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GIANT SOLITARY NEUROFIBROMA OF THIGH

General Surgery

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KEYWORDS

Neurofibroma, Nerve Sheath tumor, Neurofibroma

INTRODUCTION:

Neurofibromas are benign nerve sheath tumors in the peripheral nervous system. They evolve anywhere along a nerve from the dorsal root ganglion to terminal nerve branches. [1, 2] Neurofibromas are commonly, but not always associated with neurofibromatosis type 1, especially the solitary type.

Case Report:

A 25 year man farmer by occupation presented to us in OPD with complains of swelling in right thigh posterior aspect since three years which he noticed incidentally while working. The swelling was painless and gradually increased in size from being less than few centimetres to currently involving the upper part of posterior aspect right thigh. There was no associated limitation of movement though he felt some discomfort and dragging pain lately while walking. He has reported no weight loss and had no similar mass in any other part of his body. He had no significant past history. No history of similar illness in any family member was noted.

Examination revealed a well-built man with no visual or auditory impairment, no subcutaneous nodule, café- au- lait spots or axillary or groin freckling. He had an elliptical, poorly defined swelling at the posterior aspect of upper half of his right thigh. The mass measured 14 x 8 cm. It was firm, non-tender and within the intramuscular compartment with normal overlying skin. The mass was non-pulsatile with negative emptying sign. There was no associated peripheral lymphadenopathy and no neurovascular deficits were noted. A clinical diagnosis of soft tissue tumor was made

Lab investigation showed a normal blood count and urinalysis. Plain radiograph of his femur showed normal bones and a huge soft tissue density in the posterior compartment. FNAC from the mass showed features suggestive of neurofibroma. MRI revealed an 18 x 10 x 6 soft tissue mass, discrete, originating from the adductor magnus and displacing the surrounding muscles of the medial and posterior compartment, and extending in the posterior compartment of the thigh. The tumor showed an iso-intense signal on T-1 weighted images and a high signal intensity on T-2 weighted images suggestive of myxoma.

Excision of the mass was done under spinal anaesthesia and was sent for histology. Intra-op findings included a well circumscribed soft tissue mass (Figure 1)18 x 8 x 6 cm with thick fibrous capsule with multiple cystic and solid regions in the posterior compartment of the thigh lying along the adductor magnus and semimembranosus but separate plane of dissection could be identified between the muscle fibres and the mass. The whole mass could be excised en-bloc(Figure 2). Following satisfactory haemostasis, surgical field was closed in layers and pressure dressing applied. The patient tolerated the procedure well and was discharged the following day.

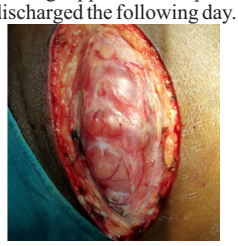

Figure1. Surgical exposure of neurofibroma of thigh

Figure 2. Resected specimen of giant neurofibroma

Pathological findings confirmed neurofibroma diagnosis and revealed tumor free margins on all sides.

DISCUSSION:

The primary treatment option for giant neurofibroma has been surgery (3). Removal of neurofibromas is considered difficult because they are known to cross tissue boundaries. However, besides pain, neurofibromas are sometimes removed due to the possibility of malignant transformation.

Solitary giant neurofibromas in uncommon sites have been reported on the oral cavity, neck scalp, presacral space, paraspinal space, retroperitoneum, labia and appendix (3).We reported a rare case of giant solitary neurofibroma of right thigh in a young male that could be surgically excised with relative ease by identification of the right plane of dissection that exists even in large neurofibromas.

CONCLUSIONS:

Plexiform neurofibromas are benign tumors originating from subcutaneous or visceral peripheral nerves, which are usually associated with neurofibromatosis type 1.Giant neurofibromas are very difficult to manage surgically as they are extensively infiltrative and highly vascularized.These types of lesions require complex preoperative and postoperative management strategies.

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