

CASE REPORT

Intravenous Lobular Capillary Haemangioma (Pyogenic Granuloma) of the Superior Vena Cava: Case Report and Literature Review

Elisabeth Blaya ^a, Vincenzo Vento ^{a,b}, Salomé Kuntz ^{a,b}, Laurence Bruyns ^{a,b}, Mickael Ohana ^{b,c}, Noelle Weingertner ^d, Anne Lejay ^{a,b}, Nabil Chakfé ^{a,b,*}

^a Department of Vascular Surgery and Kidney Transplantation, University of Strasbourg, France

^b GEPROVAS, Groupe Européen de Recherche sur les Prothèses appliquées à la chirurgie vasculaire, Strasbourg, France

^c Department of Radiology, University of Strasbourg, France

^d Department of Pathology, University of Strasbourg, France

Introduction: Intravascular lobular capillary haemangioma is a rare benign intravascular tumour, especially in large vessels. This is the report of a case and associated literature review.

Report and literature review: This is the report of the first case of an intravenous lobular capillary haemangioma (ILCH) of the superior vena cava (SVC). A 30 year old female presented with a collateral thoraco-abdominal venous circulation. Chest computed tomography angiography, thoracic magnetic resonance imaging, and positron emission tomography revealed an intraluminal SVC tumour extending from the left brachiocephalic venous trunk to the distal third of the SVC. No pre-operative biopsy was indicated. An *en bloc* tumour excision was performed, followed by reconstruction of the SVC with an L shaped, ringed polytetrafluoroethylene (PTFE) prosthesis. Histopathology revealed the presence of an ILCH with free margins. A review of the literature identified 64 cases of ILCH to date, all of which underwent total resection. When reported, no recurrences were found during follow up.

Discussion: In this case, the ePTFE reconstruction of the SVC must be checked regularly for any adverse events. Although ILCH is a benign tumour with no risk of recurrence, regular surveillance is advised.

© 2020 The Authors. Published by Elsevier Ltd on behalf of European Society for Vascular Surgery. This is an open access article under the CC BY license (<http://creativecommons.org/licenses/by/4.0/>).

Article history: Received 19 August 2020, Revised 21 November 2020, Accepted 10 December 2020,

INTRODUCTION

Primary vascular tumours of the vena cava are rare and occur mainly in the inferior vena cava (IVC). The ISSVA Classification describes two main types of malignant vascular tumours: angiosarcoma, (mainly leiomyosarcoma, generally in the IVC, none in superior vena cava [SVC]) and epithelioid haemangio-endothelioma. Lobular capillary haemangioma (LCH), also known as pyogenic granuloma, is one of the many benign vascular tumours. It occurs mainly in the skin and subcutaneous tissue of the upper limb, neck, and head. Intravascular lobular capillary haemangioma (ILCH) is a subtype of LCH and only few cases have been reported in the literature.^{1,2}

This is the first case of intravenous lobular capillary haemangioma in the superior vena cava (SVC) reported in the literature. A technique is described for reconstruction of

the SVC using an inverse L shaped synthetic graft, after an *en bloc* resection of the tumour.

CASE REPORT

A 30 year old female, without significant medical history, presented with recent aggravation of a thoraco-abdominal venous circulation developed during her pregnancy, five years earlier. She had no other complaint or symptom. Clinical abdominal and cardiovascular examinations did not reveal any specific clinical signs of portal hypertension or SVC syndrome.

Duplex ultrasound (DUS) showed dilatation of collateral veins, while the IVC was patent. Chest computed tomography angiography (CTA) (Revolution Evo G.E. Medical, Chicago, IL, USA) demonstrated an intraluminal heterogeneous mass in the SVC, extending from the left brachiocephalic venous trunk to the distal third of the SVC (Fig. 1A). No other mass or adenopathy was present. Thoracic magnetic resonance imaging (Ingenia Philips 3T, Eindhoven, the Netherlands) confirmed the intraluminal SVC location of the tumour, measuring 80 × 33 mm, with a cystic portion demonstrating heterogeneous T1 weighted and T2 weighted signal (Fig. 1B and C). The azygos vein had not been invaded. Positron emission tomography (Vision Siemens, Munich, Germany) demonstrated moderate

* Corresponding author. Department of Vascular Surgery and Kidney Transplantation, Hôpitaux Universitaires de Strasbourg, B.P. 426, 67091 Strasbourg Cedex, France.

E-mail address: nabil.chakfe@chru-strasbourg.fr (Nabil Chakfé).

2666-688X/© 2020 The Authors. Published by Elsevier Ltd on behalf of European Society for Vascular Surgery. This is an open access article under the CC BY license (<http://creativecommons.org/licenses/by/4.0/>).

<https://doi.org/10.1016/j.ejvsf.2020.12.021>

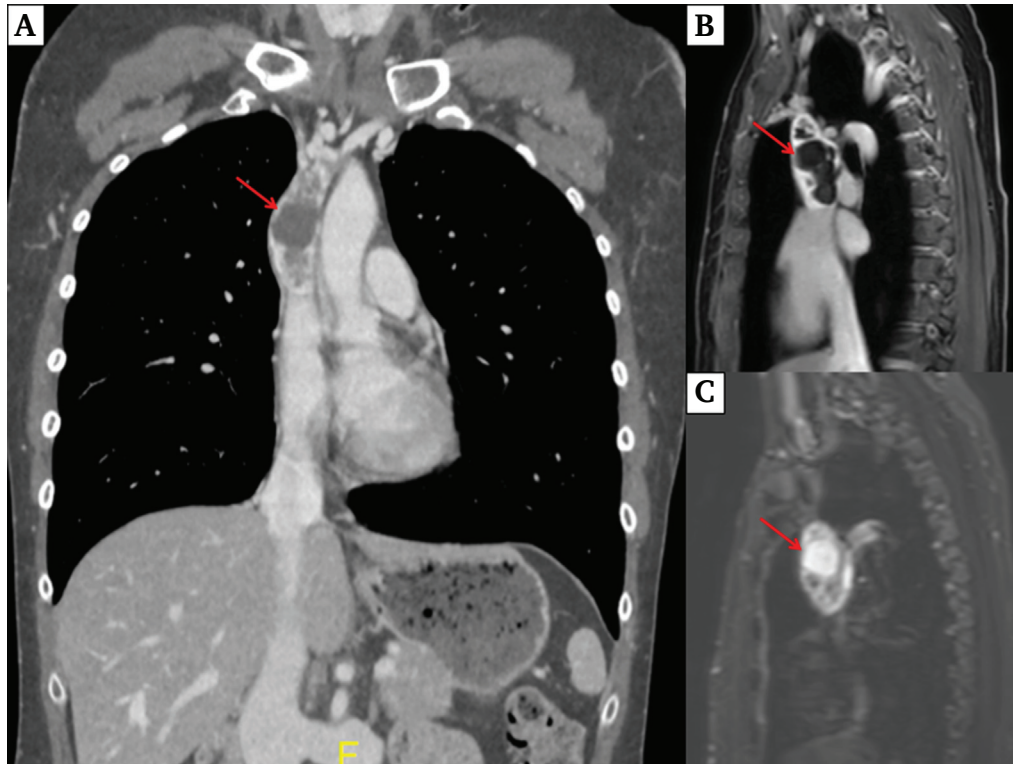


Figure 1. (A) Thoracic computed tomography angiography, coronal reconstruction showing the tumour (red arrow) filling the superior vena cava lumen. (B) Thoracic magnetic resonance imaging T2 weighted, and (C) T1 weighted with gadolinium injection.

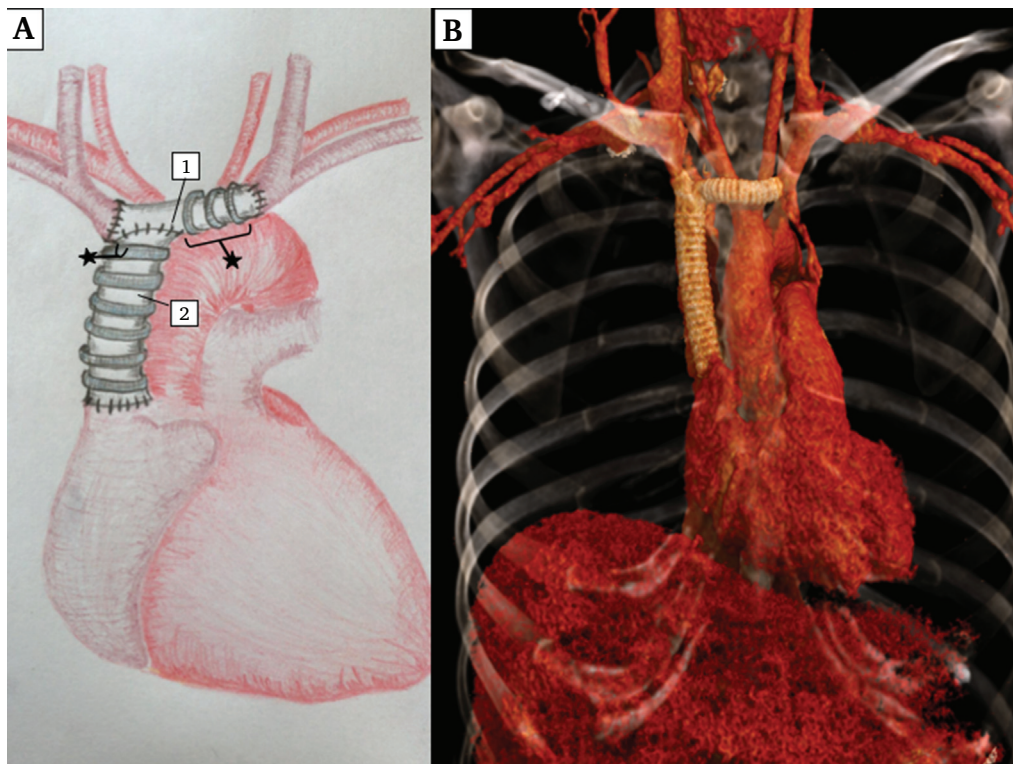


Figure 2. (A) Post-operative schematic representation of the superior vena cava reconstruction with 1) 8 mm polytetrafluoroethylene graft and 2) 16 mm polytetrafluoroethylene graft. (B) Post-operative thoracic computed tomography angiography 3D reconstruction.

fluorodeoxyglucose enhancement (SUV max: 4.8). Trans-thoracic sonography confirmed no extension into the right atrium. The oncology multidisciplinary staff reviewed the preliminary findings and suspected a primary SVC tumour or SVC thrombosis. A direct biopsy was too risky in terms of spread if malignant, and an endovascular biopsy would not have been sufficient. It was decided to perform surgical resection without radiotherapy or chemotherapy.

Surgery consisted of an *en bloc* resection of the tumour through a median sternotomy. The SVC was resected from its junction with the right atrium without myocardial breach and taking both left and right brachiocephalic veins. The venous reconstruction consisted of a 16 mm diameter, ringed, expanded polytetrafluoroethylene (ePTFE) prosthesis (Gore-Tex, W.L. Gore & Associates, Flagstaff, AZ, USA) sutured distally to the right atrium junction and proximally, perpendicularly on an 8 mm diameter ringed ePTFE prosthesis, connecting both right and left venous confluences (Fig. 2A). There was no need for cardiopulmonary bypass during the intervention. A left brachio basilic fistula was created to increase blood flow and avoid thrombosis of the prosthesis. The post-operative course was complicated by a pericardial effusion with signs of right heart failure, without any complications of the venous reconstruction. A pericardial drain was placed as an emergency. The patient was

discharged on day 26 with anticoagulant therapy (warfarin 7.5 mg once a day), as recommended in venous bypass. Six month DUS and CTA follow up showed graft patency. Fistula closure and anticoagulation interruption were conducted seven months post-operatively.

Pathological examination showed a tumour measuring 55 × 34 × 25 mm, filling the SVC lumen (Fig. 3A and B). Microscopic analysis identified a vascular lesion with lobular architecture, consisting of multiple capillaries lining unstratified endothelium without atypia separated by a fibro-oedematous stroma (Fig. 3C and D). Immunohistochemical analysis showed vascular markers, CD31 and ERG, characterising the endothelium. No sign of malignancy was found and the Ki67 proliferation index was low. After confirmation with the *Réseau de Référence en Pathologie des Sarcomes des Tissus Mous* (National French Reference Network of Soft Tissues Sarcomas), the final diagnosis was an ILCH in the SVC, also known as pyogenic granuloma.

LITERATURE REVIEW

A systematic search was performed of the Medline database from 1979 to 2020 by a combined search strategy of MeSH terms (intravenous lobular capillary haemangioma, pyogenic granuloma). All titles and abstracts collected from the search strategy were screened for relevance. The first 20 related

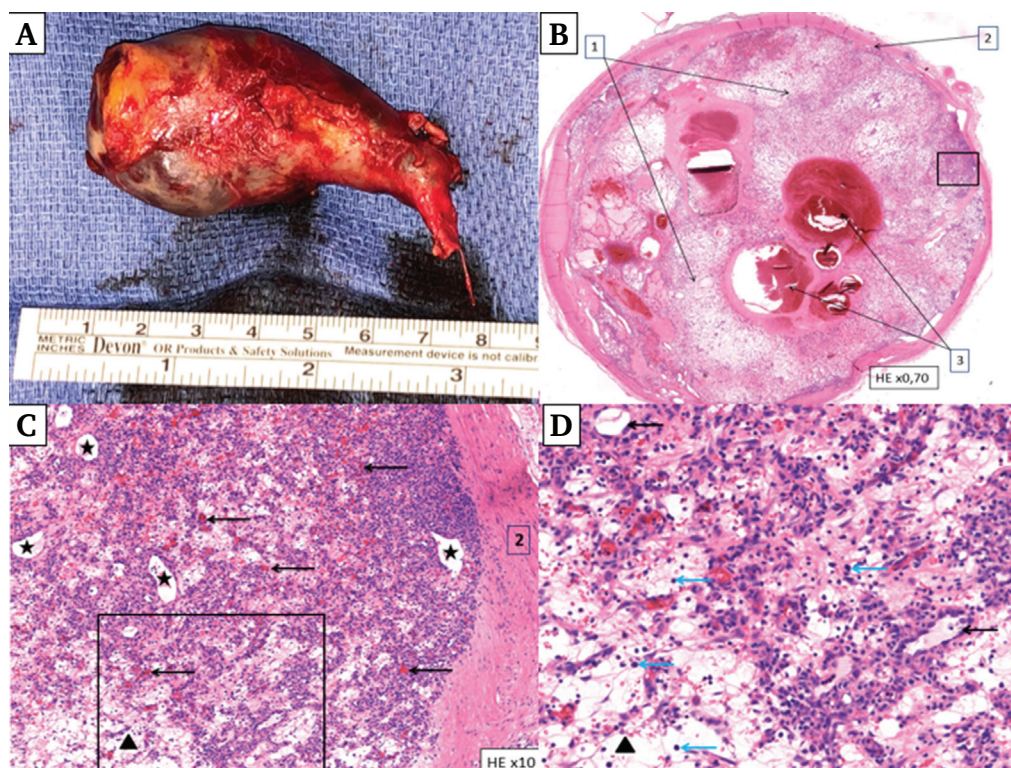


Figure 3. (A) Macroscopic view of the superior vena cava specimen: the tumour was strictly intraluminal. (B) Histology of a transverse cross section of the tumour demonstrated an intravenous (venous wall [1]) mass obstructing the lumen (2) with lobular architecture and thrombotic re-organisation (3) (haematoxylin and eosin stain [H&E] ×0.70). (C) Close up picture of the mass showed numerous small capillaries (arrows) and larger vessels (black stars) within fibro-oedematous stroma (black triangle) (H&E ×10). (D) At higher magnification, capillaries showed unstratified endothelium without atypia (black arrow), accompanied by inflammatory cells (blue arrow) (H&E ×20).

Table 1. Review of literature regarding intraluminal capillary haemangioma.*

Authors, date	Sex/age	Anatomical location	Clinical data	Therapy	Outcome data
Cooper, 1979 ⁴	18 cases NR	Neck×6 Arm×2 Forearm×6 Wrist×2 Finger×2	Not specified	Surgical resection	No recurrence Average of follow up 8.2 y
Ulbright, 1980 ⁹	M/12	External jugular vein	Neck nodule	Surgical resection	No recurrence at 1 y
Anderson, 1985 ¹⁰	F/62	Palmar vein	Palmar nodule	Surgical resection	No recurrence at 21 mo
Truong, 1985 ¹¹	M/44	Branch of angular vein	Lachrymal sac nodule	Surgical resection	No recurrence at 7 y 11 mo
	M/68	Not precise	Inner canthus nodule	Surgical resection	No recurrence at 4 y 7 mo
DiFazio, 1989 ¹²	F/37	Palmar vein	Palmar nodule	Surgical resection	No recurrence at 16 mo
Saad, 1993 ¹³	M/35	Left temple	Nodule	Surgical resection	NR
Margo, 1994 ¹⁴	M/27	Temporal artery	Temporal nodule	NR	NR
Pesce, 1996 ¹⁵	M/20	Lip vein	Nodule	Surgical resection	NR
Danz, 1997 ¹⁶	M/79	Portal vein	Asymptomatic — autopsy finding	NR	NR
Hull, 1999 ¹⁷	M/73	Renal vein	Asymptomatic — investigation for benign prostate hyperplasia	Total nephrectomy	NR
Domanski, 1999 ¹⁸	F/15	Neck vein	Neck nodule	Surgical resection	NR
Sarteschi, 1999 ¹⁹	F/56	External jugular vein	Neck nodule	Surgical resection	No recurrence at 2 y
Song, 2001 ²⁰	M/43	External jugular vein	Neck nodule	Surgical resection	NR
Qian, 2001 ²¹	F/26	Forearm vein	Forearm nodule	Surgical resection	NR
Hayashi, 2001 ²²	F/35	Thenar vein	Thenar nodule	Surgical resection	NR
Panchagnula, 2001 ²³	F/12	Neck vein	Neck nodule	Surgical resection	No recurrence at 1 y 6 mo
Kocer, 2003 ²⁴	F/58	Palm vein	Palm nodule	Surgical resection	No recurrence at 5 mo
Ghersin, 2004 ²⁵	M/21	Basilic vein	Elbow nodule	Surgical resection	NR
Hung, 2004 ²⁶	F/44	Palmar vein	Ulcerative nodule of the palm	Surgical resection	No recurrence at 6 mo
Ghekiere, 2005 ²⁷	M/50	Cephalic vein	Forearm nodule	Surgical resection	NR
Madison, 2006 ²⁸	M/20	Superficial palmar branch of radial artery	Thenar nodule	Surgical resection	NR
Jung, 2008 ²⁹	M/51	Cephalic vein	Forearm nodule	Surgical resection	NR
Pradhan, 2008 ³⁰	F/75	Right internal iliac vein	Abdominal pain and diarrhoea	Surgical resection	No recurrence at 2 mo
Vijayan, 2008 ³¹	M/16	Superficial vein	Finger nodule	Surgical resection	No recurrence at 18 mo
Kamishima, 2009 ³²	F/56	Non-specified vein	Finger nodule	Surgical resection	NR
	M/66	Non-specified vein	Finger nodule	Surgical resection	NR
Winn, 2009 ³³	F/47	Angular vein	Medial canthus nodule	Surgical resection	No recurrence
Maher, 2010 ³⁴	F/41	External jugular vein	Supraclavicular fossa nodule	Surgical resection	NR
Joethy, 2011 ³⁵	M/32	Non-specified	Finger nodule	Surgical resection	No recurrence at 7 mo
Johnson, 2011 ³⁶	F/12	Finger vein	Finger swelling	Surgical resection	No recurrence at 1 ½ mo
Wu, 2011 ³⁷	F/38	Internal jugular vein	Neck nodule	Surgical resection	No recurrence at 1 y
Trombetta, 2011 ³⁸	M/59	Right renal vein	Incidental finding	Total nephrectomy	No recurrence at 5 mo
Takeuchi, 2012 ³⁹	F/36	Left renal vein	Abdominal pain	Total nephrectomy	No recurrence at 4 mo
Turtay, 2012 ⁴⁰	M/34	Arteriovenous fistula	Pain and swelling of ankle	Surgical resection	NR
Ahn, 2013 ⁴¹	F/39	Cephalic vein	Forearm nodule	Surgical resection	NR
Taguchi, 2013 ⁴²	M/53	Forearm vein	Painful forearm nodule	Surgical resection	NR
Umari, 2013 ⁴³	M/50	Renal vein	Asymptomatic — investigation benign prostate hyperplasia	Total nephrectomy	No recurrence at 9 mo
Risio, 2013 ⁴⁴	F/55	Adrenal gland vein	Abdominal discomfort, anorexia, and nausea	Right adrenalectomy	Uneventful No follow up mentioned
Cera, 2014 ⁴⁵	M/51	Internal jugular vein	Incidental finding	Surgical resection	Graft patency at 1 mo
Nguyen, 2014 ⁴⁶	F/79	Azygos vein	Right pleural effusion	Surgical resection	NR
Matsuzaki, 2016 ⁴⁷	M/73	Right subclavian vein	Right upper arm oedema	Surgical resection	No recurrence
Gameiro, 2016 ⁴⁸	M/54	Penile corpus spongiosum	Nodule of penal coronal sulcus	Partial biopsy	Regression within 2 weeks No recurrence at 6 mo
Loftus, 2017 ²	M/51	Subcutaneous vein	Forearm nodule	Surgical resection	No recurrence at 10 mo
Bongiolatti, 2018 ⁴⁹	F/32	Left subclavian vein	Left arm and face oedema	Surgical resection	No recurrence at 1 y

Continued

Table 1-continued

Authors, date	Sex/age	Anatomical location	Clinical data	Therapy	Outcome data
Reimold, 2019 ⁵⁰	F/78	Right renal vein	Asymptomatic, routine check up	Total nephrectomy	NR
Woo, 2019 ⁵¹	M/31	Jugular vein	Neck mass	Surgical resection	No recurrence at 6 mo

NR = not reported; y = years; F = female; M = male; mo = months.

* References 9–51 are listed in [Supplementary material](#).

items of all relevant articles were scanned for other potentially relevant studies. Full texts of all relevant articles were obtained and reviewed for relevance. The reference lists of each article were scanned for other potentially relevant studies. The systematic search identified 43 full text English studies, one full text German study, and one full text Korean study, including 39 single case reports, two case series, and four reviews, corresponding to a total of 64 cases.

Ten cases (15.6%) were symptomatic, with symptoms such as abdominal pain, limb oedema, or pleural effusion, six (9.4%) were incidental findings after routine examinations, and 48 (75%) presented with nodules, pain free or not. The male/female ratio was 10:9 and the median age was 44 years old (interquartile range [IQR] 35, 51).

The treatment was not reported in two cases (3.1%). Total resection was the chosen treatment for 61 cases (98.3%), only one case (1.7%) had a biopsy and regressed spontaneously. Follow up was reported in 22 cases and all had no recurrence, from two months (see reference³⁰ in [Supplementary material](#)) to eight years of follow up.⁴

DISCUSSION

Primary SVC tumours are rare. Three cases of epithelioid haemangio-endothelioma of the SVC and 12 leiomyosarcomas have been reported in the literature.^{2,3} No cases of SVC ILCH were reported.

LCH was described for the first time in 1979.⁴ ILCH is a rare type of LCH as, according to the present literature review, only 64 cases have been reported to date (Table 1). This benign vascular tumour is derived from endothelial cells, characterised by an anarchic capillary proliferation in a fibromyxoid oedematous stroma, with endothelial markers such as CD31 and/or CD34, or smooth muscle alpha-actin. Differential diagnoses of ILCH are other intraluminal lesions such as venous thrombosis, papillary endothelial hyperplasia, intravenous atypical vascular proliferation, histiocytoid haemangioma, and angiosarcoma.¹

Despite being a benign tumour, surgical removal of ILCH is the first choice approach to make a histological diagnosis and to prevent tumour related complications, such as SVC occlusion or thrombosis, local compression and pulmonary embolisation. As SVC resection is not a frequent procedure, the replacement strategy remains controversial. The main risk is graft thrombosis, occurring between one and five months after implantation.^{5–7} Long term patency rates can vary depending on graft material, length, or shape (ePTFE or a biological graft like bovine pericardium). In 2019, Maurizi et al. reported the results of SVC reconstruction with either bovine pericardial conduit (12 cases) or ePTFE grafts (13

cases). In their experience, follow up showed no statistical difference in terms of graft patency.

In this case, an ePTFE vascular graft was chosen. As shown in Fig. 2, the graft was L shaped. A left brachiobasilic fistula was created to increase blood flow and prevent graft thrombosis.

Despite the excellent long term prognosis of the benign characteristics of the tumour, immediate post-operative anticoagulant treatment and close follow up with DUS of the graft will be mandatory to assess graft patency.⁸

Conclusion

This case reports successful management of *en bloc* resection of an SVC ILCH associated with SVC prosthetic reconstruction.

APPENDIX A. SUPPLEMENTARY DATA

Supplementary data to this article can be found online at <https://doi.org/10.1016/j.ejvsf.2020.12.021>.

REFERENCES

- Lahon B, Fabre D, De Montpreville V, Darteville P. Epithelioid haemangioendothelioma of the superior vena cava. *Interact Cardiovasc Thorac Surg* 2012;**15**:186–7.
- Loftus WK, Spurrier AJ, Voyvodic F, Neuhaus SJ, James CL. Intravenous lobular capillary haemangioma (pyogenic granuloma): a case report and a review of imaging findings as reported in the literature. *J Med Imaging Radiat Oncol* 2018;**62**:217–23.
- de Chaumont A, Pierret C, de Kerangal X, Le Moulec S, Laborde F. Leiomyosarcoma of the superior vena cava. *Ann Thorac Surg* 2014;**98**:725–7.
- Cooper PH, McAllister HA, Helwig EB. Intravenous pyogenic granuloma. A study of 18 cases. *Am J Surg Pathol* 1979;**3**:221–8.
- Maurizi G, Poggi C, D'Andrilli A, Vanni C, Ciccone AM, Ibrahim M, et al. Superior vena cava replacement for thymic malignancies. *Ann Thorac Surg* 2019;**107**:386–92.
- Lee GD, Kim HR, Choi SH, Kim YH, Kim DK, Park SI. Prosthetic graft interposition of the brachiocephalic veins or superior vena cava combined with resection of malignant tumours: graft patency and risk factors for graft occlusion. *J Thorac Dis* 2016;**8**:61–7.
- Nakano T, Endo S, Kanai Y, Otani S, Tsubochi H, Yamamoto S, et al. Surgical outcomes after superior vena cava reconstruction with expanded polytetrafluoroethylene grafts. *Ann Thorac Cardiovasc Surg* 2014;**20**:310–5.
- Oizumi H, Suzuki K, Banno T, Matsunaga T, Oh S, Takamochi K. Patency of grafts after total resection and reconstruction of the superior vena cava for thoracic malignancy. *Surg Today* 2016;**46**:1421–6.