Alopecia may be the presenting feature in lipoatrophy: A case of centrifugal lipoatrophy

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Abstract

Disorders of fat are uncommon in paediatric practice. It can broadly be classified as disorders with increased fat tissue and disorders with decreased fat tissue. The latter is also known as lipoatrophic disorders. Lipoatrophy could be congenital or acquired. It can be further classified as localized, partial or generalized types. Here we report a case of centrifugal lipoatrophy involving head in a four years and six months old girl for its rarity.

Introduction

Fat atrophy, the loss of subcutaneous adipose tissue can result from many congenital or acquired conditions. These conditions may result in localized, partial or generalized fat involvement. The most common form of lipoatrophy in children is localized lipoatrophy and mostly it is secondary to inflammation or a scarring. In these cases, lipoatrophy may be the only finding[1,2].

Case history

A four years and six months old girl was referred by a paediatric neurologist for evaluation of hair loss. She was born to non consanguineous healthy parents following uneventful pregnancy by an emergency caesarian section due to prolonged labour. Her birth weight was 2.9 kg. Her post natal period was uneventful. Her younger sibling was healthy.

At three years and nine months of age mother noticed a patch of hair loss in left parietal region of scalp. This has gradually progressed rounded shaped alopecia. She was treated with ayurvedhic oil. After five months there was regrowth of hair in the center of patch resulting in ring shaped alopecia. At the same time mother noticed an asymmetry of face and child was brought to the neurology clinic. There was no history of trauma or administration of injections to affected site. There was no past history or family history of autoimmune diseases.

On examination she was an averagely built child. There was a depressed area in her left cheek. Skin over the depressed area was normal. There was no tenderness. Underlying muscle power and function was normal. Her cranial nerve examination was normal. She had a ring of non scarring alopecia over the scalp involving parietal and vertical region. Area of alopecia showed an orange colour hue. In the center of alopecia there were short hairs which were gray in colour. Hair pulling test was negative. Light microscopic examination of hair adjacent to alopecia was normal. Her system examination was normal.

Figure 1. *Lipoatrophy in leftcheeck.*

Figure 2. *Lateral view.*

Figure 3. *Ring of alopecia with orange colour hue.*

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Skin biopsy was done under local anesthesia from the ring of alopecia. Histology showed reduction in hair follicles in epidermis and underneath there was lobular panniculitis with lymphocytes, histiocytes and plasma cells. Inflammation appears to extend into adnexial structures. No granulomata, vasculitis or a neoplastic infiltrate were seen. This appearance was compatible with lobular panniculitis.

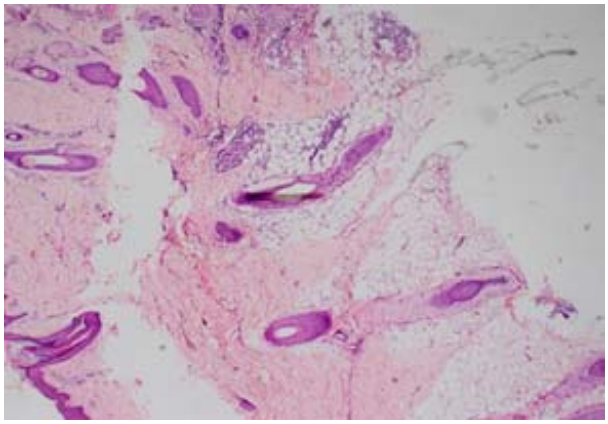

Figure 4. *Reduction in hair follicles and panniculitis (H&E).*

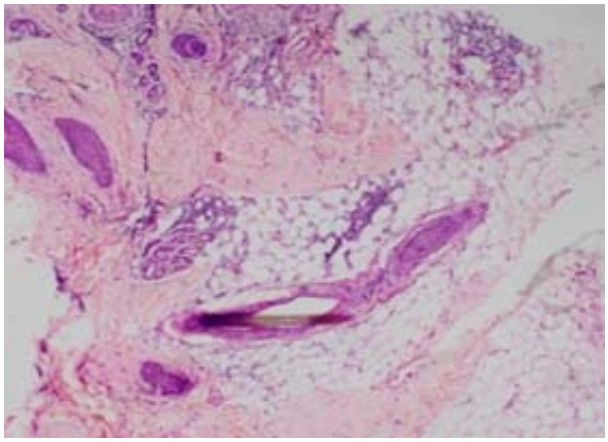

Figure 5. *Inflammation in fat lobules with septal sparing (H&E).*

Discussion

Localized fat atrophy is mainly related to acquired disorders. Congenital disorders are not identified yet[1]. Acquired localized lipoatrophy can be primary or secondary. Primary or idiopathic lipoatrophies are a group of poorly characterized diseases, with focal disappearance of fat usually in abdomen, thigh or ankle[3]. The subtypes of primary lipoatrophy are centrifugal lipoatrophy, atrophy of ankle, semi circular lipoatrophy, annular lipoatrophy and naevoid disorders. Secondary localized lipoatrophy is mainly following inflammatory or scarring process associated with conditions like trauma, injections, infections, neoplasms, connective tissue diseases or thrombophlebitis[1].

Among primary localized lipoatrophies centrifugal lipoatrophy is the variant presenting in childhood. Centrifugal lipoatrophy is common in females with a 2:1 ratio of female to male. In 90% of patients the lesions develop before 8 years of age. This is characterized by abdominal location and most commonly around groin. However few patients are reported with extra abdominal lesions involving head, neck and lumbar region. About one half of patients present with erythematous bluish macules or ecchymosis associated with regional lymphadenopathy. Other half as in index case may only have well defined depression or atrophy of skin. The lesions gradually spread centrifugally leaving central lipoatrophy[1].

Treatment with topical and oral corticosteroids may be beneficial but it did not seem to prevent the extension of the disease. In most of the cases progression of the lesion spontaneously stops before 13 years of age. Persistence of lesions in to adulthood can be associated with angioblastoma[1,4].

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