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A Rare Presentation of Solitary Giant Neurofibroma

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Abstract

Neurofibroma is common benign peripheral nerve sheath tumor. It may occur anywhere in the body. Including skin and soft tissue, nervous system muscle and skeleton and visceral organ. Solitary neurofibroma is rare in the giant type. We are presenting a rare case of solitary giant gluteal neurofibroma, which was first confirmed with true cut biopsy which was completely excised afterwards and histopathological confirmation was performed. This case had no generalised neurofibromatosis. Presentation, diagnostic features, imaging and treatment are represented here.

Keywords

Neurofibroma; Skin; Tumor; Sciatic Nerve Injury

Introduction

Neurofibromas are common benign peripheral nerve sheath tumor. They often appear as a soft, skin coloured papule or soft subcutaneous nodule. Overlying skin is often hyper

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pigmented. Neurofibroma tend to be soft almost gelatinous in consistency. It may occur anywhere in the body Including skin and soft tissue,nervous system , muscle and skeleton and visceral organ. Most of neurofibromas are solitary.

Neurofibroma arises from cells of peripheral nerve including schwan cells, perineurial cells and fibroblasts. Peripheral nerve sheath tumors can arise within the dura as well as along the peripheral course of nerve. Peripheral nerve sheath tumors are also associated with two forms of neurofibromatosis.

Neurofibroma can present as -

1. Localized mass-most commonly as cutaneous neurofibroma or in peripheral nerve as solitary neurofibroma.Localized can occur anywhere in the body with a predilection for the trunk, head/neck as ill-defined indurated plaques with thickened skin

2. Diffuse

3. Plexiform-Infiltrative lesion growing within and expanding a peripheral nerve. Plexiform tumors result in significant neurologic deficit with potential of malignant transformation

Presence of multiple neurofibroma or plexiform neurofibroma suggest the diagnosis of neurofibromatosis type 1 [1].

Majority of neurofibroma occurs sporadically and have an extremely low risk of malignant transformation.

They affect men and women equally. Usually complete excision of the tumor is curative. Neurofibroma has tendency for local recurrence after excision.

Case Report

An 8 years old boy, presented with painless, firm swelling over left gluteal region for the period of 8-9 months. On initial examination there was a firm swelling noted on left gluteal region of size 10x8 cm, without any constitutional symptoms [2,3]. Patient had underwent incisional biopsy outside hospital but reports were not contributory. On physical examination, there was healed scar mark over left gluteal region, normal temperature over the swelling, firm to hard in consistency, immobile, overlying skin is free ,there was no tenderness, margin is smooth and defined, normal range of motion of left hip, no distal neurovascular deficit.

Blood investigation were within normal limits.

On X-ray examination-There was no evidence of bony involvement.

MRI(CEMRI, 17/9/2019)-showed a large lobulated heterogeneous mass lesion is seen in left gluteal region displaying hyperintense signal on T2W1, intermediate signal on T1W1 and shows prominent heterogeneous enhancement on post-contrast scan, the mass measures 10.4x8.9x7.4 cm in dimension (Fig. 1).

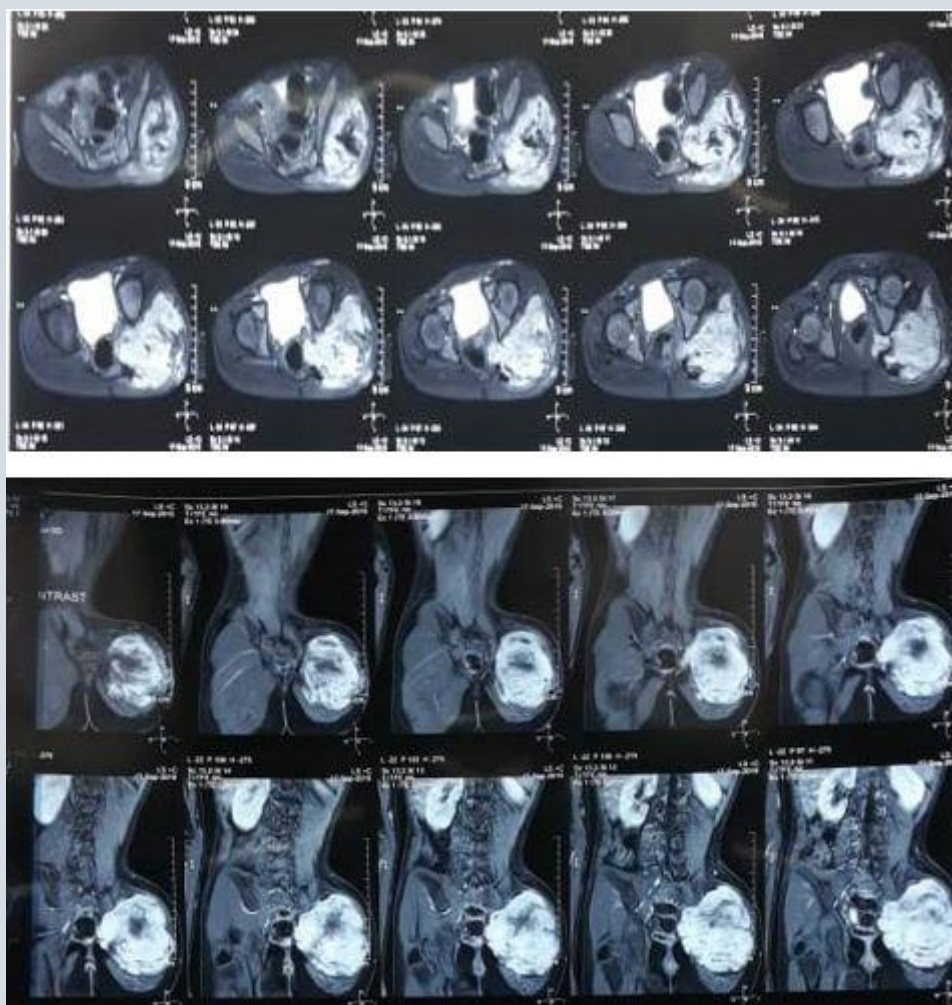


Figure 1: MRI scan results.

We planned for truecut biopsy of the mass and reports were suggestive of neurofibroma. After that, patient underwent excisional biopsy of mass and mass sent for HPE.

Procedure-Patient under adequate anesthesia, positioned in right lateral position, part was painted and draped, incision line incorporated the previous scar, gluteus maximus retracted, sciatic nerve isolated and mobilized carefully. The whole extension of the tumor was approached from 360 degree angle (Fig. 2). Its root appeared to arise through sciatic notch and

it encircled around the sciatic nerve and tumor is made free from its bed all around and whole mass excised (Fig. 3) [3-5].

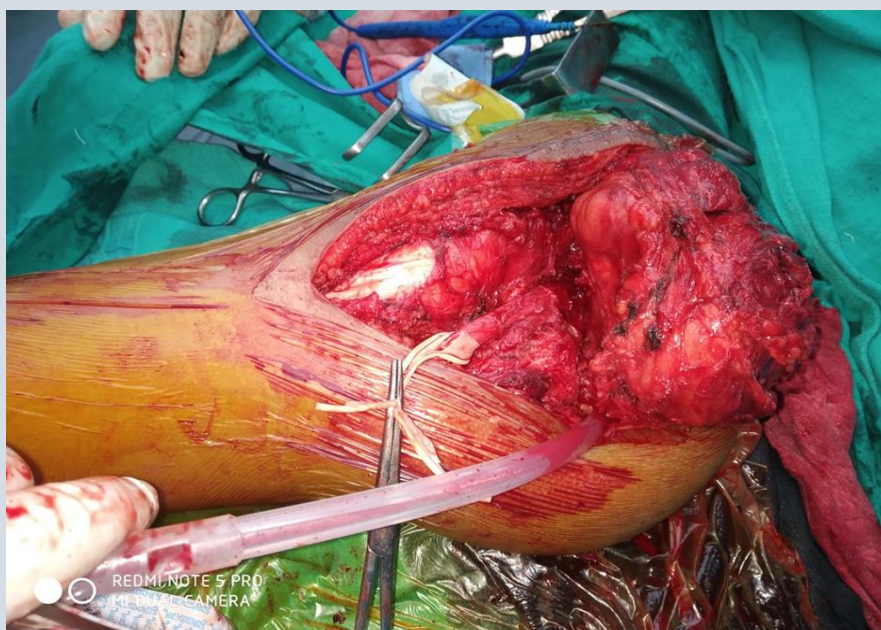


Figure 2: Intra operative picture showing giant neurofibroma in gluteal region.



Figure 3: Giant encapsulated tumor after resection, having no adhesion to adjacent tissue or vascular involvement.

Post operatively, patient had no symptoms /signs related to sciatic nerve injury, wound healed with primary intention (Fig. 4), discharged after 10 days and was on regular follow up. Hip movements are normal. Wound healed with primary intention without features of recurrence.



Figure 4: Postoperative picture showing healed scar mark.

HPE studies confirmed a diagnosis of neurofibroma

Gross description-Circumscribed grey white tumor measuring 11.5x10x6.5 cm, cut section homogenous white, no necrosis, hemorrhage noted, P/E-section A1-A5 tumor with capsule.

Microscopic description-multiple section examination shows a well circumscribed tumor composed of spindle cells. The tumor cells are arranged in sweeping fascicles and sheets cells are having elongated spindle shaped wavy nuclei, indistinct nucleoli and scanty cytoplasm. No nuclear atypia seen. Mitosis is infrequent. Focal myxoid changes noted [6].

Immunohistochemistry-Tumor cells shows focal S100 positivity (Fig. 5).

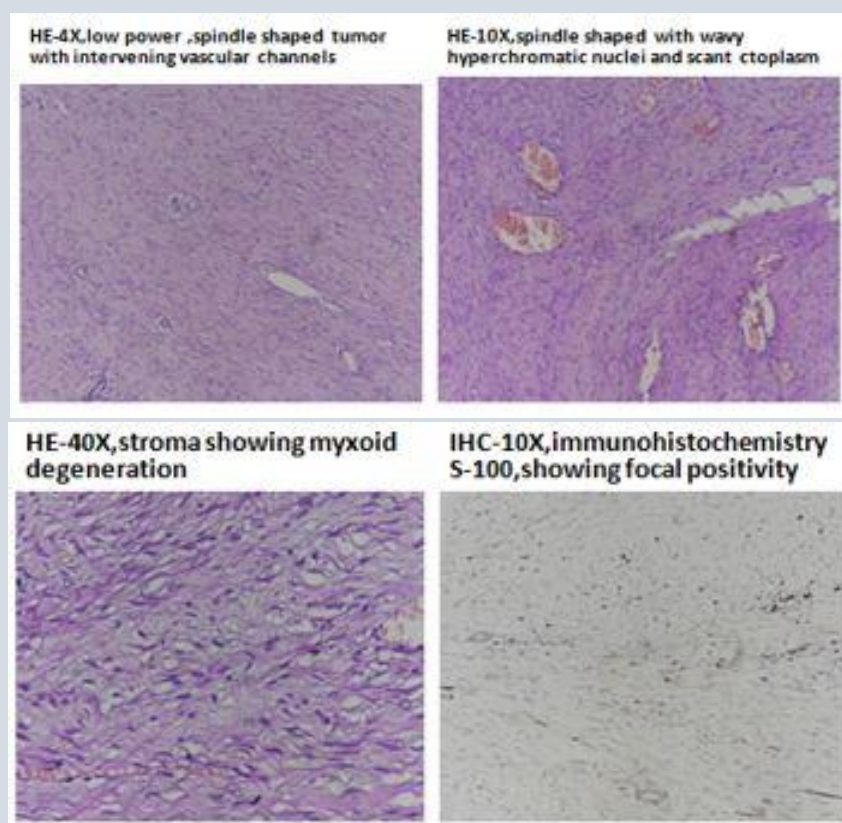


Figure 5: Tumor cells shows focal S100 positivity.

Discussion

We have presented a case of solitary giant neurofibroma in the gluteal region which was rare in size and location. Neurofibroma can be cured by resection. The neurofibroma can recur after surgical excision. However surgical intervention is the most effective means of treatment [7].

Conclusion

This case report validates the use of surgical intervention to excise the lesion is curative and it restores appearance and improves quality of life.

Consent for Publication

Written informed consent was obtained from the parent for publication of this case report and any accompanying image.

Conflict of Interest

The authors declare no conflict of interest.

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