A Huge Axillary Lipoma In A 3year Old Girl With Concomitant Macrodystrophia Lipomatosa And Syndactyly Of Left Upper Limb - Case Report

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Abstract: Lipomas are the most common subcutaneous soft-tissue tumors of mesenchymal origin. The axillary region is an unusually reported localization for lipomas. Macrodystrophia lipomatosa is a rare cause of congenital macrodactyly, characterized by progressive proliferation of all mesenchymal elements, with a disproportionate increase in fibroadipose tissue. This developmental anomaly may be associated with anomalies like syndactyly. Lipoma in other part of the body has also been reported. We report a huge recurrent cervico-axillary lipoma in a 3-year old girl with with concommitant macrodystrophia lipomatosa and syndactyly of left upper limb. To our knowledge and according to a review of English literature, has not been reported before in

Keywords: Lipoma, Macrodystrophia lipomatosa, Syndactyly, Macrodactyly.

I. Introduction

Lipomas are the most common subcutaneous soft-tissue tumors of mesenchymal origin; the estimated annual incidence is one per 1,000 persons [1] and the overall recurrence rate is 5% [2]. The axillary region is an unusually reported localization for lipomas [3]. Macrodystrophia lipomatosa (MDL) presents as localized gigantism of the hand or foot and comes to clinical attention because of cosmetic reasons, mechanical problems secondary to degenerative joint disease, or development of neurovascular compression due to large osteophytes. [4]. This developmental anomaly is reportedly more common in the foot than in the hand, In this case ring and little finger were involved which is rarely reported in the literature.

II. Case Report

A 3 year old presented with a huge lump in left axilla and unusual enlargement of left hand finger, little and ring [fig1-3]. Physical examination revealed a soft lump measuring 8cmsx4.1cmsx3.5cms. Cervical lymphnodes were not palpable. Lump was nontender subcutaneous with ill defined margins. The little and ring finger were joined together and non tender along with dorsal angulation of the finger [fig----]There were no overlying cutaneous changes or pitting oedema. Patient was otherwise healthy and asymptomatic. FNAC of the axillary lump shows features of lipoma. FNAC from finger revealed only scanty cellularity and biopsy was advised. Adebulking procedure of the finger was performed and extraction of the mesenchymal tissue was done and sent for histopathological examination along with the excised axillary lump.

Histopathology of the axillary lump confirmed it to be lipoma. Histopathology of tissue from the finger shows abundant adipose tissue with some interspersed fibrous tissue. Bony elements were unremarkable.

III. Discussion

MDL is an uncommon congenital, nonhereditary, localized gigantism involving the digits or extremities. This term was first used by Feriz in 1925 to describe unilateral overgrowth of the lower limb.[5] MDL usually presents at birth and may be associated with anomalies like syndactyly, polydactyly, brachydactyly, or clinodactyly.[6] Association with small osseous protuberances, which resemble osteochondromas and lipomas, in other parts of the body has also been reported.[7] The disease is almost always unilateral, with an equal incidence in males and females.[8] The lower limb is more frequently involved than the upper limb. The abnormal area is usually along a specific sclerotome. The second and third digits of the hands and feet are most frequently involved, Involvement of the ring and little finger as seen in our case is is extremely unusual. Adipose tumors account for only 5% of soft-tissue tumors in children, of which 95% are benign [9]. Superficial lipomas are smaller than 5cm in 80% of cases, with only 1% of lesions greater than 10cm in size (5).

Lipomas most frequently affect the upper back, neck, proximal extremities (particularly the shoulder), and abdomen. The axillary region is an unusually reported localization for lipomas; A major concern facing a

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giant lipoma should be to rule out malignancy; however, such a transformation for cutaneous lipomas is exceedingly rare [10].

IV. Conclusion

Huge axillary lipomas are very rare in small children; this benign lesion needs careful histopathological examination though malignant transformation is rare. The differential diagnoses of MDL and macrodactyly include neurofibromatosis, fibrolipomatous hartomas. Neurofibromatosis is seen along nerves. Fibrolipomatous hamartoma (FLH) of nerve is a rare tumor-like condition in which mature fat infiltrates the neural sheath. Pathologically, in FLH, the deposition of fat is within the nerve sheath, while in our case it was present throughout the involved part of the digits.

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[fig1-4]: A huge lump in left axilla and unusual enlargement of left hand little and ring finger.