the internal iliac vein or ovarian vein. Any B, C, and D type ICL should be approached through sterno laparotomy. To establish a CPB for B, C, and D type ICL, it is needed to cannulate the ascending aorta and SVC, as well as the femoral vein on the healthy side. The dissection of the adherence and blockage for B, C, and D type ICL should be undertaken under the support of DHCA. Especially in the case of C type ICL, the hepatic falciform ligament in combination with the coronary ligament, or the deltoid ligament on its own, should be dissected free to expose the retro hepatic segment of the IVC, thus the IVC could be opened to dissect the intumescences of the ICL, in order to take out the body of the intumescences and its two ends under the support of DHCA.

The third grade classification refers to the area of origin of the ICL (I or O type). I or O type can decide the site, which is the internal iliac vein or vena ovarica, to excise the ICL pedicles. The tumour has only ever been found attached to the vascular bed at one site, either the osteium of the internal iliac vein or the ovarian vein. Research about the histogenesis of ICL revealed that the ICL does not originate from the vessel wall but from the seeding of the uterus and its annexa.[21]

For the removal of the intracaval tumour, a literature review showed a preference of conducting supra and infra renal vena cava venotomy. However, as the leiomyoma usually does not adhere to the vessel wall, for removal of the remaining caval portion it is recommended that an iliac venotomy should be taken. It is believed that this incision has the following advantages over caval and especially suprarenal caval level incisions: (1) Fast perioperative recovery due to less retro peritoneal exploration; (2) Surgical complications of the iliac region are easier to cope with than in the caval region; (3) Venous thrombosis due to the venotomy is more tolerable in the iliac region; (4) Better cosmetic results; (5) The advantage of being closer to the origin of the tumour, i.e. in the uterus.

The fourth grade classification refers to the laterality of the lesion in the pelvis (L or R type). L or R type can

decide the deviation direction of the laparotomy when needed.

One-stage or two-stage procedures

If the tumour is too extensive, or adheres to the cardiac and vascular structures requiring resection of the abdomino-pelvic and intrathoracic components, a separate operation may be mandatory.[22] Otherwise, one-stage resection under total circulatory arrest and hypothermia can be used successfully.[23–25] Although the literature review failed to explain the lower incidence of ectopic recurrence with single-stage resection under total circulatory arrest, the risk of intraoperative seeding is likely to be reduced by arrested circulation.[26] A simple and safe approach of removing the ilio-caval portion of the tumour in intravascular leiomyomatosis, iliac venotomy may be used in one- and two-stage operations. It is recommended to remove the ilio-caval portion of the tumour in both stages of operations using iliac venotomy.[27] The first stage of the operation should be intracardiac and intravenous leiomyomatectomy; and the second stage of the operation should involve hysterectomy and pelvic dissection.

The early and late efficacy of surgical treatment for ICL

According our retrospective analysis and follow-up of the 14 cases of ICL, the surgical mortality is low, and the recurrence is also low. The only recurrence is due to the incomplete excision of the lesions (Patient No. 7 with NBOL type ICL), and we surmise that this recurrence would have been prevented if this ICL case had taken a surgical procedure under the guidance of our suggested classification scheme by using DHCA. Other than Patient No. 7 who had an incomplete excision of the intumescences at the orifice of the IVC at the right atrium, there was no recurrence of ICL or IVL in the other six cases in our series by pulling out the ICL pedicles. This suggests that the recurrence of IVL and ICL can be prevented through hysterectomy and ligation of the internal iliac vein or the ovarian vein. It can be determined that surgical resection is the best treatment for intracardiac extension of intravenous leiomyoma, and the patients may regain satisfactory heart function after the excision of ICL.

In conclusion, surgical resection is the best treatment for ICL and must be performed immediately due to the risk of sudden death. The surgical treatment of ICL can result in a good mid to long term survival rate with satisfactory heart function. The quaternary classification scheme for ICL could be a useful tool used to choose the surgical strategy for the treatment of ICL and can serve as the basis for standardising the reporting of future ICL management.

References

1. Ma SQ, Bai CM, Yu XH, et al. Clinical and pathological analyses of intravenous leiomyomatosis. *Chin J Obstet Gynecol* 2005;40: 34–7.
2. Akatsuka N, Tojunaga K, Isshiki T, et al. Intravenous leiomyomatosis of the uterus with continuous extension into the pulmonary artery. *Jpn Heart J* 1984;25:651–9.
3. Ricci MA, Cloutier LM, Mount S, et al. Intravenous leiomyomatosis with intracardiac extension. *Cardiovasc Surg* 1995; 3:693–6.
4. Gawne-Cain ML, Wilson AG, Corbishley C, et al. Case report: intravenous leiomyomatosis, an unusual cause of intracardiac filling defect. *Clin Radiol* 1995;50:123–5.
5. Wakiyama H, Sugimoto T, Ataka K, et al. Intravenous leiomyomatosis extending into the right ventricular cavity: one-stage radical operation using cardiopulmonary bypass—a case report. *Angiology* 2000;51:505–9.
6. Mandelbaum I, Paulletto FJ, Nasser WK. Resection of a leiomyoma of the inferior vena cava that produced tricuspid valvular obstruction. *J Thorac Cardiovasc Surg* 1974;67:561–7.
7. Mozzola A, Gregorini R, Procaccini B, et al. Intracaval and intracardiac leiomyomatosis of uterine origin. *Ann Vasc Surg* 1986;1:134–8.
8. Lai TK, Huang HY, Chan RY, et al. Magnetic resonance venogram of intravenous leiomyomatosis. *Hong Kong Med J* 2005;11:524–6.
9. Wong YY, Chu WC, Lam WW. Intravenous leiomyomatosis: computed tomography diagnosis. *Hong Kong Med J* 2006;12: 239–40.
10. Le Bouedec G, Bailly C, Penault-Llorca F, et al. [Intravascular leiomyomatosis of uterine origin. A case of pseudo-metastatic cavo-cardial thrombus] *Press Med* 1999;28:1463–5.
11. Durck H. Ein kontinuierlich durch die untere. Hohlvene in das Herz vorwaschsendes Leiomyom. *München Med Wochnschr* 1907; 54:1154–5.
12. Topcuoglu MS, Yaliniz H, Poyrazoglu H, et al. Intravenous leiomyomatosis extending into the right ventricle after subtotal hysterectomy. *Ann Thorac Surg* 2004;78:330–2.
13. Lam PM, Lo KWK, Yu MY, et al. Intravenous leiomyomatosis: two cases with different routes of tumour extension. *J Vasc Surg* 2004;39:465–9.
14. Cooper MM, Guillen J, Dalton J, et al. Recurrent intravenous leiomyomatosis with cardiac extension. *Ann Thorac Surg* 1992; 52:139–42.
15. Gonzalez-Lavin L, Lee HR, Falk L, et al. Tricuspid valve obstruction due to intravenous leiomyomatosis. *Am Heart J* 1984;108: 1544–6.

16. Rosenberg JM, Marvast MA, Obeid A, et al. Intravenous leiomyomatosis: a rare cause of right sided cardiac obstruction. *Eur J Cardiothorac Surg* 1988;2:58–60.
17. Moorjani N, Kuo J, Ashley S, et al. Intravenous uterine leiomyosarcomatosis with intracardiac extension. *J Card Surg* 2005;20:382–5.
18. Subramaniam B, Pawlowski J, Gross BA, et al. TEE-guided one-stage excision of intravenous leiomyomatosis with cardiac extension through an abdominal approach. *J Cardiothorac Vasc Anesth* 2006;20:94–5.
19. Gehr NR, Lund O, Alstrup P, et al. Recurrence of uterine intravenous leiomyomatosis with intracardiac extension. Diagnostic considerations and surgical removal. *Scand Cardiovasc J* 1999; 33:312–4.
20. Stegmann T, Garcia-Gallant R, Doring W. Intravascular leiomyomatosis: report of a case and review of the literature. *Thorac Cardiovasc Surg* 1987;35:157–60.
21. Kir G, Kir M, Gurbuz A, Karateke A, Aker F. Estrogen and progesterone expression of vessel walls with intravascular leiomyomatosis; discussion of histogenesis. *Eur J Gynaecol Oncol* 2004;25:362–6.
22. Filsouri F, Farivar RS, Anderson C, Santerre D, Adams DH. Renal vein injury complicating removal of intravenous leiomyoma. *J Thorac Cardiovasc Surg* 2002;123:820–2.
23. Suginami H, Kaurar R, Ochi H, et al. Intravenous leiomyomatosis with cardiac extension. *Ann Thorac Surg* 1992;53:139–41.
24. Okamoto H, Itoh T, Morita S, et al. Intravenous leiomyomatosis extending into the right ventricle: one-stage radical excision during hypothermic circulatory arrest. *Thorac Cardiovasc Surg* 1994; 42:361–3.
25. Arinami Y, Kodama S, Kase H, et al. Successful one-stage complete removal of an entire intravenous leiomyomatosis in the heart, vena cava, and uterus. *Gynecol Oncol* 1997;64:547–50.
26. Katsumata T, Shinfeld A, Houel R, et al. Pelvic leiomyma in the right atrium. *Ann Thorac Surg* 1998;66:2095–6.
27. Stols NAG, Santos G, Haddad VLS, et al. Successful one-stage resection of the intravenous leiomyomatosis of the uterus with extension into the heart. *Cardiovasc Surg* 1999;7:661–4.