SPECTRUM INDIA 2024
MUMBAI VASCULAR FOUNDATION
8TH - 10TH NOVEMBER, 2024 | GRAND HYATT MUMBAI

Utilization of Non-Conventional Access in the Management of Consumptive Coagulopathy in Infants with Vascular Tumour

Registration number: 242

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Patient 1 – Clinical details

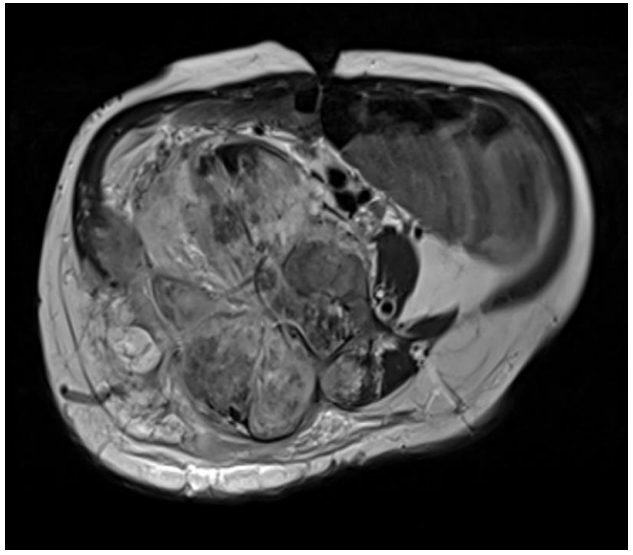
Presentation

- 6-months-old child
- Swelling - lumbar region
- ***On & off ecchymotic patches over the back and abdomen***

Lab values

- ***Platelets 8000/mm3***
- Hemoglobin 8.2 gm%
- ***Fibrinogen 52 mg/dl***
- ***D dimer 16053 ng/ml***
- PT/INR 18.7/1.36

Imaging



- Large, ill-defined, solid lesion in abdomen and pelvis on the right side, multi-compartmental
- ***Intense enhancement +***

Biopsy

- Correction of bleeding parameters – Platelets, cryoprecipitate, FFP
- General anesthesia
- USG guided, 17G-18G co-axial, automatic

Kaposiform hemangioendothelioma + Kasabach Merritt syndrome

Patient 1 – Clinical details

Medical management

- ❖ **Syp. Prednisolone 5 mg BD**
- ❖ **Inj. Vincristine 0.5 mg iv weekly**
- ❖ **Syp. Propranolol 25mg BD**

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Sub-optimal response

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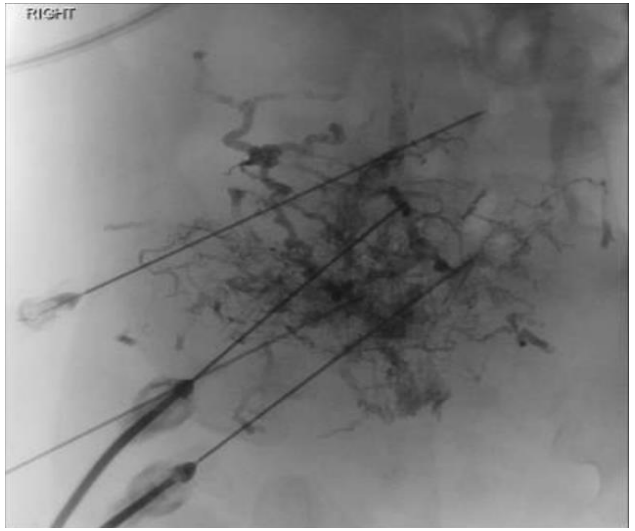
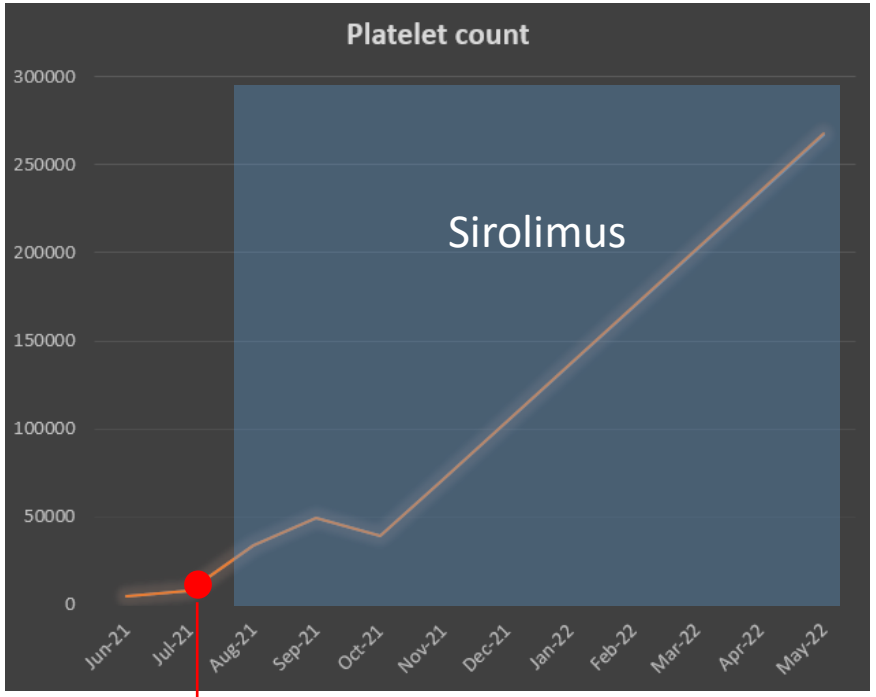
Embolization + Sirolimus 0.5 mg

Endovascular management



- ❖ **Common femoral artery diameter – 1.5 mm**
- ❖ Opted for ***direct tumor puncture***
- ❖ 22G lumbar puncture needles - 17% glue injection

Follow-up



Day of embolization

	Jun-21	Jul-21	Aug-21	Sep-21	Oct-21	Nov-21	May-22
Fibrinogen	49	26	108	50	209	165	551
Platelet count	5000	8000	34000	49000	39000	71000	267000

After 3 years

**** Reduction in size of lesion***
**** Ecchymosis settled***
**** Normal platelet c.***

Patient 2 – Clinical details

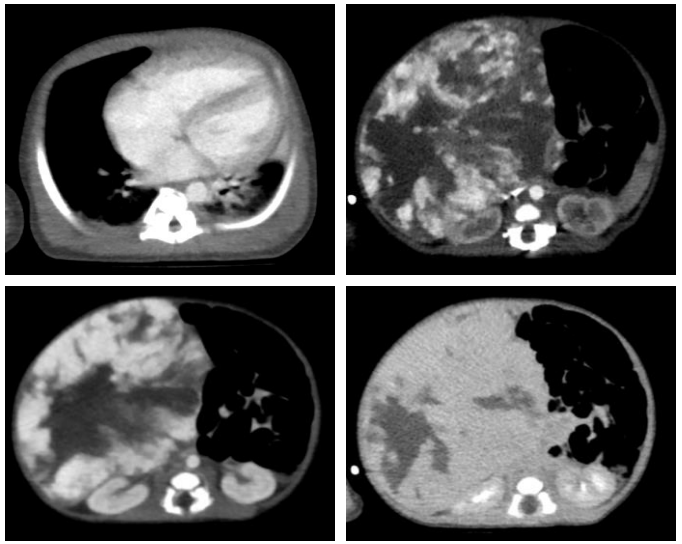
Presentation

- ***3-days-old child***
- Late preterm, LSCS
- ***Respiratory distress (on CPAP) & abdominal distension***
- ECHO – Significant patent ductus arteriosus

Lab values

- ***Platelets 50000/mm3***
- ***Fibrinogen 99 mg/dl***
- ***D dimer 62890 ng/ml***
- PT/INR 20.6/1.57

Imaging



- Large hepatic mass – **peripheral globular enhancement with centripetal fill-in pattern**

Giant hepatic hemangioma + consumptive coagulopathy

Management

Started on oral prednisolone & propranolol

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On Day 3 – platelet and fibrinogen level started declining

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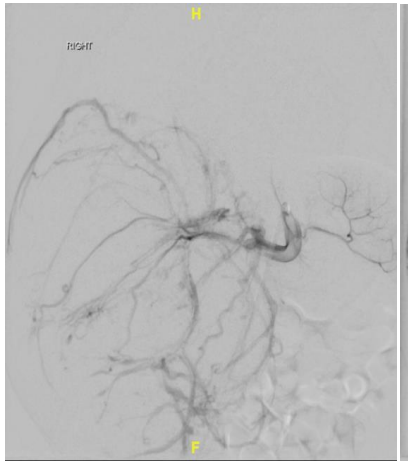
Cryoprecipitate 10 ml/kg
+
Embolization (treat coagulopathy & tumor down-sizing to combat respiratory distress)

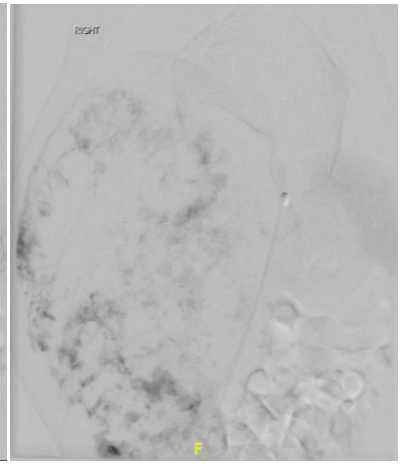
Patient 2 – Clinical details

Umbilical arterial access for embolization

Umbilical arterial line was secured surgically

UAC exchanged over glide wire to 4F glide cobra catheter





Hepatic arterial feeders selectively cannulated and embolized with PVA & gel foam

Cobra catheter exchanged to UAC

Follow-up

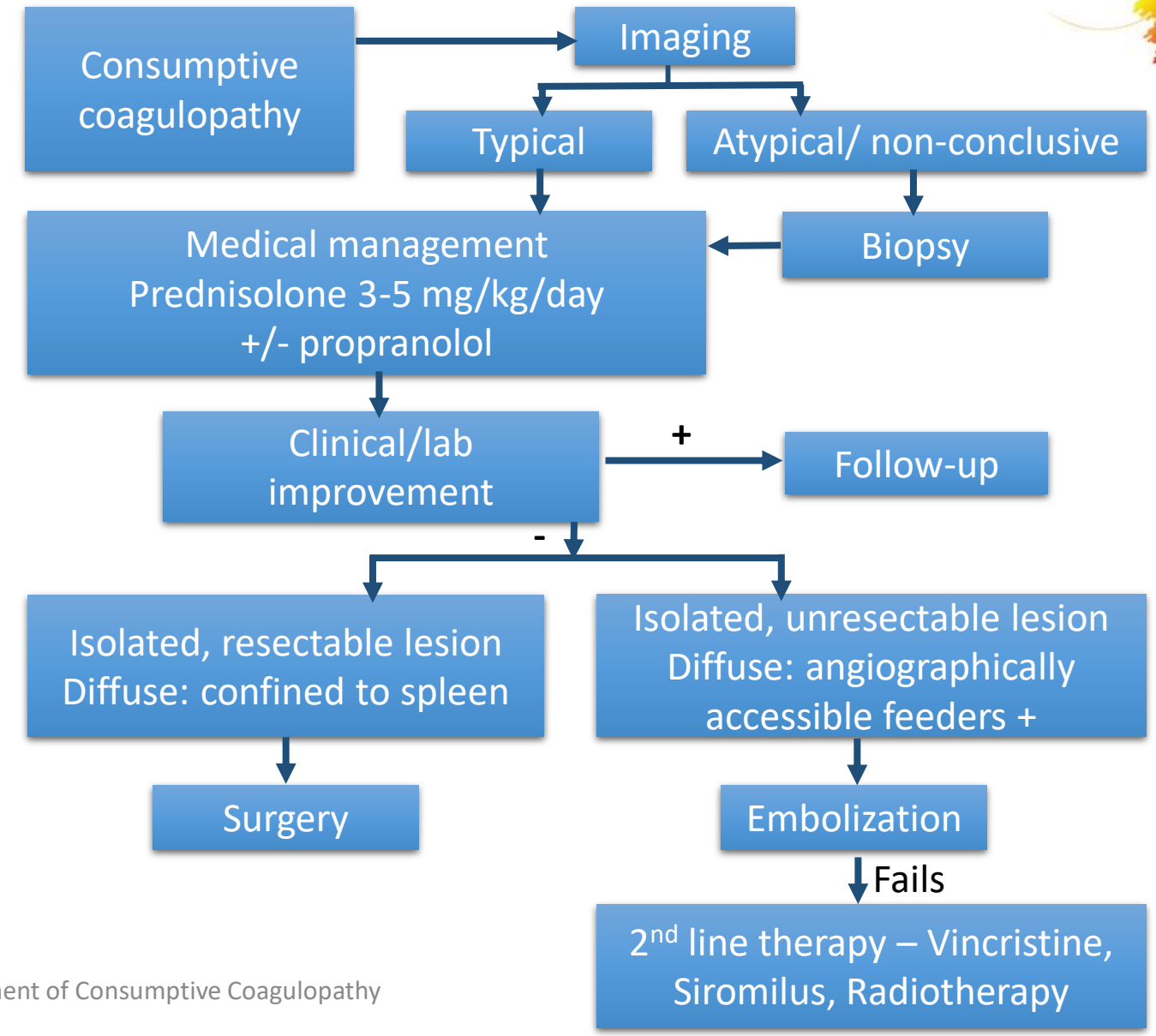
	Day 0	POD 2	POD 4	POD 20
Fibrinogen count	99	107	251	143
Platelet count	54000	90000	82000	59000

- ***Transient improvement in platelet count and fibrinogen***
- Child died on day 30 due to sepsis and worsening cardiac failure

Consumptive coagulopathy in vascular malformation

- Thrombocytopenia
- Hypofibrinogenemia
- Elevated D dimer
- Elevated PT/INR & APTT

Mild	Severe
Congenital hemangioma Venous or veno-lymphatic malformation Kaposiform lymphangiomatosis	Kaposiform hemangioendothelioma Tufted angioma ***(Kasabach Merritt syndrome)***



Various tumor accesses for embolization in infants

Various tumor access in neonate/ infants	*Advantages*	*Disadvantages*
Femoral arterial access	❖ Familiar with technique ❖ Accommodates relatively larger sheaths	❖ Smaller caliber in neonates ❖ Catheter associated thrombosis & ischemia
Umbilical arterial access	❖ Pre-existing umbilical artery catheter – simple exchange technique ❖ Femoral artery preservation for future interventions	❖ Availability of neonatologist for access ❖ Cannot be used after 5-7 days of life ❖ Risk of infection
Direct tumor puncture	❖ Easy to perform ❖ Avoids arterial access related complications	❖ Entire tumor coverage during embolization - challenging ❖ Risk of hemorrhage

Take-home points

- Consumptive coagulopathy in vascular tumor – rare, can cause life-threatening hemorrhage
- Prompt diagnosis and effective management are critical
- **Embolization – safe and effective, indicated when medical management fails or when response is slow (bridge therapy)**
- **Neonates – smaller diameter of common femoral artery – Umbilical arterial access or direct tumor puncture can be utilized**

Kindly scan the QR code below for references

thank you