A RARE CASE OF BRACHIAL PLEXUS INFILTRATION

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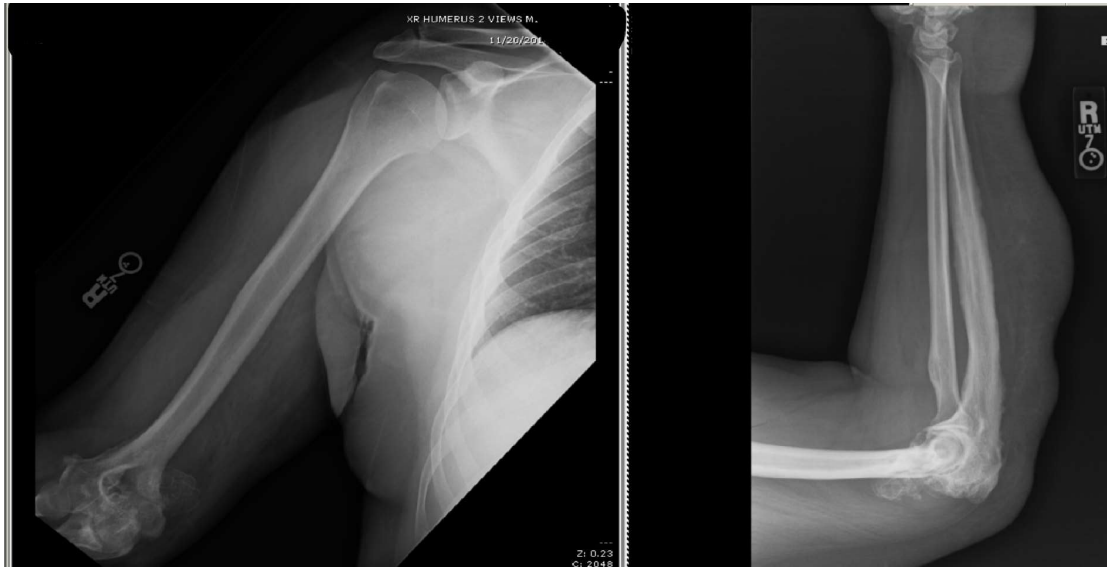
Introduction

Macrodystrophia Lipomatosa Progressiva (MLP) is an uncommon congenital disorder characterized by overgrowth of fibroadipose tissue, usually resulting in a painless enlargement of unilateral fingers or toes. This condicion is usually diagnosed before age of four. There is no gender predominance. It causes important cosmetic and functional problems to patients. Hand or foot involvement is more common than whole extremities. Foot involvement may have serious implications in the life of patients. It may present with difficulties in wearing shoes and increase injuries, affecting the daily life, social development and learning of patients.

There are two forms of MLP: progressive and static types. In the progressive type, the affected areas grow disproportionately. Whereas, in static MLP the affected regions grow proportionately.

Diagnosis is done clinically and using radiographic imaging, such as X-ray, MRI, CT scan and ultrasonography. Management consists of surgical and non-surgical options. However, surgical options are preferred over observation. Usually the type of surgery done is amputation, followed by debulking or reduction. Involvement of the brachial plexus is extremely rare; having serious implications in the functionally and prognosis of patient.



Abstract

A 38-year-old male patient was diagnosed with MLP at the age of 2 after presenting with enlargement of the digits 3, 4 and 5 of his right hand. He underwent some surgeries to debulk the fat, being the last one on June 2018. A month later, on July 2018, he noted a rapidly progressing weakness over a 24 - hour period. Weakness started distally on the right hand, progressing to triceps and shoulder muscles. He reported minimal numbness and no pain. Physical examination was remarkable for atrophy and scapular winging. Right upper extremity (RUE) was found to have weakness, affecting more the distal muscles. All sensation modalities were affected in RUE as well. Deep tendon reflexes were absent in RUE. MRI of the right brachial plexus, done after the onset of this weakness, showed normal nerve roots with expansion of the trunks divisions and cords with lipomatous tissue in between fascicles with massive expansion of the brachial plexus. Progression of MLP was determined and surgical options are currently being considered. Physical therapy was implemented as part of his rehabilitation process. Physicians should be alert of this unusual manifestation of MLP, illustrated in this case, in order to implement treatment therapies as fast as possible to prevent further deterioration and disability in patients.

EMG: Right brachial plexopathy with significant denervation in the upper trunk including the nerves from the roots (to Rhomboids). His previous study showed changes in the muscles supplied by middle and lower trunk.

MRI of right brachial plexus: normal nerve roots with expansion of the trunks divisions and cords with lipomatous tissue in between fascicles with massive expansion of the brachial plexus.

References

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Conclusion

Macrodystrophia Lipomatosa is a rare congenital disorder, but early identification can translate into significant functional outcomes in patients. Sensory and motor impairments can be seen as result of fatty infiltration and grow into norrmal tissue. Therefore, early detection and surgical evaluation could be crucial to prevent further damage. Although most of the time the diagnosis is make during childhood, some patients may present as adults with secondary osteoarthritis and compression of neurovascular elements. Therefore, it should be in the differential diagnosis of patients presenting with weakness or sensory impairments when most common causes are not found.

Our patient had sudden onset of weakness and was found with fatty infiltration into the brachial plexus, which makes it a very unusual case. Treatment options are challenging and surgery is being considered.