Gigantic Plexiform Neurofibroma of Extremities: Our Experience

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INTRODUCTION:

Plexiform neurofibroma (PNF) is among the most common complications of NF1 which cause morbidity to patients include pain, functional disturbances, cosmetic problems, and malignant transformation. Surgical excision is challenging and commonly associated with complications of bleeding, wound breakdown and infection. However, patients who have significant disfigurement and functional impairment may benefit from surgical excision. We are sharing our experience in managing 3 patients of PNF of extremities and its complications.

OUR CASES:

We had 3 patients with PNF of extremities who were managed from 2021 and 2022 with 4 excisions done. 1 patient had surgery done to both upper and lower limb, 1 patient had surgery done to right thigh and 1 patient on left lower limb. Indications for surgery were physical disfigurement, inability to walk and difficulty in maintaining perineal hygiene. All procedures were at least subtotal excision (50%-80%).

RESULTS:

2 patients had minimal post operative complication of delayed wound healing which required frequent outpatient dressing that healed a 1 month post operative. The other 2 patients had severe complications, 1 required transfusion with wound breakdown requiring debridement and prolonged hospital stay for 56 days. The other patient had bleeding, post-operative ICU care for 14 days for severe pulmonary embolism and required 2 debridement for wound breakdown. She had a hospital stay for 59 days. There was no mortality in our series. All complications were treated successfully, and patients had improved quality of life postsurgery.

Figure 1: Pre-operative



Figure 2: Post-operative



DISCUSSIONS:

Surgical excision is a treatment option for selected patients with PNF which offers promising outcomes for patients. Surgeons should be mindful of all complications that might occur during and after the procedure as mentioned in our series and prepare patients thoroughly prior to surgery.

CONCLUSION:

We believe this type of surgery should be done in highly specialized units that have all the post operative support.

REFERENCES:

1. "Plexiform neurofibromas - PubMed." 26 Mar. 1999, https://pubmed.ncbi.nlm.nih.gov/10469434/.