

Childhood Psoriasis – a challenge to all

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Abstract

Pediatric psoriasis consists broadly of 3 age groups of psoriatic patients: infantile psoriasis, a self-limited disease of infancy, psoriasis with early onset, and pediatric psoriasis with psoriatic arthritis. About one-quarter of psoriasis cases begin before the age of 18 years. A variety of clinical psoriasis types are seen in childhood, including plaque-type, guttate, erythrodermic, napkin, and nail-based disease. Like all forms of auto-immunity, susceptibility is likely genetic, but environmental triggers are required to initiate disease activity. The most common trigger of childhood is an upper respiratory tract infection. Once disease has occurred, treatment is determined based on severity and presence of joint involvement. Topical therapies, including corticosteroids and calcipotriene, are the therapies of choice in the initial care of pediatric patients. Ultraviolet light, acitretin and cyclosporine can clear skin symptoms, while methotrexate and etanercept can clear both cutaneous and joint disease. Concern for psychological development is required when choosing psoriatic therapies. This article reviews current concepts in pediatric psoriasis and a rational approach to therapeutics.

Keywords: Psoriasis, autoimmunity, *Streptococcus*, etanercept, calcipotriene, topical corticosteroids

Introduction

Twenty ninth of October is observed as World Psoriasis Day; for the year 2010, 'the theme is fixed as 'Childhood Psoriasis – a challenge to all!'

Psoriasis vulgaris is a common dermatologic disorder seen in about 3.5% of the population.¹ One-third of psoriasis cases in a dermatology center are pediatric.² Psoriasis is a T-cell mediated chronic inflammatory disorder of the skin characterized by hyper-proliferation of keratinocytes and consequent red scaly skin plaques. Pediatric onset psoriasis is somewhat different than adult disease, as pharyngitis, stress, and trauma are more common triggers of disease activity than in adulthood.^{3,4} Despite the differences in

pediatric psoriasis, the therapies used for pediatric psoriasis are essentially the same as those used in adulthood, with dosage and strength reductions calculated based on age, weight, and available formulations. This article looks at a rational approach to the diagnosis and management of pediatric psoriasis, with a careful focus on those aspects of disease unique to the pediatric patient.

Pathogenesis

The exact pathogenesis of psoriasis has not been completely elucidated; however, it is known to have a genetic basis, as 23.4% to 71% of children will have a family history of psoriasis^{3,5,6} The guttate psoriasis subset is linked to inflammatory focus in about two-thirds of cases, and is not caused by a specific subtype of group A beta hemolytic streptococcus, but rather by a host-specific response. Cross-reactivity of keratinocytes antigens with streptococcal antigens is thought to initiate psoriatic disease in this setting. Other infections that have been noted in psoriatic disease are presence of staphylococcal superantigens and HPV DNA.^{7,8,9} No single gene has been found to be responsible for psoriasis vulgaris. A series of genes have been isolated in which mutations have been associated with psoriatic disease. These genes play a role in Th2 cell and Th17 cell activity and signaling, demonstrating both a role for Th2 and Th17 lymphocytes in the pathogenesis of psoriatic disease. Th17 cells have been noted in psoriatic lesions, as have collections of Th2 and Th1 cells.¹⁰

Many patients with psoriasis will have other autoimmune conditions, often of the skin, including morphea and vitiligo vulgaris (sometimes this association is familial).¹¹ Family history of psoriasis and other forms of autoimmunity can be noted in patients with psoriasis and in patients with a personal history of other autoimmune diseases such as multiple sclerosis.¹²

Clinical aspects and diagnosis

Most children manifest with plaque-type psoriasis vulgaris (68.6%) in similar patterns to adult patients, with lesions localized to the scalp, post auricular region, elbows, and knees. Guttate disease is more common in pediatric than adult patients. Diaper involvement is very common in infancy, but involvement of the groin is uncommon in older children. Inverse psoriasis with involvement of the folds of the skin (axillae, inner thighs) represents a small minority of children. Additionally, nail psoriasis can be noted in the setting of plaque-type psoriasis vulgaris, psoriatic arthritis, or with isolated nail disease, the last sometimes being called trachyonychia, although this is controversial. Involvement of joints with psoriatic arthritis is less prevalent in younger patients; however, it does occur in childhood disease and should be considered in the differential of pediatric arthritis.⁴

Psoriasis vulgaris occurs in a variety of clinical types.

A few clinical features of psoriasis that are pertinent during physical examination include

- 1) The isomorphic response or Koebner phenomenon, which is occurrence of lesions in areas of trauma,
- 2) Altered pigmentation with lesional clearance,
- 3) The Auspitz sign – pinpoint bleeding at the base of scale that has been removed, and
- 4) Presence of nail pitting, which can aid in diagnosis of the disease. Severity grading for psoriasis is usually based on surface area and presence and co-morbid psoriatic arthritis. The Psoriasis Area and Severity Index (PASI) can be used to assess severity (given below). Others will divide disease into mild if less than 3% body surface area, moderate 3% to 10% body surface area, and severe >10% body surface area.¹³

Psoriasis Area and Severity Index (PASI) score

To determine PASI score, establish grade and severity and then use Site-based weighting

Grade: surface area as below (0–6)

0% of involved area, grade: 0

- *<10% of involved area, grade: 1*
- *10%–29% of involved area, grade: 2*
- *30%–49% of involved area, grade: 3*
- *50%–69% of involved area, grade: 4*
- *70%–89% of involved area, grade: 5*
- *90%–100% of involved area, grade: 6*

Severity: 0–4 none to severe

Erythema (redness)

Induration (thickness)

Desquamation (scaling)

Notes: The sum of all three severity parameters is then calculated for each section of skin, multiplied by the area score for that area and multiplied by weight of respective section (0.1 for head, 0.2 for arms, 0.3 for body and 0.4 for legs).

Differential diagnosis

The differential diagnosis of psoriasis includes other papulosquamous disorders of childhood including lichen planopilaris, psoriasiform ID reactions, nummular dermatitis, pityriasis rosea, and pityriasis rubra pilaris. Biopsy can be helpful in differentiating psoriasis from these other illnesses.

Treatments

The treatments of psoriasis have expanded over the past decade; however the use of topical therapy in childhood is the first line of treatment for skin-limited disease, in combination with a trial of oral antibiotics where indicated. With chronicity of illness and in more severe cases, systemic therapy and phototherapy are added to help induce remission. Significant psychological disturbances are seen in children with psoriasis, no matter what the surface area. Disease,

although cutaneous, should be treated more aggressively when it is causing psychological disturbance, to improve quality of life.

Topical therapy

Topical therapies for pediatric psoriasis include over-the-counter agents such as tar and salicylic acid, the latter of which aids in removal of hyperkeratosis. Tar has been a controversial medication in pediatric psoriasis because of demonstrable genotoxic risk, including chromosomal aberrations in peripheral lymphocytes and release of heat shock protein. Prescription agents for psoriasis vulgaris in childhood include anthralin, topical corticosteroids, topical calcipotriene with or without topical corticosteroids, and topical calcineurin inhibitors such as tacrolimus 0.3% ointment, and pimecrolimus 1% can be beneficial for pediatric psoriasis, particularly in sites where atrophy is of risk, such as the face, intertriginous areas, and groin. Use in children under the age of 2 years is not recommended.^{13, 14}

Phototherapy

Phototherapy is a safe and effective treatment for children old enough to stand still in a phototherapy booth, particularly teenagers with extensive disease. Generalized or hand foot therapy, either narrowband UVB (NBUVB) or psoralens and UVA (PUVA), can be used. Topical psoralens are preferable to oral psoralens because of the difficulty of wearing protective eyewear for a 24-hour time period after oral psoralens. As PUVA has been associated with long-term carcinogenicity in psoriatics, NBUVB is likely safer in childhood cases.^{15, 18}

Systemic agents

Systemic agents should be saved for severe psoriasis, disabling psychological ramifications, and psoriatic arthritis. Usage of systemic agents is generally limited to a 6-month time period for immunosuppressants such as cyclosporine, acitretin or methotrexate. Longer usage periods have been observed to be safe in rheumatoid arthritis patients on etanercept; however usage beyond a one year time period has not been well assessed in pediatric psoriatics. Methotrexate has been used for pediatric patients for decades longer than etanercept. Oral antibiotics are the systemic therapy of choice in early disease, due to their superior side effect profile.

1. Oral antibiotics can be useful in treating psoriasis vulgaris, particularly in the setting of positive oral pharyngeal cultures, presence of perianal bacterial dermatitis, pustular psoriasis,³⁹ or in guttate psoriasis of childhood (but not adult guttate disease), as these settings are linked to a bacterial precipitant of psoriasis.^{16, 17}
2. When pediatric psoriasis becomes chronic and severe (PASI > 10), systemic therapy provides the major source of improvements in pediatric quality of life. The original psoriatic systemic therapy used for extensive psoriasis is methotrexate, which has been used for over 3 decades for pediatric psoriasis and pediatric psoriatic arthritis. Addition of folic acid supplementation helps protect against pancytopenia and macrocytic anemia.^{18, 19}

3. Cyclosporine A, a systemic immunosuppressant used originally for prevention of transplant rejection, can be dosed for oral use at 3 to 5 mg/kg can improve cutaneous symptoms in pediatric psoriatic patients. Alterations in renal function with altered serum urea nitrogen and creatinine and high blood pressure can be observed. Therefore, close monitoring is warranted. Risks of malignancy and lymphoproliferative disorders seem to be minimal in children treated for skin diseases due to limited courses of therapy and dosages that are below 5 mg/kg/day.²⁰
4. Retinoids: Acitretin orally 0.5 to 1 mg/kg per day has been used for disorders of cornification and psoriasis with good results. Because of teratogenicity, oral contraceptives in girls of childbearing age should be used concurrently, and for 3 years after drug discontinuation. Short-term side effects, such as elevations in lipids or alterations in blood counts, require monitoring. Long-term changes such as bony abnormalities can occur and treatment periods should be limited, using a cyclic approach. Bony evaluation may be required.²⁰
5. Biologics: Etanercept and infliximab, injectable and intravenous, respectively, tumor necrosis factor alpha (TNF- α) inhibitor therapies have been used for a decade in pediatric psoriasis. Long-term use did not increase serious side effects such as tuberculosis, opportunistic infections, malignancies, lymphomas, lupus, demyelinating disorders, or death. Long-term improvements on bony disease in arthritis of childhood have been demonstrated with etanercept.^{21, 22}

Natural supplements

A common question asked by parents of children with psoriasis is “What dietary changes or natural supplements can I give my child to improve their skin disease?” While natural supplements or dietary alterations cannot cure psoriasis, they can improve disease severity. The best-known supplement is fish oil, rich in omega-3 fatty acids. The ingestion of this agent for psoriasis is based upon its preventive effects in the Inuit population. Oral and intravenous supplementation of omega-3 and, less effectively, omega-6 fatty acids have been found effective in psoriatic adults, possibly through alterations in production and alterations in arachidonic acid (20:4 omega 6) and docosapentaenoic acid. Fish meals 4 to 6 times per week can mimic the effects of omega-3 supplementation.²³

Conclusions

Recent advances in genetics and the unraveling of the processes responsible for psoriatic disease are making possible considerable advancements in the treatment of pediatric psoriasis.

Key Points

1. One-third of psoriasis cases in a dermatology center are pediatric.
2. Most children manifest with plaque-type psoriasis vulgaris
3. A few clinical features of psoriasis that are pertinent during physical examination include
 - i. the isomorphic response or Koebner phenomenon, which is occurrence of lesions in areas of trauma,
 - ii. altered pigmentation with lesional clearance,
 - iii. the Auspitz sign – pinpoint bleeding at the base of scale that has been removed, and
 - iv. presence of nail pitting, which can aid in diagnosis of the disease.
4. The treatments of psoriasis have expanded over the past decade; however the use of topical therapy in childhood is the first line of treatment for skin-limited disease, in combination with a trial of oral antibiotics where indicated.
5. With chronicity of illness and in more severe cases, systemic therapy and phototherapy are added to help induce remission
6. While natural supplements or dietary alterations cannot cure psoriasis, they can improve disease severity.

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