

Leiomyosarcoma of the inferior vena cava: 1 case

B. Bancel, A. Rode, C. Ducerf

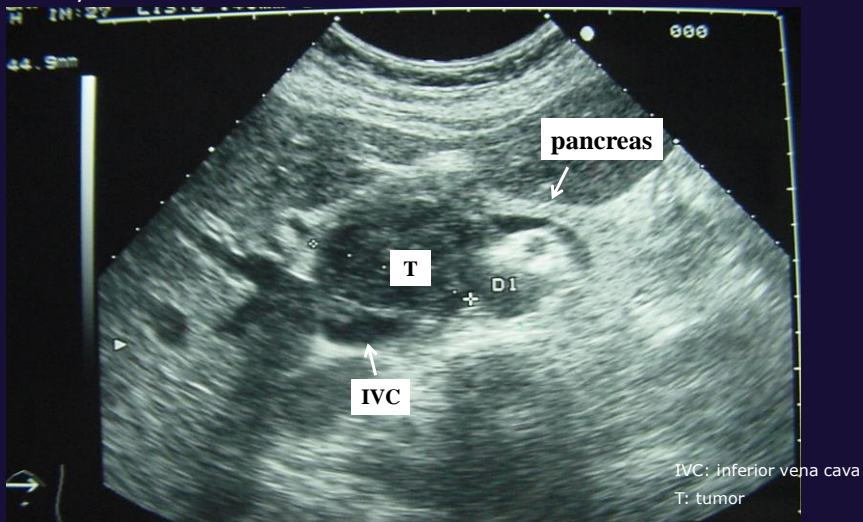
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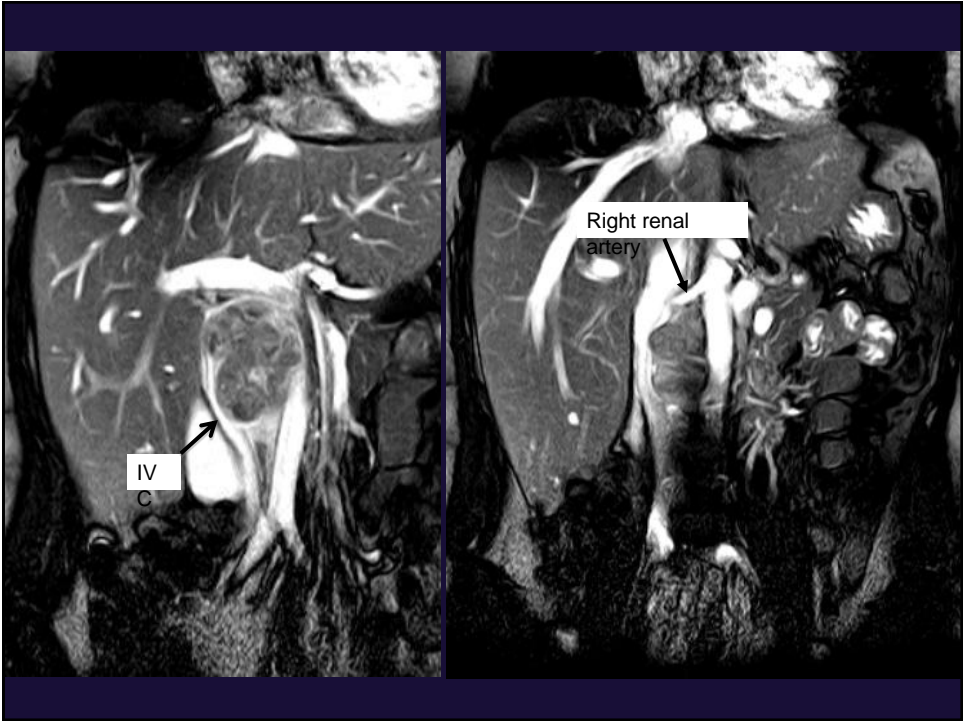


Bucharest
Nov 2011

Case report

- 34 yr-old woman, no antecedent
- Sept 2004: Abdominal upper right quadrant pain: gallstones?
- Physical examination normal





➤ Oct 2004:

Surgical resection of the tumor including the anterior wall of the IVC and a short segment of the left renal vein

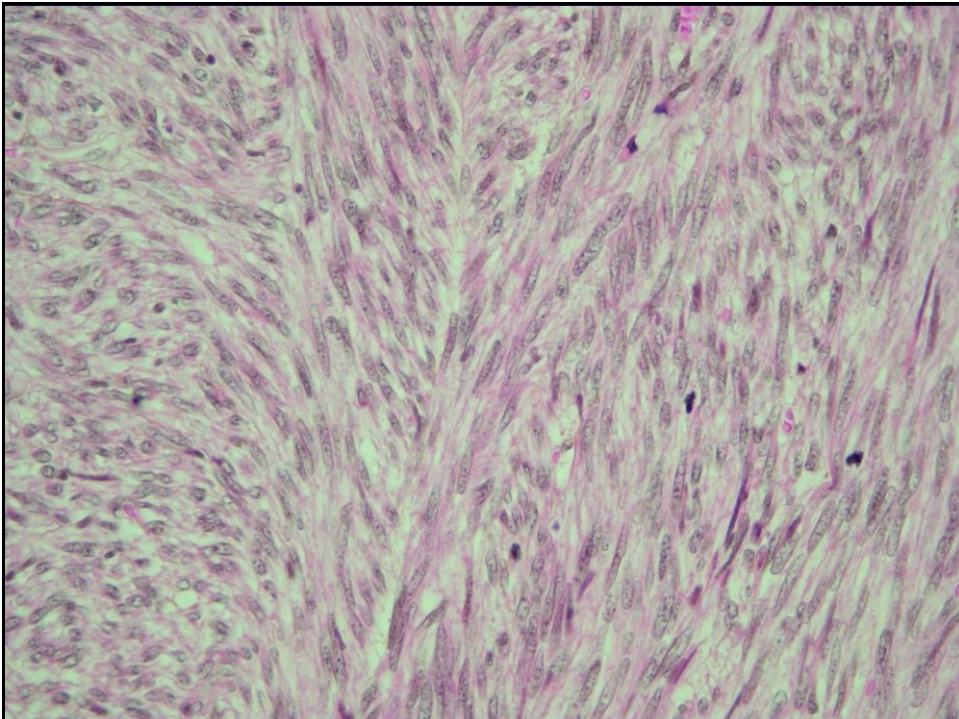
Macroscopically complete

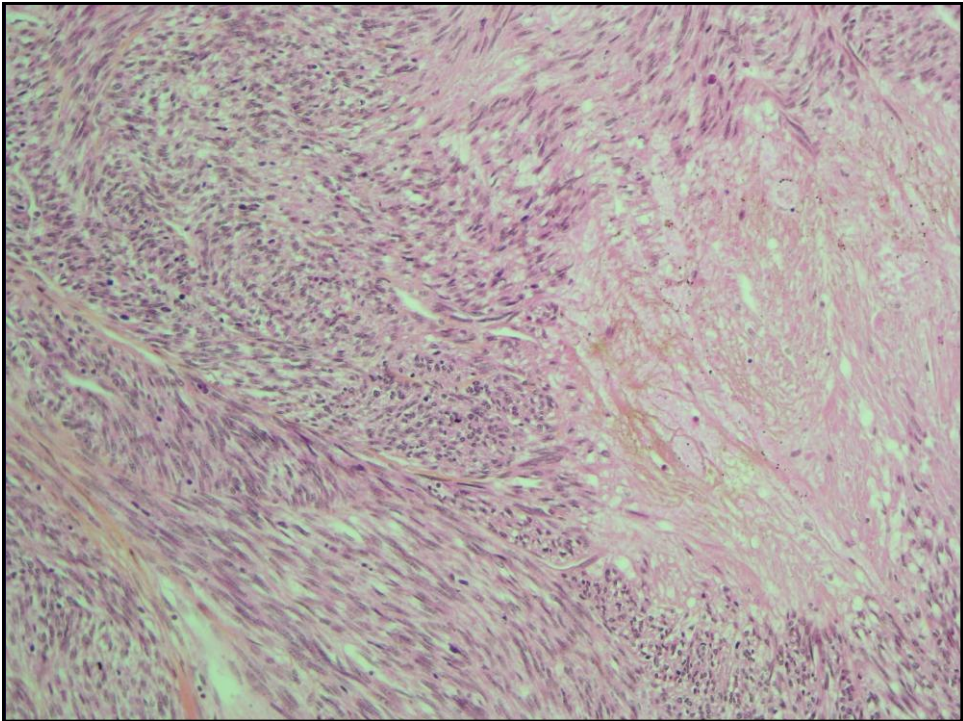
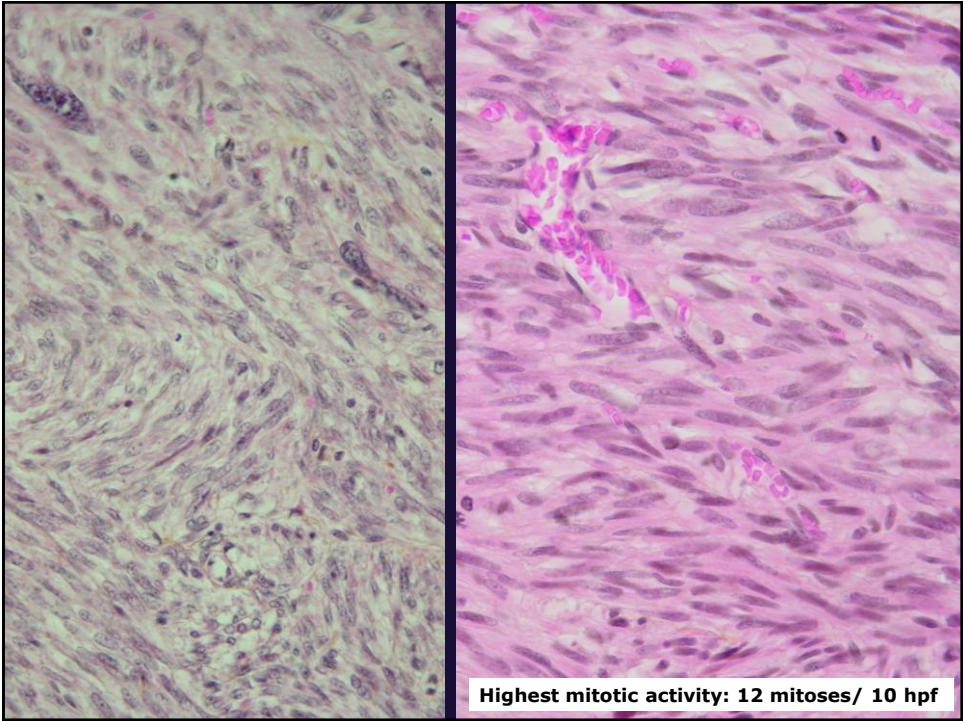
➤ Pathology:

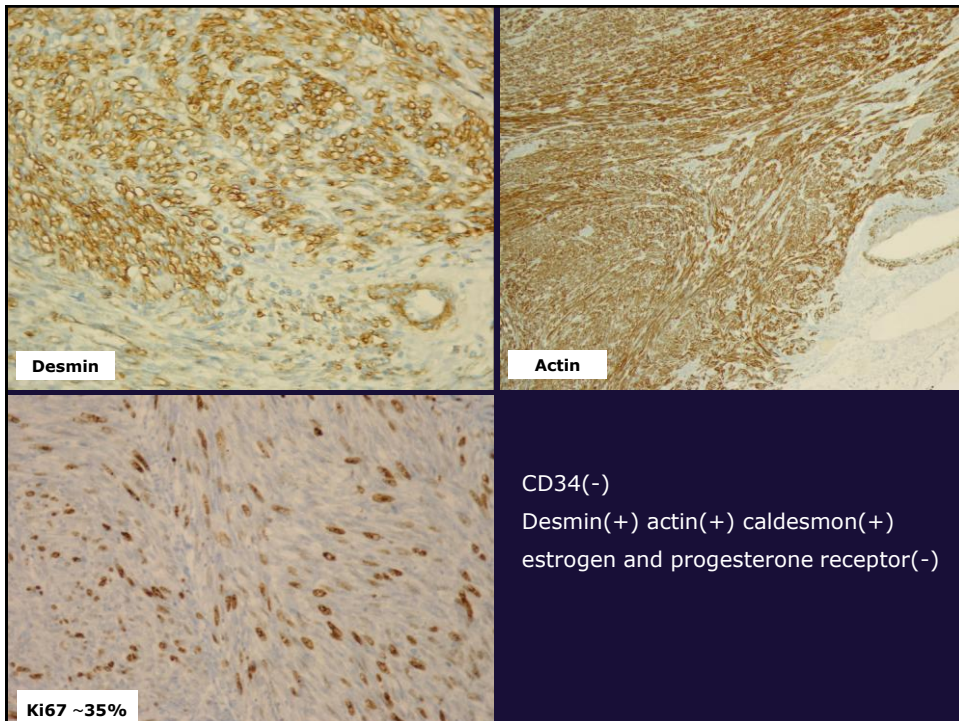
A lobulated pseudoencapsulated tumor (5,5 x 4,2 x 4 cm)

Whorled whitish cut surface; reddish-purple foci of hemorrhage; no macroscopic necrosis

No intraluminal tumor







Diagnosis:

Inferior vena cava leiomyosarcoma

lower level I¹

Histologic grading 2 (FNCLCC)

Differentiation: score 1

Mitosis: score 2

Necrosis: score 1

1. Segments of the IVC

lower level I (below the renal veins to the origin of the IVC)

middle level II (below the hepatic veins to the renal veins)

upper level III (from the right atrium to the hepatic veins)

Tumor grading (FNCLCC criteria)

- **Tumor differentiation**

Score 1: sarcoma closely resembles normal adult tissue (e.g., low grade leiomyosarcoma)

Score 2: sarcoma for which histological typing is certain (e.g., myxoid liposarcoma)

Score 3: embryonal and undifferentiated sarcoma

- **Mitotic count**

Score 1: 0-9 mitosis per 10 HPF,

Score 2: 10-19 mitosis per 10 HPF,

Score 3: ≥ 20 mitosis per 10 HPF,

- **Tumour necrosis**

Score 0: no necrosis,

Score 1: $< 50\%$ tumour necrosis,

Score 3: $\geq 50\%$ tumour necrosis,

Histological grade

Grade 1: total score 2,3,

Grade 2: total score 4,5,

Grade 3: total score 6,7,8,

- Post operative radiotherapy

51 months (jan 2009) multiple pulmonary metastasis

Chemotherapy

- 84 months patient alive with disease

Discussion

- **Primary vascular tumors:** rare (300 to 400 reported cases)
 - Most are sarcomas
 - Most sarcomas are leiomyosarcomas
- Up to **50% arise in the inferior vena cava (IVC)**
 - ~ 0.5% of adult soft tissue sarcomas

	Mingoli et al. 1996¹ International registry of IVC leiomyosarcoma up to 1993	Laskin et al. 2010² AFIP between 1976 and 2008	Mann et al. 2010³ A 2-institution analysis from 1984 to 2009
Number of patients	218*	40*	17
Female (%)	82.6%	77%	71%
Median age (y)/(range)	54 (15-85)	58 (22-86)	48 (29-79)
Symptoms	Abdominal pain, mass, lower limb's edema, Budd's-Chiari sd		
Duration (months)	Mean 7.4 ± 0.7 months	Median 12 (1-84) months	/
<p>• including 13 examples earlier reported from the AFIP²</p> <p>1. Mingoli A, et al. <i>Anticancer Res</i> 1996;16:3201-5. 2. Laskin WB, et al. <i>Am J Surg Pathol</i> 2010;34:873-81. 3. Mann G. N., et al. <i>Surgery</i> in press, 2010.</p>			

	Mingoli et al. 1996 International registry of IVC leiomyosarcoma; N=218	Laskin et al. 2010 AFIP between 1976 and 2008; N=40	Mann et al. 2010 A 2-institution analysis; N=17
Mean size (range)	10 cm (3-30)	8,5 cm (3,5-15)	11,5 cm (5-21)
Segment location	(Resected 134)	Classified according to most superior extent	
Lower I	28%	13%	53%
Middle II	33%	74%	35%
Upper III	14%	13%	12%
Multiple		42%	/
Growth			
Extraluminal	76%	60% (All cases level 1 and/or 2)	/
Intraluminal	29%	50% (main component 22%)	/
Adjacent organ involvement	5.9%	42% Microscopic 10%	44%

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Grading	(Differentiation)	(FNCLCC)	(FNCLCC)
Grade 1	22%	27.5%	14%
Grade 2	25.5%	52.5%	47%
Grade 3	24%	12.5%	41%
Mitotic index		1-48/ 10 hpf (mean, 12; median, 11)	
All non-hormonally responsive retro peritoneal smooth muscle tumor exhibiting cytological atypia and any level of mitotic activity is diagnosed as leiomyosarcoma ¹			
Miettinen M, Fetsch JF. Histopathol. 2006;48:97-105.			

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Surgical resection			
Curative	61,5%	17.6%	71%
Palliative	12%	33%	
Inoperable	26,6%	12.5%	
Local/ Metastatic spread		52%	58%
Disease-free survival After curative surgery			
5-year	31%	28%	37%
10-year		6%	/
Overall survival After surgery			
5-year	49.4%	34%	54%
10-year	29.5%	13%	/

Poor prognostic factors		
	Mingoli et al. 1996 International registry of IVC leiomyosarcoma; N=218	Laskin et al. 2010 AFIP between 1976 and 2008; N=40
Size ≤5 >5 >10 cm		No correlation
FNCLCC grade	Discussed	No correlation
Level III	/	Correlation
Budd-Chiari syndrome	/	Correlation
Right atrial involvement	/	Correlation
Intraluminal growth	/	Correlation
Residual postoperative tumor	Correlation	Correlation

Conclusion

IVC sarcoma

A rare sarcoma with a poor prognosis. The most common sarcoma affecting the venous system

Importance of the involved venous level

Histological grade - mitotic count - tumor necrosis do not predict outcome

Importance of complete surgical resection

No consensus exists regarding adjuvant treatment