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GIANT PLEXIFORM NEUROFIBROMA: SINGLE-STAGE SURGERY WITH HIP DISARTICULATION

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INTRODUCTION:

One of the many different varieties of neurofibroma is plexiform neurofibroma (PN). Although being a benign peripheral nerve sheath tumor, it is invasive and can result in significant morbidities such as pain, disfigurement, neurological deficits, and local organ compromise. It is associated with a risk of malignant transformation to malignant peripheral nerve sheath tumors (MPSNT), the leading cause of mortality in NF1.[1]

REPORTS:

A 39-year-old lady from Kelantan, complained of right lower limb swelling for 20 years with rapid progression in size for the past 3 years and had been bed-bound since then. All her activities of daily living were severely affected. On examination, she is moderately built, presence of multiple café-au-lait spots over the trunk and limbs with varying sizes of more than 1.5cm. Multiple subcutaneous and intra-mascular nodular masses can be palpated from the gluteal down to the foot region. The knee and ankle joints were deformed. Otherwise, her hearing, vision, and spine were normal, and no bowing of other long bones. CT scan showed features consistent with plexiform neurofibroma. A single-stage right hip disarticulation surgery was done. Intraoperatively, the non-standard flap of hip disarticulation was made due to presence of multiple neurofibromas at the gluteal region. We managed to control excessive bleeding by meticulously securing the major and feeding vessels including the venous tributaries and avoiding incision over the tumor mass. Tissue sample was taken and HPE was reported as neurofibroma with no malignant changes. The patient underwent series of post-hip disarticulation rehabilitation in the ward before she was discharged home. Upon her first visit to our clinic, she was able to ambulate with double crutches after 3 years of being bed-bound since 2019.

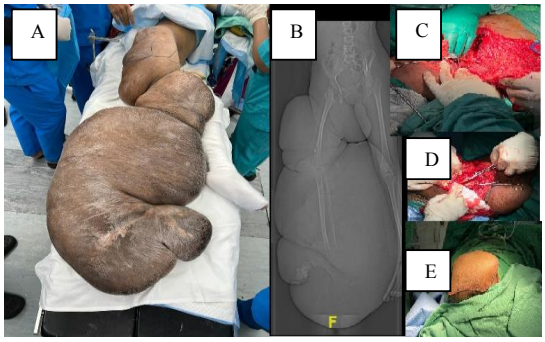

Figure 1: Preoperative image in OT (A), Radiograph (B), Division of anterior hip and groin muscles (C), Division of posterior muscles (D), Right hip disarticulation stump with non-standard flap (E).

CONCLUSION

PN is an uncommon variant (30%) of neurofibromatosis type 1. It arises from multiple nerves as bulging and deforming masses.[1] Treatment for PNs was limited to surgical debulking, which is often associated with subtotal resection, plexiform regrowth, and significant surgical risks (i.e. bleeding/ infection)[2], In our scenario, we believed that a well-planned hip disarticulation resulted in better clinical outcome than multiple debulking surgeries.

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