

Localized intravascular coagulopathy in venous malformations

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Localized intravascular coagulopathy (LIC)

- Subset VM exhibit localized LIC
 - ➔ **pain, thrombosis & excessive bleeding** during surgical procedures
- LIC can progress to **disseminated intravascular coagulopathy (DIC)** and life-threatening hemorrhage even when coagulation parameters (PT, INR, PTT) are normal

Coagulation disorders in venous malformation

LIC

D-dimer $>1,000$ ng/mL a/o

fibrinogen <200 mg/dL

incidence between 42% and 88%

lesion size ($P < 0.001$), presence of phleboliths ($P = 0.005$)

DIC

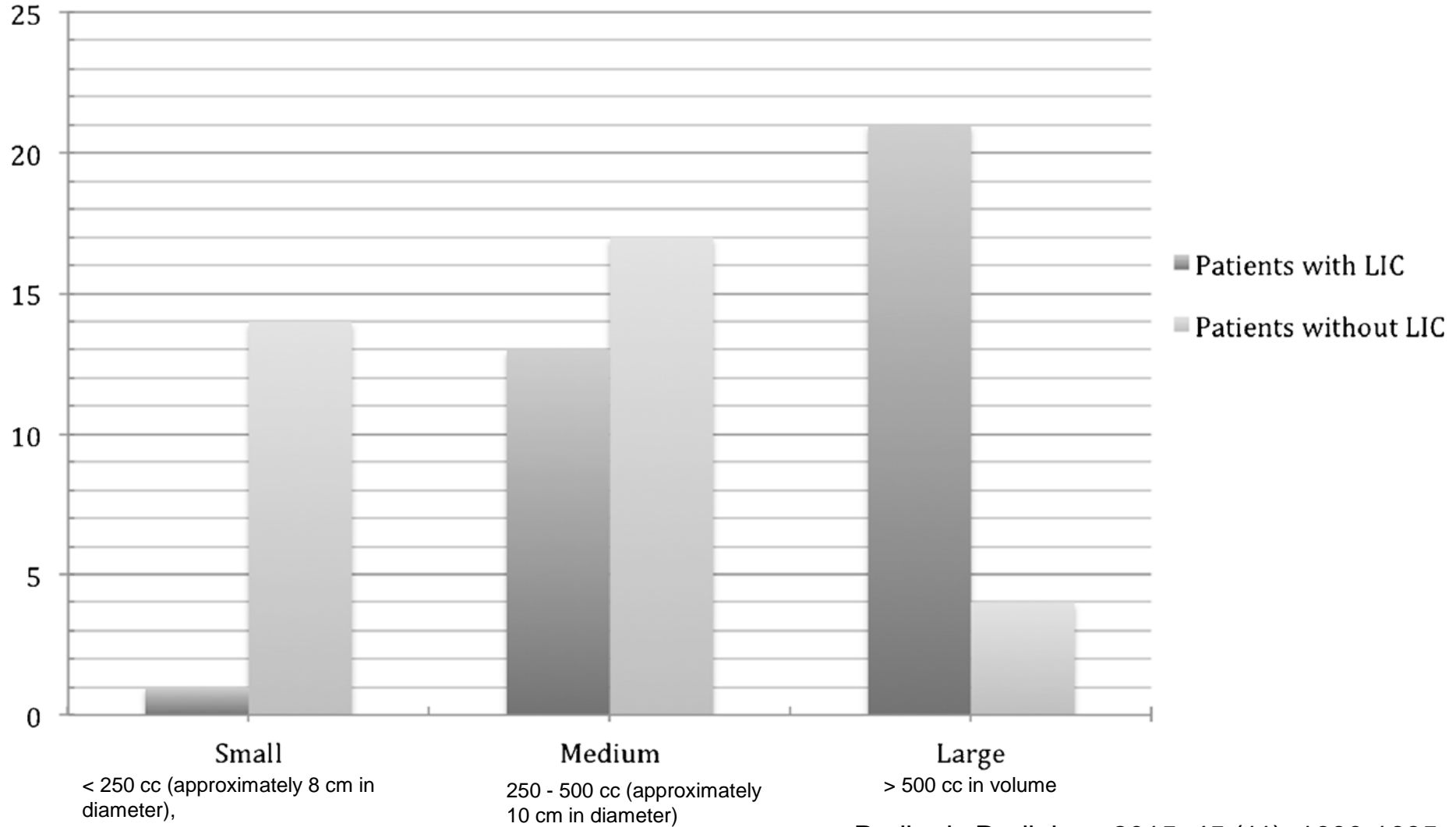
conversion of LIC to DIC

consumption of platelets and coagulation factors

increase in PT & decrease in FV earliest signs

bleeding

Localized intravascular coagulopathy (LIC) and lesion size



Pediatric Radiology 2015, 45 (11), 1690-1695

Association of LIC with venous malformations

- Prospective, consecutive series

140 patients with VM

59 (42%) high D-dimer levels

6 (4.3%) low fibrinogen (85-176 mg/dL), 1 (0.7%) < 100 mg/dL

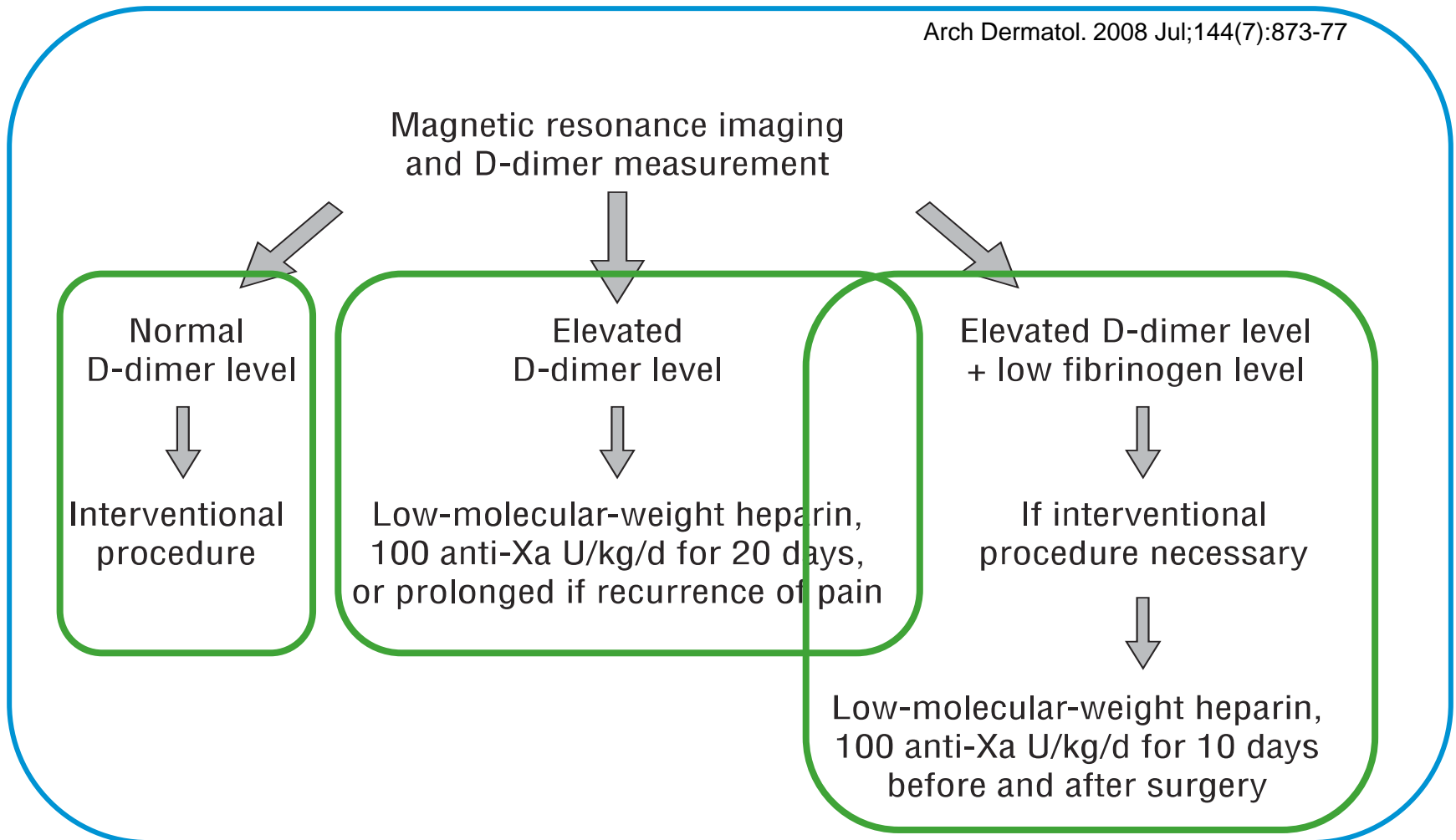
large size (>10 cm²) sig. associated with positive D-dimer test, independently of localization

59% large VMs (>10 cm²) vs 23% small VMs with high D-dimer ($p < .001$)

none exhibited PE

Management algorithm

Arch Dermatol. 2008 Jul;144(7):873-77



- ASS low efficacy (vs. Kasabach-Merritt phenomenon, platelets not involved in LIC)
- OAC decrease coagulation factors, not sufficient to prevent thrombin formation in LIC

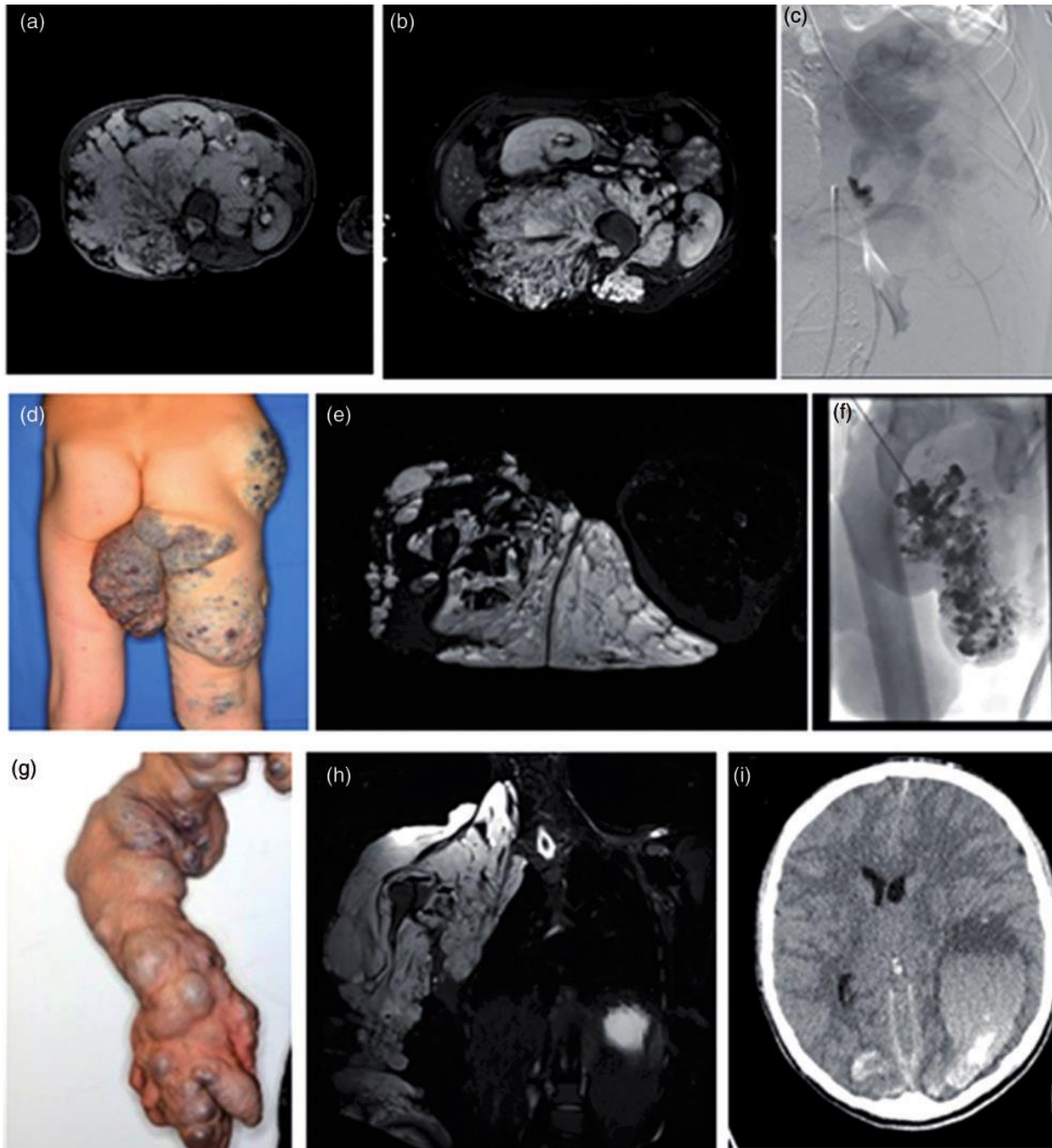
Sclerotherapy complications

N=127, sclerotherapy for venous malformation

4 (3.1%) severe complications, all related to coagulopathy

| Complication | Management | N | Grade |
|---|--|---|-------|
| Skin damage | Not necessitated | 8 | I |
| Pain or swelling or both for > 2 weeks | Analgesics and/or compression garment | 6 | I |
| Thrombophlebitis outside the malformation | Not necessitated | 3 | I |
| Unusual pain and transient sensory loss | Analgesics | 1 | I |
| Unusual swelling and loss of range of joint motion | Physiotherapy | 1 | I |
| Pain, swelling, skin damage, and loss of range of motion | Analgesics, physiotherapy | 1 | I |
| Digital loss of range of joint motion | Physiotherapy | 1 | I |
| Skin damage, infected | Antibiotics, per oral | 4 | II |
| Bleeding at injection site, skin damage and sepsis | Corticosteroids, intermediate care-unit treatment, i.v. antibiotics, | 1 | II |
| Intra-abdominal bleeding, mild | Blood transfusion (one unit), | 1 | II |
| Deep vein thrombosis at ankle level | LMWH medication | 1 | II |
| Skin damage | Skin graft under local anesthesia | 2 | IIIa |
| Intra-abdominal bleeding (6 liters) | Blood transfusion, intra-arterial embolisation, | 1 | IV |
| Bleeding at injection site, sepsis, abscess, worsening of DIC | Transfusions of blood and coagulation factors, i.v. antibiotics, abscess drainage, ICU treatment and prolonged hospitalization | 1 | IV |
| Bleeding at injection site, skin damage and sepsis | Intermediate care-unit treatment, i.v. antibiotics, long hospitalization, | 1 | IV |
| Intracerebral hemorrhage and exitus | | 1 | IV |
| Intra-abdominal bleeding (6 liters) | Blood transfusion, intra-arterial embolisation, ICU treatment | 1 | IV |
| Intracerebral hemorrhage and exitus | | 1 | V |

Phlebology 2015, October, 1-15

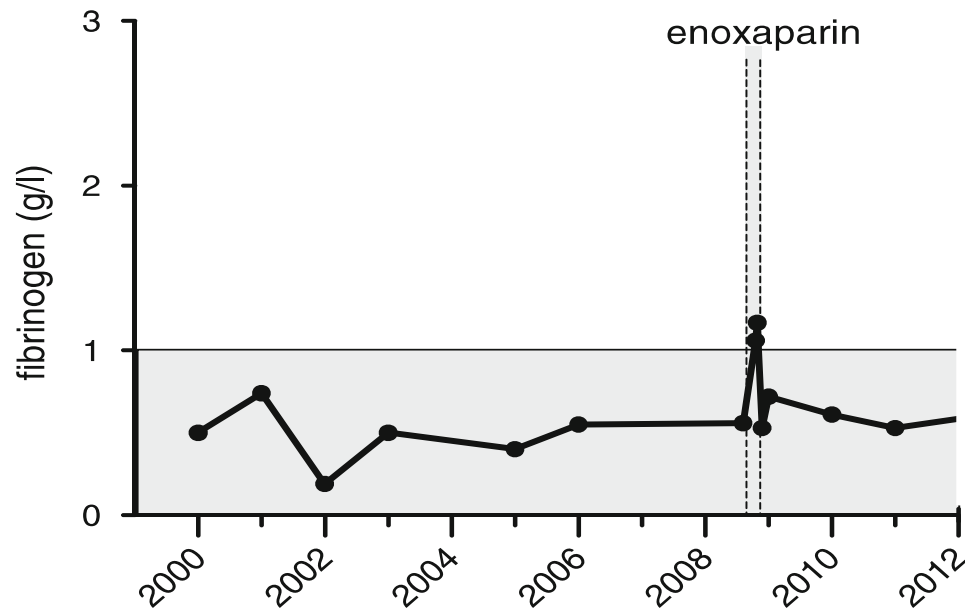


Phlebology 2015, October, 1-15

Rivaroxaban for treatment of LIC in venous malformation (case report)

LMWH recommended in LIC: invasive procedures, active bleeding, very low fibrinogen levels (<0.5 – 1.0 g/L) associated with a bleeding diathesis

Evolution of markers of intravascular coagulation



J Thromb Thrombolysis (2014) 38:121–123; Blood Coagulation and Fibrinolysis 2015, 26:00–00

Conclusion

Higher VM severity scores associated with more severe LIC

Sclerotherapy, surgery or pregnancy can trigger conversion to DIC, with bleeding related to factor consumption

Prophylactic dose of LMWH (100 IE/kg) recommended in LIC with recurrent pain and before invasive treatment

Therapeutic dose of LMWH is recommended in LIC with DVT

NOACs seem to have comparable efficacy and might become an ideal oral treatment alternative to LMWH

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