Intravenous leiomyomatosis with intracardiac extension: an unusual cause of cardiac syncope

Pui-Kee Lee, MB, BS, FRCP (Edin); Tirone E. David, MD, FRCSC; Cairrine Sloggett, RN, RDMS; John R. Ross, MD, FRCPC

Recurrent syncope is common. Careful history-taking, the details often checked with a reliable witness, and a thorough general and neurologic examination are the most useful aids that lead to appropriate investigations and diagnosis.[1] A cardiovascular cause should be considered, particularly with a history of abrupt onset and recovery. The cause is usually arrhythmia or a sudden reduction in stroke volume. Intracardiac tumours are a rare cause of syncope, atrial myxomas being the commonest. Only 15 cases of intravenous leiomyomatosis with intracardiac extension have been reported in the literature.[2,3] As described here, this condition is an unusual but curable cause of cardiac syncope.

Case report

A 38-year-old woman was referred to the Cardiology Division of the Wellesley Hospital, Toronto, for assessment of recurrent syncopal episodes. Two years earlier exploration of a pelvic mass had revealed an enlarged uterus infiltrated by a poorly demarcated mass that extended into the parametrial and the retroperitoneal tissue. Hysterectomy, bilateral salpingo-oophorectomy and excision of most of the adjacent mass had been performed. A small stump of the tumour had been left. Histologic examination had revealed the following: the tumour comprised parallel arrays of smooth muscle cells with abundant differentiation to form thick vessels; part of the tumour in the uterus was intravascular; mitotic figures were rare (observed in fewer than 1 in 20 cells per high power field); and nuclear pleomorphism was minimal. The consulting pathologist had diagnosed intravenous leiomyomatosis.

The patient had remained well for 20 months before her first episode of syncope. During the 4 months before referral she had had about a dozen syncopal episodes and a similar number of near-syncopal episodes. The near-syncopal attacks were not associated with palpitations; they were abrupt in onset, with no preceding aura or seizure activity, and recovery was always prompt and complete within minutes. She had also experienced an episode of severe shortness of breath of sudden onset that lasted several hours, without associated syncope or chest discomfort. The possibility of cardiac arrhythmia or seizure disorder had been assessed with 24-hour ambulatory electrocardiographic monitoring and electroencephalography; the findings had been normal.

At referral the woman looked healthy and in no distress; there was no pallor or cyanosis. The jugular venous pressure and the carotid pulse volume and upstroke were normal. The blood pressure was 130/80 mm Hg bilaterally, with no significant postural drop, and the pulse was regular, at 70 beats/min. The cardiac apex was not well localized. The first and second heart sounds were normal. A low-frequency, early diastolic sound and a grade 3 scratchy systolic murmur, both of which increased in intensity on inspiration, were heard at the lower left sternal border. There were no diastolic murmurs. Examination of the chest and the abdomen revealed normal findings, and pelvic examination did not demonstrate tumour recurrence.

Electrocardiograms and chest x-ray films appeared normal. However, two-dimensional echocardiography showed a large, irregular mass of various acoustic densities in the right atrium and ventricle (Fig. 1). The subcostal view revealed extension of the mass from the inferior vena cava. An inferior venacavogram demonstrated a mass in the left common iliac vein that extended to the right atrium and ventricle and into the pulmonary artery.

From the departments of Medicine and Surgery, University of Toronto

Reprint requests to: Dr. John R. Ross, Department of Medicine, Ste. 236, Wellesley Hospital, 160 Wellesley St. E, Toronto, Ont. M4Y 1J3

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During surgery the venous portion of the tumour was found to be attached to the wall of the inferior vena cava at its junction with the left common iliac vein and infiltrating the wall of the left hypogastric vein. This portion of the tumour was dissected. The remainder, which was intravascular, was found not to be attached to the vascular lining of the inferior vena cava or the heart. The tumour was divided in the inferior vena cava to allow the removal of the cardiac portion through the right atrium (Fig. 2). The intra-abdominal and intrapelvic portion of the tumour was subsequently removed. The entire specimen is shown in Fig. 3.

The findings at histologic examination were identical to those for the pelvic tumour removed 2 years earlier. The postoperative course was unremarkable. Clinical and radiologic assessment during follow-up for 3 years have revealed no evidence of tumour recurrence.

Comments

Intravenous leiomyomatosis is rare and is characterized histologically by the benign growth of smooth muscle cells in the lumen of veins. In most case reports the intraluminal extension arose from the uterine and pelvic veins,[2,4,5] although origin from the inferior vena cava has also been reported.[6] Extension into the right cardiac chambers is rare, only 15 cases having been reported.[2,3] Clinical presentations of intracardiac leiomyomatosis have varied widely. The tumour may cause no symptoms and be an incidental finding at echocardiography, or it may cause sudden death. Symptoms have included those of outflow obstruction of the inferior vena cava, the right atrium or both. A history of syncope, such as described in this case, is an indication of intermittent severe cardiac obstruction.

Intracardiac lesions can be diagnosed easily with two-dimensional echocardiography. Computed tomography, contrast angiography or both can be used to delineate the extracardiac component of the tumour. The differential diagnosis of a mass extending from the inferior vena cava to the right cardiac chambers includes direct extension of a tumour from the liver, kidney or other intra-abdominal organ and thrombus arising in the inferior vena cava.

The primary goal of therapy is the complete removal of the tumour. Recurrences up to 17 years after primary excision have been reported, but even in those cases the literature supports a good long-term prognosis after additional surgery.[7] The possible role of hormone therapy was raised by the detection of cytoplasmic estradiol and progesterone receptors in one case of intracardiac leiomyomatosis.[8] In addition, regression of uterine leiomyomas

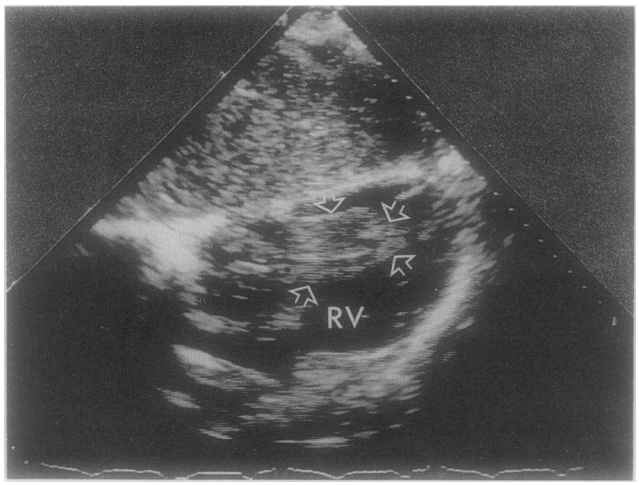

Fig. 1: Two-dimensional echocardiogram, showing subcostal view of mass in right ventricle.

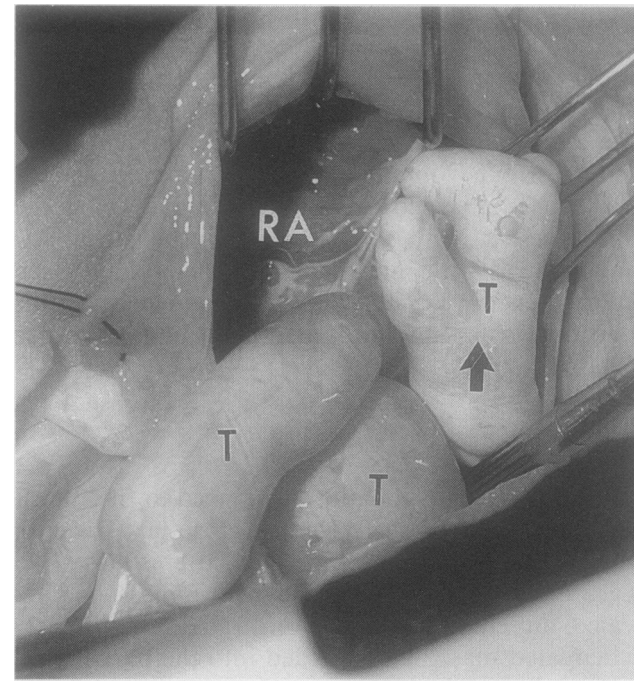

Fig. 2: Opened right atrium (RA) and three "tentacles" of tumour (T). One tentacle (arrow) had autoamputated; hence its whiteness.

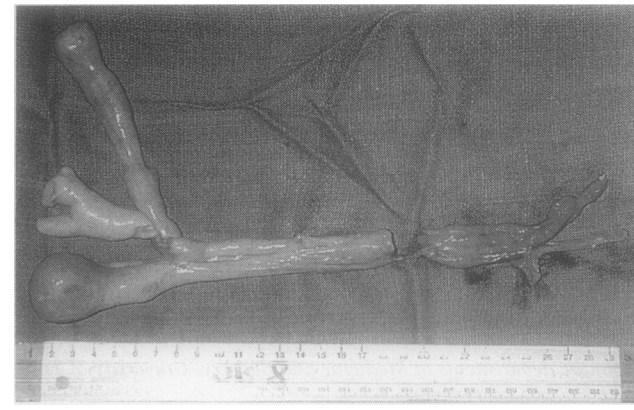

Fig. 3: Entire resected tumour.

after luteinizing hormone releasing hormone agonist therapy has also been reported.[9]

This case emphasizes the importance of long-term follow-up of patients after resection of intravenous leiomyomas. The recurrence of tumour can usually be managed surgically. Hormone therapy should also be considered in cases of unresectable residual tumour.

References

1. Kapoor W, Karf M, Wieand S et al: A prospective evaluation and follow-up of patients with syncope. *N Engl J Med* 1983; 309: 197-204
2. Politzer F, Kronzon I, Wieczorek R et al: Intracardiac leiomyomatosis: diagnosis and treatment. *J Am Coll Cardiol* 1984; 4: 629-634
3. Tsutomu S, Masahiro Y, Hisayuki C et al: Intravenous leiomyomatosis of the pelvis with re-extension into the heart. *Ann Thorac Surg* 1986; 42: 104-106
4. Marshall JF, Morris DS: Intravenous leiomyomatosis of the uterus and pelvis. *Ann Surg* 1959; 149: 126-134
5. Bahary CM, Gorodeski IG, Nilly M et al: Intravascular leiomyomatosis. *Obstet Gynecol* 1982; 59 (suppl): 73S-77S
6. Mandelbaum I, Pauletto FJ, Nasser WK: Resection of a leiomyoma of the inferior vena cava that produced tricuspid valvular obstruction. *J Thorac Cardiovasc Surg* 1974; 249: 2799-2802
7. Norris H, Parmley T: Mesenchymal tumour of the uterus: 5. Intravenous leiomyomatosis. *Cancer* 1975; 36: 2164-2178
8. Tierney WM, Ehrlich CE, Bailey JC et al: Intravenous leiomyomatosis of the uterus with extension into the heart. *Am J Med* 1980; 69: 471-475
9. Maheux R, Gwilloteau C, Lemay A et al: Regression of leiomata uteri following hypoestrogenism induced by repetitive luteinizing hormone-releasing hormone agonist treatment: preliminary report. *Fertil Steril* 1984; 41: 863-871

Conferences

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Sept. 13-15, 1990: New Brunswick Medical Society Annual General Meeting
Hotel Beauséjour, Moncton
Ms. Judy Orem, annual general meeting coordinator, New Brunswick Medical Society, 176 York St., Fredericton, NB E3B 3N8; (506) 458-8860

Sept. 13-15, 1990: Ontario Medical Association and Canadian Anaesthetists' Society Annual Fall Meeting
Niagara Falls, Ont.
Dr. F. Halliday, OMA/CAS Annual Fall Meeting, c/o Greater Niagara General Hospital, PO Box 1018, Niagara Falls, Ont. L2E 6X2; (416) 358-0171, ext. 474

Sept. 14-16, 1990: Canadian Hospital Association 7th Annual Invitational Seminar on Health Care Directives
Millcroft Inn, Alton, Ont.
Conferences, Canadian Hospital Association, 100-17 York St., Ottawa, Ont. K1N 9J6; (613) 238-8005, FAX (613) 238-6924

Sept. 14-17, 1990: Royal College of Physicians and Surgeons of Canada Annual Meeting (held in conjunction with the Annual Meeting of the Canadian Pediatric Society and Canadian Society for Clinical Investigation)
Metro Toronto Convention Centre
Anna Lee Chabot, coordinator, Royal College of Physicians and Surgeons of Canada, 74 Stanley St., Ottawa, Ont. K1M 1P4; (613) 746-8177, FAX (613)746-8833

Sept. 14-17, 1990: 67th Annual Meeting of the Canadian Paediatric Society (held in conjunction with the Annual Meeting of the Royal College of Physicians and Surgeons of Canada)
Metro Toronto Convention Centre
Dr. Victor Marchessault, executive vice-president, Canadian Paediatric Society, 401 Smyth Rd., Ottawa, Ont. K1H 8L1; (613) 737-2728

Sept. 15-23, 1990: British Medical Association Annual Scientific Meeting
Edinburgh
Meetings Department, PO Box 8650, Ottawa, Ont. K1G 0G8; 1-800-267-9703, FAX (613) 731-9013

Sept. 21-23, 1990: Dermatology '90: Therapeutic Update
New World Harbourside, Vancouver
Dermatology '90, 204-402 W Pender St., Vancouver, BC V6B 1T6; (604) 669-7175, FAX (604) 669-7083

Sept. 22-28, 1990: 23rd International Congress on Occupational Health: Sharing Solutions
Montreal Convention Centre
Secretariat, 23rd International Congress on Occupational Health, 2-58 de Brésoles St., Montreal, PQ H2Y 1V5; (514) 499-9835, FAX (514) 288-4627

Oct. 1-5, 1990: Canadian Society of Forensic Science Annual Conference
Skyline Hotel, Ottawa
Canadian Society of Forensic Science, 215-2660 Southvale Cres., Ottawa, Ont. K1B 4W5; (613) 731-2096

Oct. 11-12, 1990: Histopathologic Diagnosis of Inflammatory and Neoplastic Skin Diseases: Assessment of Patterns and Silhouettes
Halifax Sheraton
Dr. Noreen Walsh, Department of Pathology, Victoria General Hospital, Rm. 721, D.J. MacKenzie Building, 1278 Tower Rd., Halifax, NS B3H 2Y9; (902) 428-3897

Oct. 11-14, 1990: Canadian Pain Society (IASP Chapter) Annual Meeting
London, Ont.
Ms. Inese Kramins, Local Arrangements Committee, Department of Psychology, University of Western Ontario, London, Ont. N6A 5C2

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