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Case Report

LEONINE FACIES IN A CHILD – ENIGMATIC CUTANEOUS HISTIOCYTOSIS

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ABSTRACT

Leonine facies is characterized by thick, coarse, furrowed facial skin with prominent supra orbital ridge, thick glabella and deep furrows on the malar and infraorbital regions giving an appearance of "lion". Diffuse dermal infiltration of papules coalescing to form plaques on the face adds to the manifestation of leonine facies. It is also seen in conditions such as lepromatous leprosy, cutaneous T cell lymphomas, actinic reticuloid and pachydermoperiostosis. We report a case of multicentric reticulohistiocytosis(MRH) in a 9-year-old boy who presented with multiple raised skin lesions distributed all over the body including face, trunk and extremities since 17 days of birth and multiple boggy swellings in scalp for past three years.

Key words: Leonine facies, Histiocytosis, Multicentric reticulohistiocytosis.

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INTRODUCTION

Reticulohistiocytosis are a rare group of closely related non Langerhans cell histiocytosis which most commonly affects adults. The spectrum of disease ranges from a solitary cutaneous form to multicentric reticulohistiocytosis, a disease with both cutaneous and systemic features. In 1954, the term multicentric reticulohistiocytosis was introduced by Goltz and Laymon [1]. Multicentric reticulo histiocytosis is usually a disease of middle aged and occurs most frequently in females though rarely reported in children. It clinically presents with characteristic leonine facies, coral bead like lesions in nail folds and arthritis mainly destructive with 20% association with internal malignancy.

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CASE REPORT

A nine year old boy born out of non-consanguineous marriage presented to our out-patient department with complaints of multiple raised skin lesions all over body and scalp. History from the mother revealed that the child was delivered by full term normal vaginal delivery with birth weight of 2.5 kg. Antenatal and perinatal period were uneventful. Mile stones were attained normally. The child was normal at birth. From the seventeenth day, child started developing few raised lesions over scalp which progressed to involve the face, trunk, back and both the extremities. No history of itching. These lesions were initially small in size which progressed to increase in size and number and attain the present condition till six years of age. After 6 years, the child had increase in number as well as size of scalp

lesions. The scalp lesions were then infested with myiasis.

There was no history of recurrent infections, fever or lymphadenopathy. No history of loss of weight or appetite. There was no history of joint pains. No history of seizures, jaundice, difficulty in breathing or palpitation. No history of oliguria or hematuria. No history of blurring of vision, redness or pain in the eyes. There was no history of drug intake or topical application. There was no history of trauma. No history of wheezing or asthma. There was no remission of lesions at any time. There was no history of similar complaints in the family members. On general examination child was moderately built and nourished.

Child's mental development was normal. Dermatological examination revealed multiple skin colored papules and nodules distributed over face, pinna, neck, trunk, upper and lower limbs giving a leonine appearance [Figure 1]. Multiple, well defined, boggy nodules of varying sizes ranging from 2×2 cm and 5×4 cm with crusting and serosanguinous discharge with hair loss were seen over the scalp [Figure 2]. There was no tenderness. Peripheral lymph nodes were not palpable. There was no peripheral nerve thickening. Palms, soles, nails and oral mucosa were normal. Systemic examination done was normal. Complete blood count was normal with haemoglobin of 9.9mg/dl. Peripheral smear done was normal. There was no peripheral eosinophilia. Renal function tests, liver function tests, thyroid profile, lipid profile done were within normal limits. Skeletal survey done was normal. Ultrasound abdomen done was normal. Skin biopsy was taken both from skin lesions and from scalp lesions. Both sites revealed a normal epidermis with diffuse sheets of oval to polygonal cells with abundant eosinophilic cytoplasm and dark staining pleomorphic nuclei infiltrating the superficial dermis with few giant cells [Figure 3,4]. With these findings a clinico-pathologic diagnosis of multicentric reticulohistiocytosis was made.

Figure 1. Clinical photograph showing multiple skin colored papules and nodules distributed over face, pinna, neck, trunk, upper and lower limbs giving a leonine appearance.

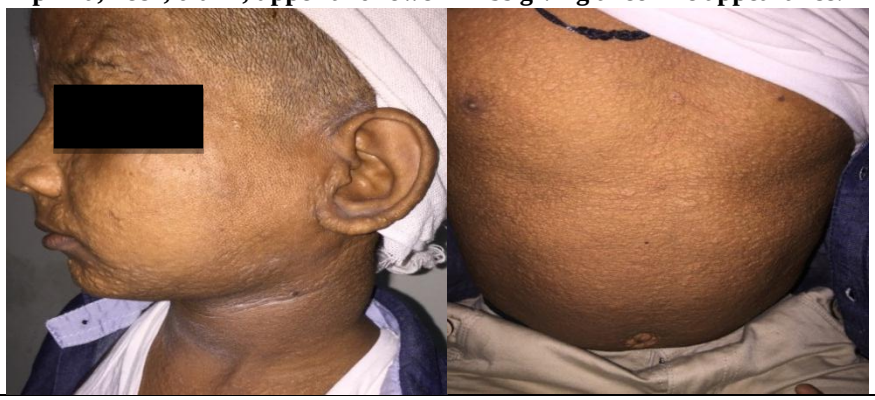


Figure 2. Clinical photograph showing multiple, well defined, boggy nodules of varying sizes ranging from 2×2 cm and 5×4 cm with crusting and serosanguinous discharge with hair loss were seen over the scalp

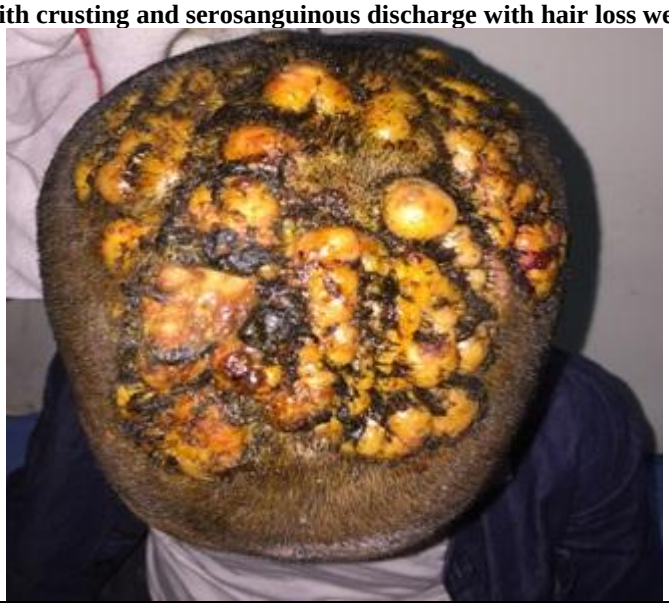


Figure 3. Haematoxylin and eosin (H&E) section of skin lesionshowing sheets of oval to polygonal cells with abundant eosinophilic cytoplasm and dark staining pleomorphic nuclei in 4 x 100 and 10 x 100 (LP) magnification

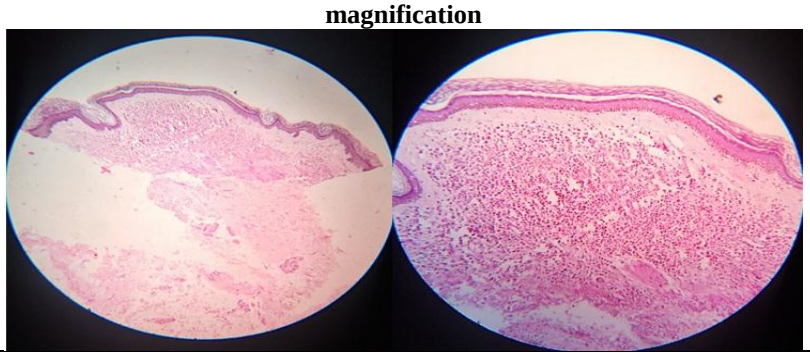
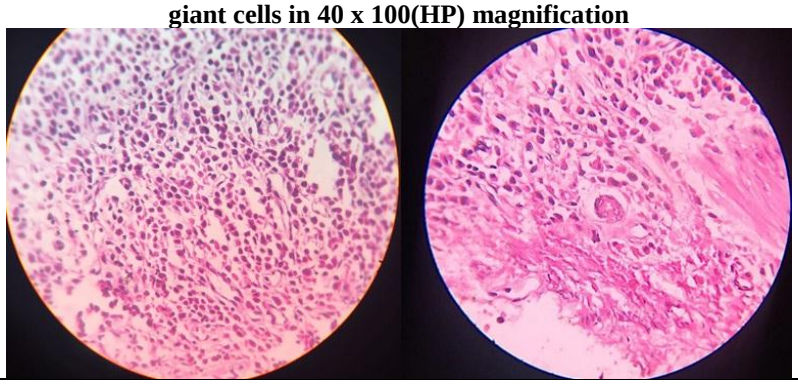


Figure 4. Haematoxylin and eosin (H&E) section of the scalp lesion showing presence of multinucleated giant cells in 40 x 100(HP) magnification



DISCUSSION

Histiocytosis includes a cluster of conditions with histiocytic infiltration of either Langerhans or non-Langerhans type in various tissues. Multicentric reticulohistiocytosis (Syn: Reticulohistiocytosis Granuloma, Lipoid Dermatoarthritis, Giant cell Histiocytoma, Reticulohistiocytoma cutis, Multicentric Giant cell Reticulohistiocytosis) is a rare normolipemic histiocytosis characterized by the presence of an extensive papulonodular cutaneous eruption and a severe destructive arthropathy [2] and 20% risk of internal malignancy.

Though the pathogenesis is unclear, it is a disease of reactive histiocytes. Anecdotally, there is a 33% evidence of contact with tuberculosis patients, though no definite infective etiology has been established. Genetic predisposition is also absent. It is usually a disease of middle age with female predisposition [3]. Rarely, it has been reported in children and adolescents [4]. Classical skin lesions include few to hundred skin colored to yellow or reddish brown papules and nodules that are 1-2mm to several centimeters in diameter predominantly distributed over extensors of forearm, hands, scalp, trunk and ears. Lower trunk and leg involvement is very rare. They may occur in isolation or in clusters or crops with a cobblestone appearance. Lesions are usually asymptomatic but one third may complain of itching. Extensive nodulation on the face can give rise to leonine facies. Coral bead like appearance may be seen as tiny lesions along the nail folds. Xanthalesmatas are common on the eyelids.

Joint pains are a very characteristic feature of multicentric reticulohistiocytosis which differentiates from other xanthomatous conditions. The arthritis is an inflammatory, symmetrical, polyarticular arthritis that affects many joints including the hands, shoulders, knees, wrist and hip. The arthritis is progressively destructive giving rise to mutilating deformities. In severe cases, it causes opera glass appearance where the fingers can be pushed in and out. Mucosal involvement affecting mouth, gingiva, pharynx, larynx and sclera occurs in 50% of cases [5]. Systemic symptoms like fever, malaise, weight loss are occasionally noted. Internal organ involvement may be associated. Death due to cardiac involvement has been reported [6]. Malignancies may later evolve in a case of multicentric reticulohistiocytosis. It may be followed by malignancy of stomach, ovary, uterus, breast, lymphoma or melanoma [7]. The term diffuse cutaneous reticulohistiocytosis is used when the multiple cutaneous lesions appear without the joint and systemic involvement.

Blood investigations are normal except for marginal elevation of erythrocyte sedimentation rate. Histopathology of early lesions shows predominant histiocytes with few lymphocytes and eosinophils.

Large giant cells may later develop with 1-20 nuclei. Old lesions shows the presence of large mono or multinucleated histiocytes with abundant eosinophilic homogenously granular cytoplasm giving a ground glass appearance and fibrosis may start developing which indicates regression of lesions. Immunohistochemistry shows positivity for CD68 and CD45 and negative for CD1, S100, and CD34 and factor XIII a [8]. Immunocytochemistry indicates positive reaction for acid phosphatase, ATPase, lysozyme and alpha 1 antitrypsin [9]. Differential diagnosis includes lepromatous leprosy [10] due to leonine facies. Extensive skin lesions can mimic eruptive xanthomas, juvenile xanthogranuloma, sarcoidosis, rheumatoid arthritis, gout and xanthomatosis. Prognosis is good with quiescence after 7-8 years if not associated with malignancy. Arthropathy and leonine skin changes persists leading to morbidity.

No established treatment is available for this condition. Trials with systemic steroids, cyclophosphamide, azathioprine and cyclosporine have been reported to be useful. TNF alpha antagonists may play a role in cases with extensive arthritis [11].

This case has been reported due to the rare presentation of MRH as leonine facies in a child especially without joint or mucosal involvement.

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CONFLICT OF INTEREST:
The authors declare that they have no conflict of interest.

STATEMENT OF HUMAN AND ANIMAL RIGHTS

All procedures performed in human participants were in accordance with the ethical standards of the institutional research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards. This article does not contain any studies with animals performed by any of the authors.

REFERENCES

1. Goltz RW, Laymon CW. (1954). Multicentric reticulohistiocytosis of the skin and synovia. *Arch Dermatol Syphilol*, 69, 717-30.
2. Toporcer MB, Kantor GR, Benedetto AV. (1991). Multiple cutaneous reticulohistiocytosis. *J Am Acad Dermatol*, 25, 948-951
3. Barrow MV, Holubar K. (1969). Multicentric reticulosis, a review of thirty-three patients. *Medicine (Baltimore)*, 48, 287-305
4. Raphael SA, Cowery SL. Faeber EN et al. (1989). Multicentric reticulohistiocytosis in a child. *J Pediatr*, 114, 266-9
5. Katz RW, Anderson KF. (1988). Multicentric reticulohistiocytosis. *Oral Surg Oral Med Oral Pathol*, 65, 721-5
6. Yee KC, Bowker CM, Tam CY et al. (1993). Cardiac and systemic complications in multicentricreticulo histiocytosis. *Clin Exp Dermatol*, 18, 558-68
7. Snow JC, Muller SA. (1995). Malignancy associated histiocytosis clinical, histological and immunophenotypic study. *Br J Dermatol*, 133, 71-6
8. Luz FB, Gaspar AP, Ramos-e-Silva M et al. (2005). Immunohistochemical profile of multicentric reticulohistiocytosis. *Skinmed*, 4, 71-7
9. Heathcote JG, Guenther LC, Wallace AC. (1985). Multicentric reticulohistiocytosis, a report of a case and review of the pathology. *Pathology*, 17, 601-8
10. Agarwal US, Mathur DK, Jain R et al. (2003). Multicentric reticulohistiocytosis misdiagnosed as lepromatous leprosy. *Indian J Lepr*, 75, 259-62
11. Sellam J, Deslanre CJ, Dubreuil F et al. (2005). Refractory multicentric reticulohistiocytosis treated by infliximab, two cases. *Clin Exp Rheumatol*, 23, 97-9.

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