Treatment of Complicated Vascular Lesions in Children

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Annotated Bibliography

Disclosures:
- I have no conflicts of interest
- Many of the therapies I discuss are off-label

This talk will review four challenging cases:

1) Ulceration in the setting of Infantile Hemangioma
2) Multifocal Infantile Hemangiomas
3) The clinical spectrum of Tufted Angioma
4) Capillary Malformation-Arteriovenous Malformation Syndrome

Learning Objectives
- Anticipate risk factors for ulceration in infantile hemangiomas
- Effectively manage infants with ulcerated hemangiomas
- Determine appropriate work up for multifocal infantile hemangiomas
- Recognize the clinical features of tufted angioma
- Describe the clinical features of capillary malformation-arteriovenous malformation syndrome (CM-AVM)
- Determine appropriate work up and coordinate care for infants with high risk vascular lesions

The following references were helpful in creating an evidenced-based discussion surrounding each case.

Introduction:
The classification for vascular anomalies was recently updated by the International Society for the Study of Vascular Anomalies (ISSVA). This classification is based on the original Mulliken and Glowacki binary classification for vascular tumors and malformations introduced in 1982. The field has grown over the last several years and so too has our knowledge of vascular anomalies in many ways: Histopathology, genetic basis and much more. A recent summary and recommendations of the new updated ISSVA classification was recently published in Pediatrics.


Case 1: Ulcerated Infantile Hemangioma

What are the risk factors for ulceration in infantile hemangiomas?
Mucosal or intertriginous sites, Large size, segmental distribution, early white/gray discoloration.
What is the best management for ulcerated infantile hemangiomas?
In general, proper wound care and topical therapies including barrier oint (Vaseline or petrolatum/aquaphor), topical metrocream. Pain control is necessary, topical lidocaine 5% oint in a small pea sized amount several times daily is helpful along with oral analgesic (Tylenol) when necessary. Topical timolol and oral propranolol are emerging as helpful treatments for ulcerated hemangiomas. For small uncomplicated ulcerations, topical timolol could be an initial measure. For larger, segmental or already ulcerated lesions, oral propranolol has become standard of care. Predicting/anticipating ulceration and preventing ulceration from occurring is ideal.

Case 2: Multifocal Infantile Hemangiomas
A prospective study in 2011 identified that 5 or more cutaneous IH are associated with hepatic involvement. Regular screening with abdominal ultrasonography can pick up hepatic involvement early. In the past, diffuse hepatic involvement carried significant mortality, however with early screening and swift treatment if needed, outcomes have improved. An additional complication of multifocal or diffuse hepatic hemangiomas includes hypothyroidism, as hemangioma tissue produces type 3 iodothyronine deiodinase which consumes thyroid hormone. Propranolol is effective in the treatment of diffuse hepatic hemangiomas.

Case 3: Tufted Angioma
Tufted angioma (TA) is a rare vascular tumor which shares clinical, histopathologic features and lineage with kaposiform hemangioendothelioma (KHE). Tufted angiomas are heterogenous in their clinical presentation, but have a predilection for the head and neck and extremities. They often present as ill-defined pink to violaceous plaques or nodules, hypertrichosis and hypertrichosis are often observed. They are generally more superficial and less invasive than KHE. The most significant possible complication is Kasabach Merritt Phenomenon, a consumptive coagulopathy resulting in extreme thrombocytopenia. Treatment options depend on size, location and symptoms but may include excision, systemic chemotherapeutic agents such as vincristine and oral steroids. Sirolimus is also emerging as an effective treatment modality for both TA/KHE.
Case 4: Capillary malformation- arteriovenous malformation syndrome (CM-AVM)
Infants presenting with numerous small capillary stains should be evaluated for CM-AVM syndrome. Due to the asymptomatic nature of individual lesions, this condition is likely under-reported. Genetic counseling for affected family members is important due to significant genotype-phenotype variability and risk for large or intracranial AVMs.

