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Housestaff Puzzler

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A 14-MONTH-OLD WHITE MALE presents for a 20 minute acute visit with a complaint of itching rash on hands and feet. His mom said the rash had begun 8 months earlier with multiple groups of pustules on his hands and feet which would last a few weeks and then disappear, only to recur. He was treated on two previous occasions with permethrin without a change in clinical course. Sometimes the rash makes him significantly irritable because of the pruritis but this lasts only 1-2 days at onset of the lesions.

PMHx is non-contributory. His parents and his adolescent sister had mild eczema in their first few years of life before resolving. He has had no known sick contacts and stays at home with his mother.

PE: Afebrile, with normal pulse, respiratory rate and blood pressure for age. He

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Small-Vessel Vasculitides in Children

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The two most important small vessel vasculitides in children are Henoch-Schonlein purpura and Hypersensitivity vasculitis. I will discuss these disorders in a question and answer format the way we do in rheumatology rounds. Next time you visit our clinic you will be able to answer those question and impress our fellows:

1. What are the small vessel vasculitides?

Small-vessel vasculitis includes a variety of conditions that are grouped because of involvement of small vessel of the skin, especially arterioles and postcapillary venules. Leukocytoclastic vasculitis (LCV) and necrotizing vasculitis are terms used to describe the usual histopathology, in which small vessels are infiltrated with polymorphonuclear neutrophils and/or mononuclear cells. As the process evolves, fibrinoid necrosis of

the vessel wall with leukocytes fragments (leukocytoclasia) and destruction of the blood vessel wall is seen. The conditions associated with LCV include (Table 1)

2. What causes this group of small vessel vasculitis?

The cause is dependent on the underlying associated conditions. The vascular injury is believed to be trigger by the deposition of immune complexes in the vessel wall with activation of PMN's to the area, and release of lysosomal enzymes, and damage to the vessel wall.

3. What is the major clinical manifestation in small vessel vasculitis?

Palpable purpura is the most common primary lesion in cutaneous vasculitis. Typically, hundreds of discrete subtly palpable, purpuric spots suddenly appear on the lower extremities. The hands, arms and other body sites also may be affected. These lesions are dynamic, often beginning as asymptomatic, nonpalpable, purpuric macules and eventually becoming palpable. Some may become nodular, bullous, infarctive, and

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Urinary Tract Infections in Children: What If The Studies Are Normal?

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Host/Pathogen Interactions

Over 40 years ago, the Urologist Hugh Cabot proposed the Doctrine of the Prepared Soil... "It is not rare to find in the active, devoted young hospital surgeon a state of

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Table 1

Condition	Comments
a. Hypersensitivity vasculitis	Drug reaction or idiopathic
b. Urticarial vasculitis	Rare in children
c. Henoch Schonlein purpura	Most common vasculitis in children
d. Mixed cryoglobulinemia	Rare in children, Hep B and C associated
e. Rheumatic disorders	SLE, Dermatomyositis, JRA
f. Infectious	SBE, Neisseria, Mono, HIV, Hep B and C
g. Malignancy	Lymphoproliferative disorders
h. ANCA associated vasculitis	Wegener's, CSS, MPA

ulcerative.

4. Can patients with small vessel vasculitis have systemic manifestations?

Yes. Constitutional symptoms, including fever, arthralgias, and malaise, frequently accompany the appearance of the skin lesions.

5. Are any laboratory findings specific for small-vessel vasculitis?

The laboratory abnormalities are usually non-specific. Normocytic, normochromic anemia, elevated ESR, or eosinophilia is seen in approximately two-thirds of the patients. In those patients with renal involvement, hematuria and proteinuria may be present, as well as an increase in serum creatinine. The ANA is positive in 10%, and RF in 20%. The significance of these serologic findings is uncertain. Hypocomplementemia is infrequent (except in cryo and SLE).

6. How does one make the diagnosis of small vessel vasculitis?

The evaluation of these patients requires a full medical evaluation and appropriate lab tests depending on the clinical situation. Diagnosis is made by skin biopsy. Immunofluorescent study of the skin is helpful in differentiating systemic diseases as microscopic PAN and HSP. The lack of immune complex deposits distinguishes MPA from HSP and cryo, in which immunoglobulins are deposited in the vessel wall. However, skin biopsy cannot discern the etiology

of cutaneous vasculitis (infections, drugs, cryoglobulinemia, malignancy, etc.). Therefore, a complete evaluation must be undertaken with history, physical exam, and selected laboratory tests.

7. How is small vessel vasculitis treated?

Treatment has to be determined individually. If an associated disorder can be identified, treating this problem will suffice. Any potential drug or antigen should be discontinued or removed. An underlying infection must be properly treated. If systemic symptoms are present and skin lesions are diffuse, or if internal organ involvement is present, glucocorticoids are usually the treatment of choice.

8. What are the histopathologic features of HSP?

The histopathologic features of HSP are LCV or necrotizing vasculitis. The characteristic direct immunofluorescence finding is IgA deposition in affected blood vessels. The presence of IgA deposition makes this syndrome pathologically different from any other form of vasculitis.

9. What is the role of IgA in the pathogenesis of HSP?

IgA plays a pivotal role in the pathogenesis of HSP. There are two sub-classes of IgA, IgA1 and IgA2. IgA1 accounts for 80-90% of serum IgA, whereas only 50% of secretory IgA. HSP is only associated with

IgA1 deposition and not IgA2. Notably, IgA nephropathy involves IgA1 exclusively. For both HSP and IgA nephropathy, investigators have found that the hinge region O-linked glycans of IgA1 are deficient in galactose and/or sialic acid content. IgA1 molecules with these deficiencies have a tendency to form macromolecular complexes and have the ability to activate alternative complement pathway. In addition, IgA can bind to mesangial cells in the kidney leading to proliferation and release of pro-inflammatory cytokines. This kidney receptor binds to IgA1 at the hinge region more readily when IgA1 is deficient in sialic acid or galactose.

10. Describe the clinical manifestations of HSP.

The classic tetrad of palpable purpura, arthritis, abdominal pain, and renal disease occurs in 80% of cases. The rash may begin as a macular erythema and urticarial lesions, but progresses rapidly to purpuras. The lower extremity and buttocks are the most common sites for the rash. The joints are involved in 60-84% of patients. The involvement is symmetrical and most commonly involves the ankle and knees. Gastrointestinal lesions may cause severe cramping, abdominal pain, intussusception, hemorrhage and rarely ileal perforation. Renal involvement is seen in 50% of patients and is usually manifested by asymptomatic proteinuria and hematuria. However, nephritic syndrome and acute renal failure can also occur.

HSP is often acute in onset and resolution is rapid and complete in 97% of cases, except for a minority of patients with chronic renal insufficiency (3-5%). The disease occurs between ages 2 and 15, onset is common in winter months and often following an upper respiratory tract infection.

Patients with suspected HSP but without IgA on biopsy should

be evaluated for Wegener Granulomatosis.

11. How is HSP treated?

The disease is generally self-limited, lasting 6-16 weeks. For mild cases, supportive treatment alone may be adequate. Arthritis responds to NSAID. Systemic glucocorticoids may be used in patients with GI involvement or bleeding. Progressive renal disease is difficult to treat and usually does not respond to glucocorticoids. Aggressive treatment with high dose pulse glucocorticoids and cytotoxic agents should be considered in patients with poor prognostic factors: proteinuria > 1gm/day, nephritic syndrome, and crescentic glomerulonephritis.

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mind in which he almost believes that bacteria are the cause of infection. He appears to forget that infection is a result, and that bacteria in and of themselves can do nothing except in contact with living tissue and then, often, only under highly specialized conditions." This doctrine is as true today as it was 40 years ago. Management has to search for why a patient has an infection.

Bacterial Adhesion

Physicians and families can easily grasp the significance of anatomic abnormalities. The child with recurrent UTI's and a normal radiographic evaluation or the child with recurrent UTI's after successful ureteral reimplantation is a quandary. Both host and pathologic factors are important in these patients. While some of these factors are beyond physician and patient control, others are easily modified.

Blood group antigens are genetically controlled carbohydrate molecules, many of which are expressed on urothelial cells. These cell surface antigens may play a role in bacterial adherence and host susceptibility to UTI. Jantusch, et al, have shown that patients with UTI have a higher than expected frequency of blood group antigen Lewis (Le)(a-b-) and a lower than expected frequency of Le(a+b+). It has also been suggested that enteropathic *Escherichia coli* adhere to epithelial cells via filaments termed bundle-forming pili. These bacterial pili are also know as adhesins. Adhesin positive isolates are also more resistant to antibiotic treatment. By itself, expression of virulence factors on periurethral *E. coli* is not enough to predict subsequent infection. Altered host factors come into play which allow the bacteria to establish an infection.

Introital Colonization

Because the female urethra is very short, it poses only a minimum barrier between the bladder and perineal bacterial flora. This means that introital colonization with enteropathic bacteria may play a role in UTI development in females. The make up of the introital bacteria may shift with puberty and its associated hormonal changes. This is thought to account for the spontaneous resolution of recurrent infections in some girls at the time of puberty. Alternately, in older females, the act of sexual intercourse may increase the risk of bacterial inoculation of the bladder.

In a similar fashion, prepuccial bacteria may have a role in UTI development in boys. While the pros and cons of circumcision are beyond the scope of this chapter, circumcision has been shown to decreased the risk of UTI's. Meta-analysis of 10 published studies on the relationship of circumcision and UTI shows a 12-fold increase in UTI's in uncircumcised infants. This protective effect may decrease the risk of symptomatic UTI in preschool boys.

Dysfunctional Voiding

Pathophysiology

The bladder is a complex, dynamic organ with distinct anatomic and functional regions. It should store urine at a low pressure, and maintain continence until a socially acceptable time, when it should fully empty. These dynamic phases require the detrusor muscle and bladder neck to work in harmony. During storage, the bladder must stay relaxed, maintaining low detrusor pressures, despite increasing bladder volumes. At the same time, the bladder neck/internal sphincter maintains sufficient tone to preserve continence. The external sphincter is normally passive during

storage. It serves to counteract socially inappropriate bladder contractions by temporarily increasing outlet resistance and by reflexively relaxing the detrusor contraction. It is the “on/off” switch for the bladder. During voiding, the detrusor contracts while the bladder outlet relaxes.

Detrusor sphincter dyssynergy is manifested by disruption of the coordinated responses of the detrusor muscle and bladder outlet. The detrusor muscle may contract during attempted storage or the bladder outlet may fail to relax during voiding. This dyssynergy produces elevated storage pressures and incomplete emptying with increased post void residuals. Like any muscle that is forced to work against increased resistance, it can hypertrophy leading to decreased compliance and VUR. This dyssynergy may be neurological in origin but is more commonly a behavioral or functional problem in a neurologically normal child.

Evaluation

Just as a child must learn the socially important skills of walking and talking, so too, a child must learn bladder and bowel control. Children may go through a period of bladder dyssynergy during this learning process. The responsible physician needs to thoroughly examine the voiding habits of any child with a UTI and pay special attention to infrequent voiding. Symptoms of urgency, diurnal enuresis and constipation are common in these children. Children are busy playing and exploring their world and may attempt to postpone bladder emptying until the last possible moment at which a crisis is reached. The history in these children often reveals infrequent voiding. Urgency may be a sign of resultant detrusor instability which occurs at bladder capacity. When this unwanted contraction occurs,

the external sphincter is used in the attempt to shut off the detrusor contraction. Vincent’s curtsy is considered pathognomonic in which the child squats and places the heel of the foot into the perineum to help augment the external sphincter, often in a failed attempt to maintain continence.

Treatment of Voiding Dysfunction

Treatment needs to be directed at improving voiding dynamics. This involves the work and cooperation of both the child and parent. Without the cooperation of both, behavioral treatment is doomed to failure. The child should be instructed to attempt to void every two hours during the day. This usually requires a responsible adult to remind the child. Relaxation of the pelvic muscles should be stressed and straining to void discouraged. Double voiding is the technique of attempting a second void several seconds after initial bladder emptying. This may decrease post void residual and reinforces the concept of completely emptying the bladder. Anticholinergic medication may help with detrusor instability. Finally, constipation needs to be addressed. There may be a global elimination dysfunction of both the bladder and bowel. Treatment of voiding dysfunction and constipation has been shown to decrease the frequency of UTI’s.

Asymptomatic Bacteriuria

Treatment of asymptomatic bacteriuria (ABU) remains controversial. The first priority is to establish that it is truly asymptomatic. There may be associated symptoms such as enuresis or a finding on history such as infrequent voiding. Studies have shown that antibiotic treatment of ABU often makes little difference as far as the rate of reinfection and that the risk of renal damage is low. Theoretically, antibiotic treatment

may alter the bacterial colonization to a more virulent strain of E. coli. In such cases, follow-up without antimicrobial treatment may be preferred.

Antibiotic Prophylaxis

The ideal antibiotic for urinary prophylaxis should be safe, effective, inexpensive and have no side effects. While no antimicrobial is ideal, some are preferable in children. Prophylactic dosage is usually one quarter of the therapeutic dose given once per day. Too high a dose will increase side effects such as gastrointestinal upset and may alter fecal flora.

Trimethoprim alone or in combination with sulfamethoxazole is the most commonly used antibiotic for both treatment and prophylaxis of UTI. It is inexpensive and has minimal adverse effects on the bowel and vaginal flora because it is excreted and concentrated in the urine. It may cause a hypersensitivity rash and gastrointestinal upset. It may rarely cause a megaloblastic anemia through folate antagonism and should be used with caution in patients with G6PD deficiency. It is not appropriate during the first month of life due to the risk of jaundice and hemolytic anemia in the newborn.

Another common prophylactic antimicrobial is nitrofurantoin or macrodantin. It also is excreted in the urine, which allows urinary levels to be high while having few effects on fecal flora. It is inexpensive and comes in both a liquid and tablet preparation. It may also cause gastrointestinal upset. Rarely, it is associated with a peripheral neuropathy and pulmonary hypersensitivity. It should not be used in patients with impaired renal function or during the first month of life.

Amoxicillin is the prophylactic antibiotic of choice for the newborn because it can be safely metabolized in the first month of life. Its primary

side effect is a hypersensitivity reaction in some patients. It also can alter gastrointestinal flora which can lead to diarrhea and rarely pseudomembranous colitis. The liquid preparation requires refrigeration which may be inconvenient. The cephalosporins have similar properties to amoxicillin but are more expensive.

A brief warning should be stated about the quinolone antibiotics. While quinolones are used for prophylaxis in adults, they are inappropriate in children due to arthropathic effects which may damage growth plates.

Summary Algorithm

The following algorithm provides a rational basis for the basic evaluation of any child with the diagnosis of UTI. Using such a treatment plan should minimize the

risk of missing significant genitourinary pathology and preventable renal damage.

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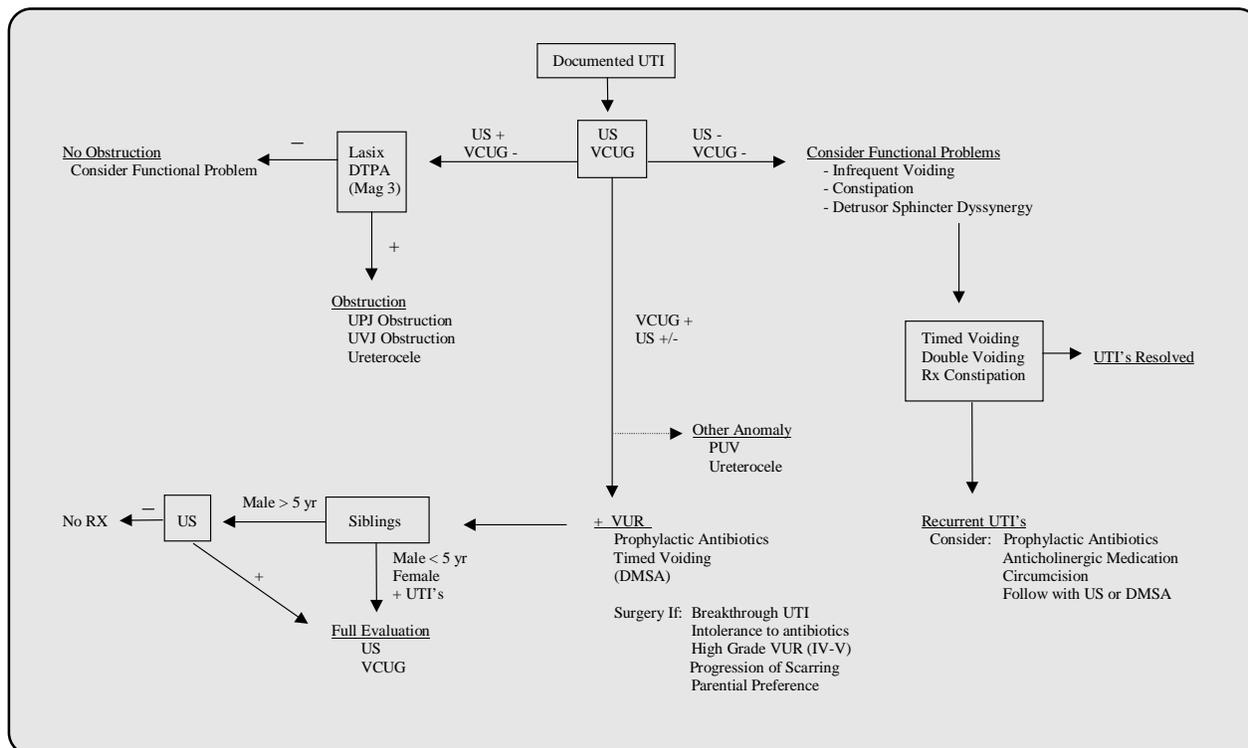
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Vascular Anomalies in Infants and Children

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A Plastic Surgeon and a Biochemist wrote the landmark article that should have quickly led to our better understanding and treatment of the various vascular anomalies in infants and children almost twenty years ago.¹ The article received due notice, but many of us went about the care of children affected by these disorders as though we were unaware of this important information. In 1996, the *International Society for the Study of Vascular Anomalies* accepted the classification as proposed by Mulliken and Glowacki in 1982.

One problem with the dissemination of knowledge about these important problems is that their care crosses the boundaries of multiple specialties: General Pediatrics, Pediatric Pulmonology, Pediatric Hematology-Oncology, Pediatric Radiology, Interventional Radiology, Neuro-radiology, Pediatric Gastroenterology, General Surgery, Plastic Surgery, Pediatric Surgery, Dermatology (General and Pediatric), Ophthalmology (General, Pediatric, Oculoplastic, Retinal), Neurosurgery (General and Pediatric), Otolaryngology (General, Pediatric, Head & Neck Oncology), Pathology, and other disciplines diagnose and treat these conditions. There is no primary specialty or “clearing house” of information to keep us all on the right track.

Several of the disciplines have developed their own system of classification and nomenclature for these anomalies, producing much confusion, anguish and improper treatment of patients.

To properly care for infants and children with Vascular Anomalies, we must forget much of what we have previously learned and learn a new system. The hard part is the forgetting, as the new system is easy and intuitive.

Classification (ISSVA, 1996)

Vascular anomalies are divided into two types, the Vascular Malformations and the Vascular Neoplasms. Vascular Malformations are lesions that arise by dysmorphogenesis while Vascular Neoplasms are truly neoplastic in nature.

Examples of Vascular Malformations are Capillary Malformations (e.g. “Port Wine Stains”), Venous Malformations (many have been incorrectly called “cavernous hemangiomata”), Lymphatic Malformations (previously called “Cystic Hygromas” or “Lymphangiomas”), Hereditary Hemorrhagic Telangiectasia (Osler-Weber-Rendu), and arteriovenous malformations.

Examples of vascular neoplasms are Infantile Hemangiomas, Juvenile Nasopharyngeal Angiofibromas, Hemangiopericytomas, Pyogenic Granulomas, Tufted Angiomas and Kaposiform Hemangioendotheliomas. The last two are of special interest due to their association with the Kasabach-Merritt Phenomenon.

Behavior of Vascular Malformations

These lesions are present at birth (90% noted) and grow with the patient. They may have size changes related to fluid content but neither proliferate nor involute. A major treatment implication here is that these lesions will not respond to anti-neoplastic chemotherapeutic agents.

Behavior of Vascular Neoplasms

These lesions are theoretically present at birth but may not be recognized. They manifest cellular growth at some time in life (intrauterine only in at least one instance). The most common of these lesions (the common hemangioma of childhood) undergoes rapid proliferation in infancy followed by slow involution in childhood. Some of these lesions, most notably the most common one, will show response to ant-neoplastic chemotherapeutic agents.

Diagnosis

The major diagnostic call is between the common hemangioma of childhood (Infantile Hemangioma) and the various Vascular Malformations. The diagnosis is typically a clinical one, rarely requiring tissue sampling or even imaging. Superficial hemangiomas are typically red, raised and bosselated (strawberry-like). Some Venous Malformations can be strawberry-like but will differ in that they will be fully formed at birth and quite compressible. Infantile Hemangiomas may be detectable at birth, but will show

Examples of Changes With New Classification

<i>Common Name</i>	<i>Old Classification</i>	<i>New Classification</i>
Port Wine Stain	Capillary Hemangioma	Capillary Malformation
Lymphangioma; Cystic Hygroma	Lymphangioma	Lymphatic Malformation
	Cavernous Hemangioma	Venus Malformation

rapid enlargement. Also, they will show some resistance to compression due to their cellular nature.

Deep hemangiomas are more difficult to separate from Vascular Malformations. History alone will often cinch the diagnosis (growth with the infant with the VM, rapid growth with the VN). In the occasional case where history and examination fail to make the diagnosis, imaging studies may provide the needed information. Consult with your radiologist before ordering a study, as their guidance may prove most helpful.

Treatment Principles Vascular Malformations

Watchful neglect is probably the most common appropriate therapy for these lesions. The most compelling reasons for active treatment are threats to life or limb. The most common examples of such threats are airway encroachment, heart failure with large AVMs and threats to vision. Cosmetic concerns may also be legitimate reasons to treat some of these lesions.

Among the active treatment options are sclerotherapy, surgical excision, laser ablation, embolization and various combinations of these.

Treatment Principles Vascular Neoplasms

Because the common hemangioma of infancy/childhood undergoes rapid proliferation followed by involution, it is subject to the effects of agents that influence such processes. Corticosteroids such as prednisone are effective in slowing/stopping proliferation and, perhaps, speeding up involution. Some chemotherapeutic agents usually reserved for malignancies will likewise show effects on these lesions, but are reserved for life-threatening situations. An example

is the child with visceral hemangiomata which are causing profound heart failure and have not responded to corticosteroids.

Again, watchful neglect is the appropriate treatment for many of these lesions. Compelling reasons to treat include airway encroachment, eye/vision concerns, high-output heart failure (especially with visceral hemangiomas and large truncal/extremity hemangiomas), major impairments in function and major cosmetic concerns. As noted above, chemotherapeutics, especially corticosteroids and alpha-2 interferon, are effective for the common hemangioma of childhood. Other means of treatment include conventional surgery, laser surgery, embolization, sclerotherapy, and combinations of these.

Special Things to Remember About the Eye

The eye is at risk with many vascular lesions of the eyelids and adjacent structures. Don't forget that even the lesion that is going to involute with time may cause blindness via the amblyopia route due to temporary mechanical blockage of the sight. Also, anything that puts the lightest pressure on the cornea will cause astigmatism.

Summary

Vascular anomalies are common in infants and children. It is important, and usually easy, to diagnose them so that accurate predications about their course may be made and the best management chosen.

Vascular Malformations are non-neoplastic, present at birth (90% noted), grow with the child and do not respond to corticosteroids or interferon. They must be treated for life or limb threats, but may be treated for lessor concerns in

selected patients. Sclerotherapy is effective for many and is preferred over surgery most of the time.

Vascular Neoplasms are either present or represented by a nidus at birth (60% noted). The most common of these, the "Infantile Hemangioma", will undergo rapid growth via cell proliferation in infancy and will show slow involution during childhood (40% by age 4, 90% by age 9). Only those with "life or limb" threats must be treated. Others may be treated on a case-by-case basis, but watchful neglect is the indicated treatment for the majority. The common hemangioma of childhood will respond to corticosteroids and/or interferon alpha-2 about 90% of the time.



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is well appearing and is occasionally scratching his hands and feet. His exam is only remarkable for multiple 1- to 2-mm vesicular-pustular lesions over his palms, the soles of his feet, and the lateral regions of his hands, wrists, feet and ankles. No burrows are appreciated and the intertriginous areas appear to be spared. No oral lesions are present. There is no HSM or LAD.

You obtain skin scrapings and observe them under mineral oil examination and are negative for any infestation. You go back and aspirate the contents of several of these lesions and send them to the lab for gram stain and culture. You send the patient home with Bactroban and a prescription for benadryl. They leave your office with you scratching your head. Two days later the lab reports many

PMNs and the culture is negative for growth.

Your most likely diagnosis is:

- A. Pustular psoriasis
- B. Infantile acropustulosis
- C. Impetigo
- D. Recurrent Coxsackie virus infection

The most appropriate therapy is:

- A. A course of oral antibiotics and continue the Bactroban you Rx'd.
- B. Reassurance and watchful neglect.
- C. A high-potency topical corticosteroid.
- D. Topical and/or oral diphenhydramine.

Answers: B and C respectively.

This is Infantile Acropustulosis (IA). It is characterized by recurrent crops of intensely pruritic vesicular-pustular lesions in the regions described above. It is very pruritic and sometimes causes significant irritability in infants. The lesions are initially red papules that quickly develop within 24 hours to the classic vesiculo-pustular lesion. The hands and feet are usually involved with lesions located on palms and

soles and on lateral surfaces, and occasionally over the dorsal surfaces. They usually last for 2-4 weeks and recur at regular intervals.

IA is more common in males and in blacks and occurs more frequently in the summer. Onset usually occurs between ages 6 to 12 months but can occur later in childhood. The recurrent episodes invariably stop by 3 years of age.

The etiology of IA is unknown. At presentation, these patients are often diagnosed with scabies infestation (which should always be ruled out) and receive several courses of treatment with anti-scabitic preparations.

Differential Diagnosis

The DDx includes Transient neonatal pustular melanosis, erythema toxicum, milia, cutaneous candidosis, impetigo, pustular psoriasis and dyshydrotic eczema. Scabies can be ruled out by not having similar symptomatic family members and by skin scraping with visualization by mineral oil examination.

Staining of the contents of the

lesions usually shows many PMNs and sometimes a significant number of eosinophils. A KOH prep will be negative for yeast/fungus. Cultures are negative for bacterial growth.

The standard treatment is high potency topical corticosteroids with or without the addition of topical and/or oral antihistamines as adjunctive therapy for the pruritis. There is no cure and the aim of treatment is to relieve the pruritis and irritability. Dapsone (2mg/kg/day) has been used for severe cases but has a significant risk of severe adverse side effects (hemolytic anemia and methemoglobinuria), so, it should be used with caution. .

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