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Case Report
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MACRODYSTROPHIA LIPOMATOSA: CASE REPORT

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SUMMARY

Lipomatosa macrodystrophia is a not well know pathology worldwide, which is characterized by a rare form of localized gigantism that can be seen from birth. This has a slow and progressive growth pattern in which the increase in size of the mesenchymal tissues and fibroadipose tissue is evidenced. These alterations cause a great clinical, functional and psychological repercussion in affected patients. This is the case of a 14-year-old male patient with lipomatosa macrodystrophia in the left hand who presents clinical and radiological characteristics of this pathology.

CASE PRESENTATION

The case of a 14-year-old male patient from the city of Quetzaltenango, Guatemala, who attends the outpatient clinic of the Hospital Regional de Occidente, with a history of enlargement of the left hand, is presented, referring that since the birth was observed to be larger than his right hand and that in the last two years it had a progressive increase, for which he decided to attend a consultation, in addition to continuous and disproportionate growth, in the last months it was accompanied by mild pain, paresthesia and functional limitation for movements.

On physical examination, a marked increase in size of the left hand is observed compared to its contralateral one, with an increase in soft tissues that predominantly affects the back, palm, first, second and third fingers, in addition to ulnar deviation of the middle finger, functional limitation for the flexion. (Fig. 1)

Figure 1: Lipomatosa macrodsitrophia of the first, second and third fingers of the left hand, in its dorsal view, accompanied by an increase in size at the expense of adipose tissue, likewise an increase in the size of the index finger and volar displacement is observed, difference in size ratio of both hands.

Laboratory tests are requested within them, complete blood count and blood chemistry which are within normal parameters, in addition to arterial and venous Doppler ultrasound of the left upper limb which does not show alterations. An anteroposterior radiograph of both hands was performed, which showed asymmetry of the left hand compared to the right hand, with an increase in soft tissues (Fig. 2)

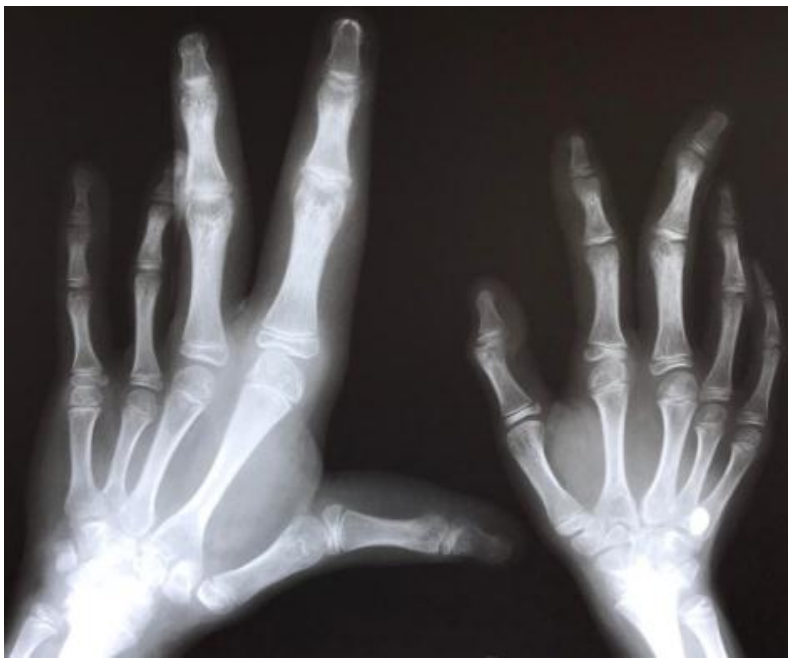

Figure 2: Asymmetry of the left hand is observed compared to the right hand.

An increase in soft tissues and in the length and thickness of the first, second and third metacarpals as well as the phalanges of the fingers mentioned in the left hand is observed.

In the right hand, a slight increase in soft tissue and in the length and thickness of the index and middle fingers is observed, as well as volar deviation of the middle and distal phalanx of the middle finger.

DISCUSSION

Lipomatosa macrodystrophia is a rare non-hereditary congenital disease, characterized by slow and progressive localized gigantism at the expense of mesenchymal and fibroadipose tissue.[1,2,4]

As it is an infrequent malformation, statistical data on its incidence worldwide are scarce, with only 108 cases published in the medical literature.[2,5,13]

This entity was first described by Feriz in 1925 as a rare form of localized gigantism that is characterized by a progressive growth of the mesenchymal elements of the fingers, with a disproportionate increase in fibrofatty tissue, affecting the lower limb.[1,9]

Later in 1960, Golding and later Ranawat in 1968 accepted these terms and applied them to the affectation of the hands.[1,10,11]

The cause of lipomatous macrodystrophia is unknown, however there are several theories proposed for the development of this disease, among them the intrauterine alteration of a growth restriction factor, the alteration of fetal circulation, lipomatous degeneration, errors of the segmentation and trophic influence of a nerve.[7,12]

On clinical evaluation, the localized gigantism that is associated with lipomatous macrodystrophia is evident in most cases from birth. Generally, this involvement is unilateral, where the lower limb and the second and third toes are the most frequently affected and in some less infrequent cases, the hands and upper limbs may be affected.[1,4,5,8]

The involvement occurs most frequently in the distribution of the median nerve in the upper limb and in the plantar nerve in the lower limb.[3]

In radiological evaluation, the first method of choice is the radiological study, however in some cases computerized axial tomography or nuclear magnetic resonance can be used if necessary to complete the imaging diagnosis.

During the simple radiography studies, we can observe the presence of hypertrophy of the soft tissues and excessive growth similar to exostosis at the bone level. Computerized axial tomography can show excessive bone growth and fat proliferation. Nuclear magnetic resonance studies show excess proliferation of non-

encapsulated adipose tissue and fat infiltration at the muscular level.[1,2,5]

To diagnose this pathology, it is necessary to adequately evaluate the family history, the presence of skin manifestations, imaging and histopathological studies.[3]

Among the differential diagnoses should be considered, type 1 neurofibromatosis, Kippel-Trenaunay-Weber syndrome, Proteus syndrome, fibrolipomatous hamartoma, Maffucci syndrome, Ollier disease, Milroy disease, Bannayana-Riley-Ruvalcalba syndrome, dysplasia polyostotic fibrosa, Temtamy syndrome, isolated hemihypertrophy, chronic lymphedema, and lymphangiomatous malformations.[1,2]

The treatment of this entity will depend on the intensity of its affectation, however, several techniques have been described for mild or moderate cases, these treatments range from partial amputations with nail grafts, fingertip flaps, to more severe cases where the final option becomes total amputation.[2,6]

CONCLUSION

Lipomatosa macrodystrophia is a rare entity worldwide, with very few published cases and an unknown incidence. At the national level, no published bibliographic references have been found on this subject, which is why it is important to publicize this pathology, the clinical manifestations, its radiological characteristics and emphasize the aesthetic and psychological affectation that it produces in the lives of our patients.

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