5. Couderc T,Chritiun F, Schilte C, Disson O, Brigitte M, Guivel-Benhassine F *et al*. A mouse model for Chikungunya: young age and inefficient type-I interferon signaling are risk factors for severe disease. Plos Pathog. 2008;4:e29.
6. Chandak NH, Kashyap RS, Kabra D, Karandikar P, Saha SS, Morey SH, *et al*. Neurological complications of chikungunya virus infection. Neurol India. 2009;57:177-80.
7. Tournebize P, Charlin C, Lagrange M. Neurological manifestations in Chikungunya: about 23 cases collected in Reunion Island. Rev Neurol. 2009;165:48-51.

Pemphigus Foliaceus

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Pemphigus foliaceus is an autoimmune blistering disease, which affects the skin but rarely affects the mucosae. There are two variants of pemphigus foliaceus : endemic and sporadic. Erythroderma due to pemphigus foliaceus is unusual and its occurrence in a child is very rare. We describe a case of erythrodermic pemphigus foliaceus in a 12- year- old boy.

Key words: *Foliaceus, Iran, Pemphigus.*

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Pemphigus foliaceus is an autoimmune blistering disease, which affects the skin but rarely affects the mucosae [1]. Erythroderma due to pemphigus foliaceus is unusual and its occurrence in a child is very rare . We describe this entity in a 12- year- old boy.

CASE REPORT

A 12-year-old boy presented to us with widespread skin lesions of 4 months duration. Erythematous and crusted lesions first appeared on his scalp and within a few days it became generalized and then erythrodermic (***Fig.* 1**). Scaling and exudation were also seen. Different antibiotics and topical steroids were prescribed without significant improvement. The child also complained of hearing loss since one month. On physical examination, erythroderma with severe scaling and malodorous discharge was seen. There was mild palmoplantar keratoderma and scales covered the entire scalp. There were two small vesicles along the ulnar side of right palm. The mucosal surfaces and nails were normal. He had two small non tender submandibular lymph nodes. External auditory canal was filled with scales and crusts. Pinna was tender on palpation. After removing the crusts, external auditory canal was found to be red and swollen. Routine laboratory tests were normal except erythrocyte sedimentation rate which was elevated (52mm/hr). *Giardia* cyst was found in stool exam. KOH examination from the scalp scales was negative for dermatophytes. Lesional and perilesional biopsy were taken with impression of pemphigus foliaceus, eczema, psoriasis, and erythroderma due to dermatophytosis.

The microscopic examination showed a subcorneal cleft in the granular layer. A few acantholytic cells were also seen. Mixed inflammatory infiltrate with lymphocytic predominance was seen in dermis. Direct immunofluorescence performed on prelesional specimen showed deposits of C3 and IgG in the upper part of the epidermis compatible with pemphigus foliaceus. Oral prednisolone 30 mg/d and azathioprine 50 mg/d were started. Proper treatment was instituted for external otitis and *Giardia* cyst. Prednisolone was increased to 50 mg/d due to poor response to treatment. A few days later the lesions began to improve. The child was discharged after 45 days of admission. Follow-up was not possible.

DISCUSSION

Pemphigus foliaceus is an autoimmune disease that is characterized by the presence of autoantibodies against the cell surface of keratinocytes, which leads to destruction of epidermal cell junctions. Blistering in this group of autoimmune disease occurs in upper parts of the epidermis, either in the granular layer or just beneath the stratum corneum. Pemphigus foliaceus comprises of two major categories: (*i*) sporadic form; and (*ii*) endemic pemphigus foliaceus also known as fogo selvagem (wild fire) [1-4].

Fogo selvagem primarily affects children in contrast to the sporadic form of pemphigus foliaceus which is

generally a disease of the middle aged and elderly people. It is rare among children [2] with the average age at presentation being 7.7 years [5]. The youngest patient reported so far was a 18- month-old child [5].Childhood pemphigus foliaceus appears to be slightly more common in boys [1]. The most common sites of involvement are scalp and face, followed by the trunk or upper extremities [6]. Lesions of the groin or pubic area are rarely reported. Mucous membranes are generally not involved. Pemphigus foliaceus may remain localized or become generalized. Progression of disease to multiple cutaneous sites occurrs in more than half of the cases [1].The most common primary lesions appear on the face and the scalp as superficial bullae which rapidly rupture and leave scales and crusts. Sometimes only scales and crusts with erythematous bases may be seen [6,9]. Generalized erythroderma has been reported in 2 cases of childhood pemphigus foliaceus [3]. Triggering factors such as UV exposure, drugs and various infections have been proposed as provoking factors for the disease [1]. Our patient suffered from otitis and giardiasis which may have precipitated pemphigus foliaceus. In most cases of pemphigus foliaceus, the diagnosis is based on histopathology. Pemphigus foliaceus may initially be misdiagnosed as impetigo, seborrheic dermatitis, eczema, psoriasis and dermatitis herpetiformis. In majority of cases topical and systemic corticosteroids are used for the treatment.

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FIG.1 *Generalized crusted lesions in Pemphigus foliaceus.*

REFERENCES

1. Metry DW, Hebert AA, Jordon RE. Non endemic pemphigus foliaceus in children. J Am Acad Dermatol. 2003;49:187-9.
2. Amagai M. Pemphigus. *In:* Bolognia JL, Jorizzo JL, Rapini RP, Horn TD, Mascaro JM, Mancini AJ, *et al*., editors. Dermatology. Edinburgh: Mosby. 2008. p.417-28.
3. Mehravaran M, Morvay M, Molnar K, Oláh J, Korom I, Husz S, *et al*. Juvenile pemphigus foliaceus. Br J Dermatol. 1998;139:496-9.
4. Diaz LA, Sampaio SA, Rivitti EA, Martins CR, Cunha PR, Lombardi C, *et al*. Endemic pemphigus foliaceus (fogo selvagem), I: clinical features and immunopathology. J Am Acad Dermatol. 1989;20:657-9.
5. Kahn G, Lewis HM. True childhood pemphigus. Pemphigus foliaceus in an 18-month-old child: immunofluorescence as a diagnostic aid. Am J Dis Child. 1971;121:253-6.
6. Goodyear HM, Abrahamson EL, Harper JI. Childhood pemphigus foliaceus. Clin Exp Dermatol. 1991;16:229-30.
7. Qureshi WA, Ali A, Bhol KC, Ahmed AR. Juvenile pemphigus foliaceus treated with sublesional corticosteroids. Int J Dermatol. 1997;36:848-50.
8. Rosella M, Masia IM, Satta R, Cottoni F. Pemphigus foliaceus in a child. Pediatr Dermatol. 1996;13:259-60.
9. Sotiriou L, Herzenson S, Jordon RE. Childhood pemphigus foliaceus: report of a case. Arch Dermatol. 1980;116:679-80.