

32nd World Pediatrics Conference

December 04-05, 2019 | Barcelona, Spain

Limb salvage in an extensive and complicated vascular lesion in an infant

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Background: Extensive vascular lesions can endanger the life of a child by their virtue of consumptive coagulopathy or cardiac failure. A conservative surgical approach is difficult and can be life-threatening due to uncontrolled bleeding. We report successful limb salvage in an infant despite an extensive and infected arterio-venous malformation of upper limb, complicated by severe consumptive coagulopathy.

Case: A term male, 3.2kgs, presented with a swelling over left arm detected antenatally. Antenatal scan- soft tissue swelling 10.8 x 6.8 x 5.4cm from left shoulder to elbow with heterogeneous cystic spaces and minimal vascularity. O/E- non compressible, no bruit heard.

Investigations: Doppler – hypoechoic lesion+ heterogeneous echotexture, numerous cystic spaces, weak color Doppler signal- S/O- Hemangioma MRI- lobulated soft tissue mass 17 x 16 x 9.5cm with an intensive enhancement, necrotic and hemorrhagic areas, involving flexor and extensor muscles of the arm. Supplied by branches of axillary and brachial arteries. Diagnosed as- Hemangioma, hence was started on propranolol 0.5 mg/kg/day. Presented at 2 months of age with severe anemia and consumptive coagulopathy refractory to medical therapy. At 4 months of age surgery became an imperative lifesaving mode of treatment.

Operative procedure: Planned for amputation/disarticulation, Axillary vessels identified and controlled, Tumor excised in toto, Flexor and extensor muscles preserved as much as possible, Radial nerve through the tumor was divided and re-anastomosed, Disarticulation was hence avoided, HPE suggestive of AVM, Arm and hand movements preserved at 10th follow-up.

Discussion: Vascular malformations are usually present at birth; they expand secondary to trauma, sepsis or hormonal modulation. They are less likely to cause consumptive coagulopathy but can destroy, distort or cause hypertrophy of the adjacent bone. They may shunt the blood leading to cardiac failure. Most of the vascular tumors and a few of vascular malformations can be managed conservatively. The indication for surgery is functional or aesthetic. When medical management fails, surgery is unavoidable.

Conclusion: A conservative limb salvage surgery is possible even in case of an extensive vascular anomaly. A good vascular control is a must to prevent life threatening hemorrhage in a conservative surgery.