Recurrent Thrombosis and Pulmonary Aneurysm

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Recurrent Thrombosis and Pulmonary Aneurysm

- **45 ♂non-smoker with recurrent hemoptysis**
- Relevant MHx:
  - 2003 **DVT (L) lower limb** in →
  - 2004 **DVT relapse (R) lower limb** → continue warfarin (INR =2-3)
  - 2008 Abdominal pain → Thrombosis of **Inferior vena cava** (from the common iliac vein up to 1.5 cm above the renal veins) + **partial thrombosis of portal and superior mesenteric vein + PE + stenosis at the origin of abdominal aorta** → Extensive collateral network (see picture). Thrombophilia work up (-) → warfarin (INR =2-3)
  - 2012 Hemoptysis (2 cupfulls) = new **PE** → stop warfarin→ fondaparinux (Arixtra).→rivaroxaban (Xarelto).
  - 2013 Hemoptysis (1 cupfull)→ CTPA → **Aneurysm of (L) pulmonary aneurysm (d = 3.2cm)**

+ Recurrent oral and scrotum ulcers
+ Severe acneiform rash

→ **Adamantiades-Behcet dis.**

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- Treatment plan:
  1. GC+ iv CY for 2 years
  2. Stop anticoagulation

- Disease course:
  - ⬇ in the frequency and severity of hemoptysis & no new thrombosis
  - Repeated CTPAs = progressive decrease in the diameter of PAA → unfortunately misinterpretation due to mural thrombus and inappropriate technique in the measurement
  - Switch to infliximab but just before initiation → **massive pulmonary hemorrhage due to PAA rupture**

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- **Question: Alternative management plan esp. for PAA?**

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CLINICAL INVESTIGATION

Effectiveness and Safety of Endovascular Aneurysm Treatment in Patients With Vasculo-Behçet Disease

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Purpose: To compare long-term outcomes of endovascular and surgical treatment of arterial aneurysms in patients with vasculo-Behçet disease (VBD).
Methods: The medical records of 912 patients with Behçet disease who were seen between May 1995 and January 2007 were reviewed. Among them, 34 (3.7%) patients with 39 non-cerebral aneurysmal lesions were diagnosed with VBD. Between February 1998 and November 2006, 16 VBD patients (14 men; mean age 39.2±9.2 years, range 25–63) were treated for 20 arterial aneurysms with an endovascular technique (stent-graft and/or coil embolization). All patients received immunosuppressive therapy (prednisolone 60 mg/d) before endovascular therapy to induce remission. From February 1993 to January 2007, 8 arterial aneurysms in 7 patients (all men; mean age 33.0±7.9 years, range 25–51) were treated with surgical graft interposition over 31.5±23.2 months.
Results: The endovascular procedure was successful in all lesions. The mean follow-up was 47.6±41.8 months, during which 4 complications in 3 patients (3/16, 18.8%) occurred (2 occluded stent-grafts and 2 access site pseudoaneurysm). There were no deaths. The cumulative primary patency rate in the endovascular group was 89% at 24 months. In the 7 patients undergoing graft interposition for 8 arterial aneurysms, 3 (42.9%) events occurred in follow-up: 2 recurrent pseudoaneurysms and 1 aneurysm-related death.
Conclusion: In Behçet disease, aneurysm treatment is performed whenever possible because of the high risk of rupture. Endovascular treatment of arterial aneurysms was effective and safe, with an acceptable vascular complication rate and excellent patency of the treated site.
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- Background:
  - High mortality rate = 50% in 2 years from hemoptysis onset
  - Open surgical management → high incidence of anastomotic dehiscence (≤ 50% of pseudoaneurysms at anastomotic sites[1])
  - Endovascular stent-grafts → long term outcomes?
- 912 pts with BD → 34 pts with VBD with 39 non-cerebral aneurysms

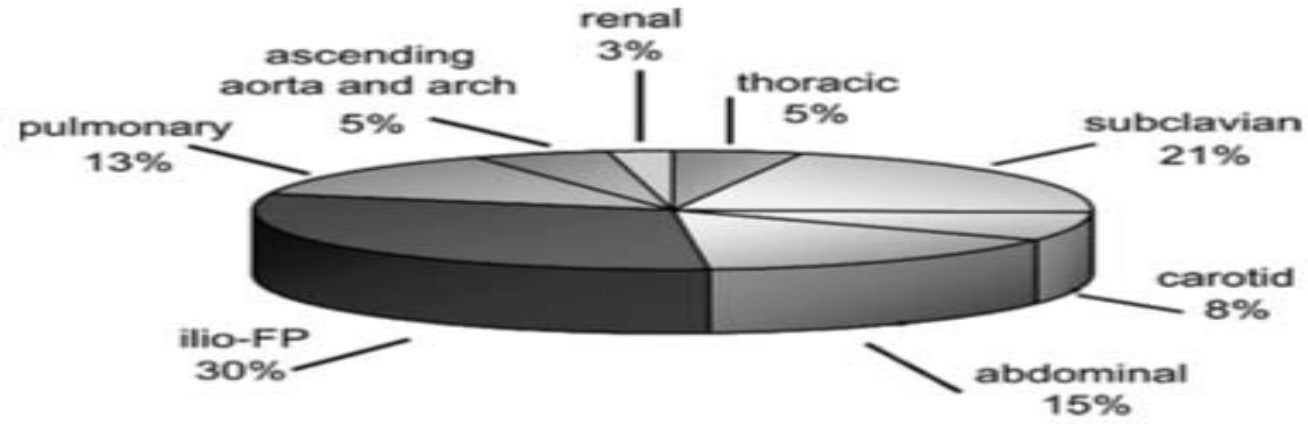

Figure 1 ◆ Distribution of 39 non-cerebral aneurysmal lesions in 34 (3.72%) of 912 patients who were diagnosed with VBD. FP: femoropopliteal.

[1]Freyrie A,et al. Arch Surg. 1993;17:762–767

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- 16 pts undergoing endovascular treatment for 20 arterial aneurysms (**PAA not included**)
- Mean follow-up = **47.6 ± 41.8 mo**
- 4 complications in 3 patients (3/16, **18.8%**)
- 2 stent- graft occlusions and 2 pseudoaneurysms at the puncture site.

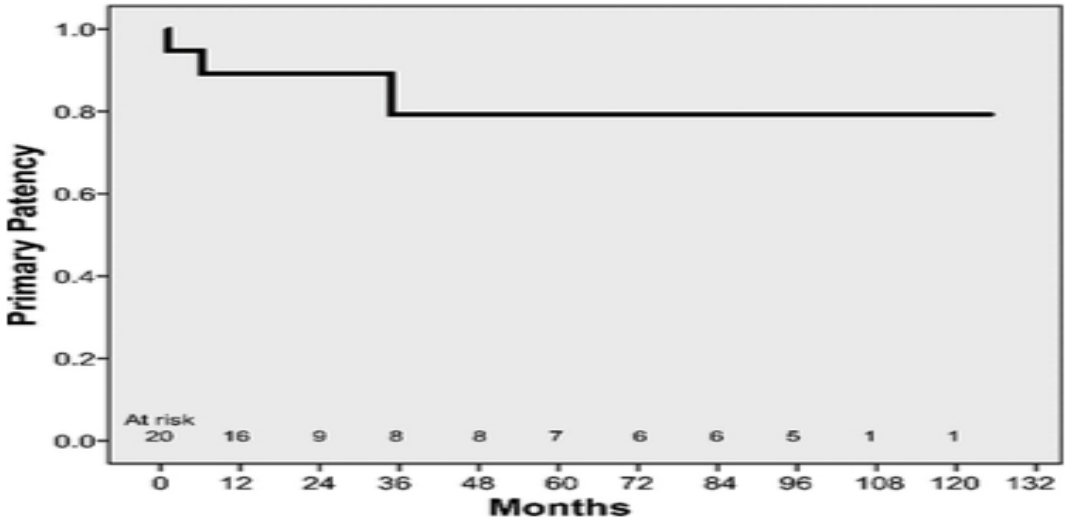

Figure 3 ◆ Kaplan-Meier graph depicting the cumulative primary patency for an endovascular procedure per lesion (n=20). The standard error was <0.10 up to 24 months.

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- **Surgical interventions for PAA in BD*:**
  a. Open surgery = 7 cases (incl. ligation, endoaneurysmorrhaphy, excision)
  b. PAA embolisation = 11 cases
- BUT NO LONG-TERM DATA available

- ISs pre- AND post- intervention warranted

Take Home message:
PAA Px = GC+CYC ± INF/ADA
Endovascular interventions should be discussed due to high associated mortality

*Yuan SM ,et al. *Clin Exp Rheumatol 2014; 32 (Suppl. 84)*