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Principles of CTCL management: what's the evidence

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Three common challenging management issues that arise in caring for cutaneous T cell lymphoma (CTCL) patients were discussed. Based on published evidence, and consensus guidelines, 1. management and prognosis of folliculotrophic mycosis fungoides (FMF)/CTCL 2. traditional vs low dose radiation therapy in CTCL 3. impact of immunosuppressive medications in MF/CTCL.

Compared to classic MF, folliculotropic mycosis fungoides (FMF) has a different clinical presentation with more varied histopathological features and a different prognosis. Epidermotrophism only occurs in 86% of FMF cases, while folliculotropism is a mandatory diagnostic feature for FMF. Other features include: follicular mucinosis (42%), eosinophilic folliculitis (3.5% and Pautrier microabscess (14.9%). Early plaque stage FMF has relatively sparse infiltrates, and small neoplastic T cells while advanced plaque stage FMF has denser infiltrates and medium/large neoplastic T cells. Head and neck involvements occur more often in FMF (70.2%). Early plaque FMF actually has good prognosis with 100 % disease-specific survival in contrast to only 35% in advanced stage FMF at 200-month post-diagnosis.

Independent risk factors for reduced survival include advanced age at diagnosis (>65 years), presence of large cell transformation, stage and secondary bacterial skin infection. Concerning the optimal treatment, Early-stage skin-limited FMF has an excellent prognosis (5-year overall survival 92-94%). Monotherapy such as topical corticosteroid, NBUVB can be offered to patch stage MF. In plaque type, PUVA is better than NBUVB. In advanced stage skin-limited FMF it has less favourable prognosis (5-year overall survival 55%). Multiple agents are needed to achieve complete clearance: PUVA +Radiation therapy, PUVA+ retinoid or interferon (IFN), or Total skin electron beam (TSEB). Low dose electron beam can be useful in clearing refractory lesions while oral bexarotene should also be considered. Compared to traditional radiation therapy, low dose RT/TSEB used in MF/CTCL is effective for palliation, doses range 8-12 Gy (NCCN), it can be used multiple times with less toxicity and cost. Higher doses would be more useful in single bulky folliculitropic lesions.

In the era of treatment 'explosion' of immunosuppressive (IS) and biologics, while rare, it has been known for years that IS increases risks of lymphoma, especially Non-Hodgkin Lymphoma (NHL). In the world of targeted therapeutics, monoclonal antibodies, and utilising immunosuppressants to treat other conditions, it has also increased the incidence of NHL. While mild psoriasis has a 4.1-fold increased relative risk of developing CTCL, severe psoriasis has 10.75-fold increased risk.

The crucial question is whether the psoriasis transformed into CTCL versus CTCL misdiagnosed as psoriasis. A small study of 22 patients with CTCL or B-cell lymphoma showed that most cases were misdiagnosed as psoriasis or eczema. Of these, 75% cases received anti-TNF agents for a presumed inflammatory eruption

Biopsy confirmation before starting biologics is required. Caution should be taken in cases with adult-onset atopic dermatitis without a personal or family history of atopy, atypical psoriasiform eruptions, pityriasis-rubra-pilaris (PRP)-like erythroderma, morphological changes in disease with or without immunosuppressive treatment. In patients with erythroderma, CBC, LDH and flow cytometry should be checked.

Learning points:

There are challenges in patient management in CTCL, evidence-based data and consensus guidelines are invaluable for the management of these cases.

Vascular anomalies symposium: classification, clinical challenges and management updates. What every dermatologist should know

Speaker: LJ Frieden
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The recent published American Academy of Paediatrics (AAP) Clinical practice guidelines in January 2019 highlighted early referral for problematic infantile haemangiomas (IHs) and its management. Our understanding of the growth characteristics IH has changed. Rapid IH growth occurs between one and three months of age. Although IHs involute, this process may be incomplete, leaving permanent skin changes that may be significant. This is especially true for thick IHs.

For lesions that are potentially high-risk, early consultation is strongly advised because of the following associations:

1. Potential for disfigurement: nasal tip/lip of any size, any facial lesions >/=2 cm or <1 cm if <3 month of age, scalp IH>2 cm, thick superficial IHs >2 mm; any lesions with ulceration.
2. Life-threatening complications: >/=5 cutaneous IHs (multifocal IHs) with possible risk of liver haemangioma, cardiac failure and hypothyroidism; 'beard-area' IH with potential for airway obstruction.
3. Functional impairment: periocular IH>1 cm with potential visual impairment, IH involving oral cavity with possible feeding problem.
4. Underlying abnormalities: segmental IH of face/scalp in PHACES syndrome, segmental IH of lumbosacral/perineal area in LUMBAR syndrome.

Oral propranolol is the treatment of choice for problematic IHs that require systemic therapy. Dosing is between 2-3 mg/kg/day unless there are comorbidities or adverse effects (e.g. sleep disturbance) that necessitate a lower dose. Topical timolol may be used to treat some thin and/or superficial IHs. Surgery and/or laser treatment are useful for the treatment of residual skin changes after involution. They may be used early to treat selected IHs.

Learning points:

The best treatment approach is risk stratification with triage and referral for those IHs with high risk. Clinicians in primary care should recognise IHs of high-risk and the window of opportunity for early intervention and prevention of complication is by one month of age. Special attention is needed for those with rebound growth and poor responders. No routine imaging is required unless there is uncertain diagnosis, or to exclude underlying structural/ anatomical abnormalities.

Phototherapy: in the era of biologics and small molecule inhibitors. Shall we still use it for psoriasis in 2019?

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According to the guidelines published on NBUVB, common indications include: psoriasis, vitiligo, cutaneous T cell lymphoma, polymorphic light eruption, atopic dermatitis, and pruritus. In psoriasis management, NBUVB, as well as PUVA often are used as combination therapy with namely, topical corticosteroid, calcipotriene ointment/cream, and traditional systemics such as methotrexate, and acitretin. NBUVB has been shown to be safe and synergistic with biologics including etanercept, adalimumab, ustekinumab, and apremilast.

NBUVB in CTCL has shown good efficacy in patch/plaque stage mycosis fungoides (MF). In one study.

It has been shown that after 52 weeks of NBUVB therapy, >95% (13/24) cases achieved complete or almost complete clinical clearance and 9/10 patients resulted in histological clearance, and over a 32-week follow-up, 34% relapsed at 12.5 weeks.

Comparing NBUVB with PUVA for early stage MF (1a and 1b), NB-UVB achieved complete clearance in 81% (21/56) patients compared to 71% (35/71) who received PUVA. Remission was maintained at 24.5 months after NBUVB as compared to 22.8 months after PUVA. Therefore, NBUVB can be considered in early stage MF, switching to PUVA in case of relapse.

For vitiligo, more than 75% repigmentation was observed in 13% patients receiving 3-month of NBUVB, compared to 19% after 6-month therapy, 36% after 12-month therapy. This suggests that long-term therapy may be necessary. However, the repigmentation as natural course of the disease is could not be excluded by these non-randomised trial and duration of disease may play a role. Nevertheless, NBUVB was considered potentially beneficial as combined therapy with other treatment modalities such as oral antioxidants, tacrolimus ointment (targeted phototherapy), and oral tofacitinib.

Other indications include: Polymorphic light eruptions (PMLE) desensitisation by NBUVB given three times per week for five weeks, with 91% patients good response and no loss of efficacy with subsequent treatments.

In a randomised prospective study, patients with steroid-dependent chronic urticaria who had received 90 days PUVA or NBUVB, an improvement was observed in average urticaria activity score (aUAS7) in both groups at Day 90. This continued to improve at day 180. Patients who received NBUVB responded better than PUVA group. NBUVB should be considered before omalizumab, cyclosporine and other aggressive treatment modalities.

In morphea, NBUVB upregulates matrix metallo-proteinases (MMPs) and inhibits collagen production. It has been found to be effective for patients with localised scleroderma. Other indications for NBUVB include lichen planus, pityriasis lichenoides chronica, lymphomatoid papulosis, and generalised granuloma annulare. Home phototherapy is an emerging option and is convenient for patients. Limitations include patient compliance, maintenance and monitoring of clearance. In a study of 4665 patients, with psoriasis as major diagnosis (55%), Skin photo type (SPT) I-III (97%), there was no increase in BCC, SCC or melanoma. However, there was a slight increase in BCC in patients received both NBUVB and PUVA.

Learning points:

Dermatologists should be familiar with the application, latest protocols, side effects, therapeutic options of phototherapy.