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Don't Miss the Target

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A 27-year-old gentleman, with no diagnosed comorbidities, presented to the vascular surgery clinic, referred as a case of lymphangioma, after an ultrasound was done elsewhere. He presented with a history of a progressively enlarging left lateral arm swelling, that he noticed two years earlier, when receiving an intramuscular injection for analgesia, following an injury to the leg, while playing football. Examination revealed [Figure 1] hyper-pigmented spots, Cafe-au-lait macules (CALMs), distributed over his body. The swelling was overlying the lateral aspect of the left arm, measuring about 12 x 7 cm. It was non-tender and firm with limited mobility. There were similar smaller swellings over his chest, back and other limbs. MRI was performed after a clinical diagnosis of Neurofibromatosis [Type 1]. This revealed a classical appearance of neurofibroma's known as the: Target sign, shown in Figure 2. Patient's consent was obtained prior to taking and using his photos and images.

Comment

Neurofibromatosis is an autosomal dominant neurocutaneous disorder. It has two types: Neurofibromatosis type I (NF-1) and neurofibromatosis type 2 (NF-2). NF -1 makes up about 96% of all cases. It affects 1 in 3000 individuals worldwide and presents with cutaneous and non-cutaneous manifestations. Commonly, it presents with Café au lait spots, neurofibromas,

axillary or groin freckling and optic manifestations. Other complications of the disease includes seizures, problems with digestion, psychological burdens and developmental delay including delay in learning to walk and talk. NF-2 usually presents with schwannomas and meningiomas.[1,3] There are three varieties of neurofibroma's: Localized, diffuse and plexiform.

Classically, radiologists have recognized two types of neurofibroma: The discrete mass/nodular form and the diffuse or plexiform variety.[1] On MRI, plexiform neurofibromas present with the pathognomonic "target Sign" depicted as lesions with central hypo - intensity and surrounding hyperintense rim on T2-weighted images, representing fibro collagenous and myxoid components respectively. A "Reverse target sign", on T1-weighted images after IV gadolinium-based contrast administration, is seen as central enhancement with peripheral hypo intensity.[4] Venous malformations mimic the classical "target sign" seen in plexiform neurofibromas, whereby the central hypointense focus represents a phlebolith in a dilated venous channel.

Phleboliths are also noted as a focal hyperintensity on gradient-recalled echo sequences. Other MRI-differentiating features of venous malformations include a hyperintense, lobulated and septated lesion on T2-weighted images & IV contrast enhancement of its cavernous section without arteriovenous shunting on dynamic MR imaging.[4] NF-1 with its various manifestations is a clinical diagnosis and its management requires the involvement of a multidisciplinary team. Patients are referred to surgeons to excise the neurofibroma when the patient is symptomatic with pain, a recent increase in size, the tumor is resulting in a neurological deficit or has a high risk of rupture and bleed. Plexiform neurofibromas, carries significant morbidity due to the way it infiltrates surrounding structures, it's unpredictable growth and its potential to bleed.[5]

Authors' Contribution

ES conceptualized the manuscript. OH, BY, IA and RS performed the literature review. OH, BH, IA, HM, RS and KW drafted the manuscript. ES revised the manuscript. All authors approved the final version of the manuscript.

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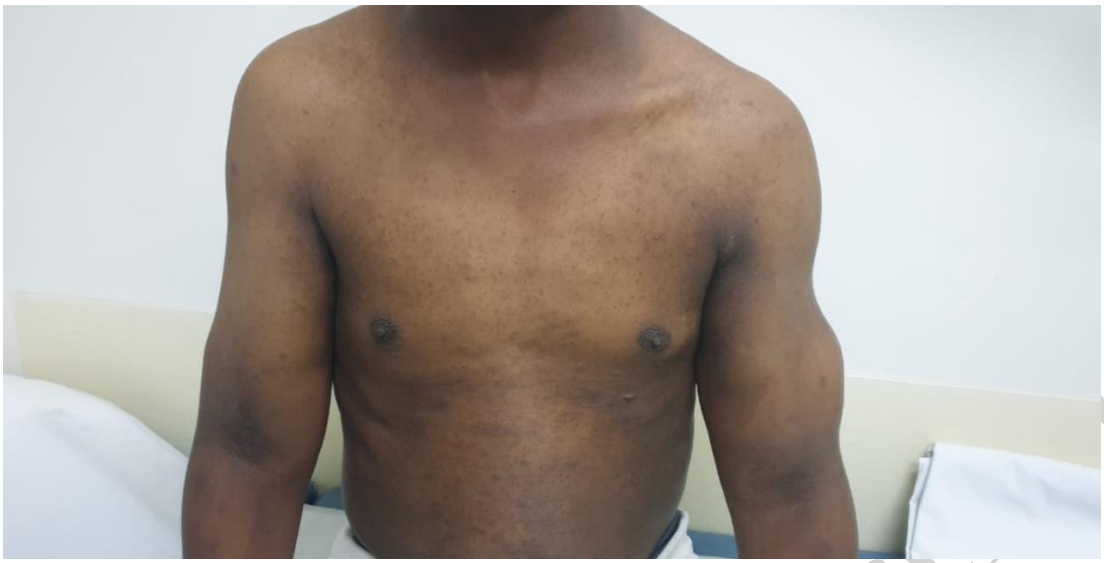

Figure 1: Café-au-lait macules, and multiple swellings in the arms.

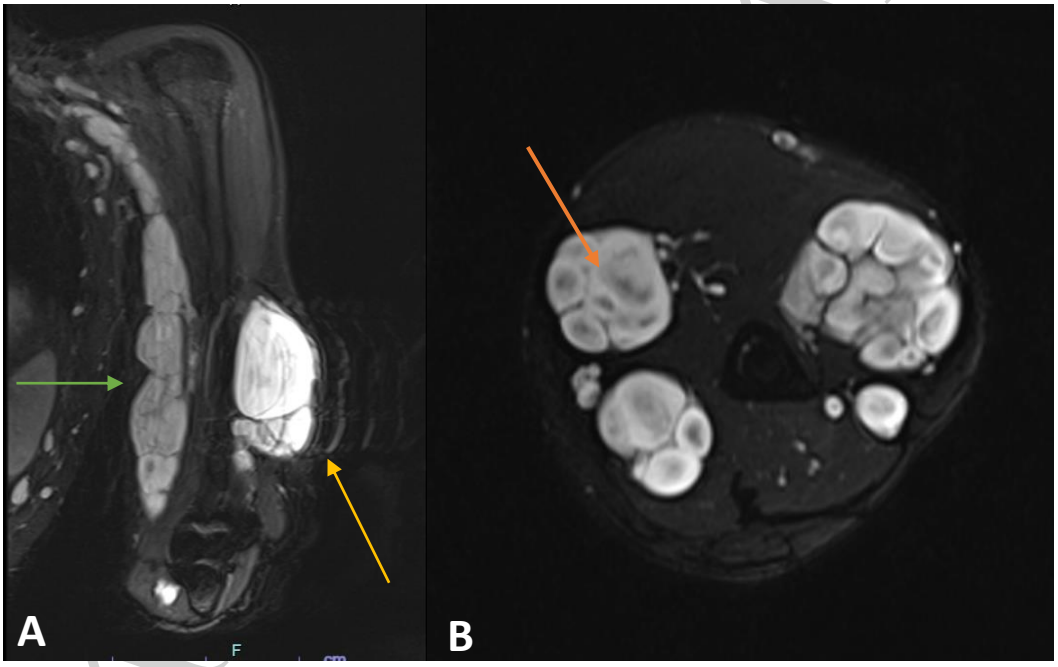

Figure 2A: MRI image, left arm, showing localized lesion laterally (yellow arrow); with plexiform lesion medially (Green arrow); **2B:** MRI axial image, showing the Target sign (Orange arrow). The target sign refers to a central area with low signal intensity, surrounded by a high signal intensity and is sometimes referred to as a bull's eye sign.