



ISSN: 0975-833X

## CASE REPORT

### NEURALFIBROLIPOMA AT A RARE SITE-A CASE REPORT

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#### ARTICLE INFO

##### Article History:

Received 05<sup>th</sup> February, 2015  
Received in revised form  
19<sup>th</sup> March, 2015  
Accepted 03<sup>rd</sup> April, 2015  
Published online 25<sup>th</sup> May, 2015

##### Key words:

Neural fibrolipoma,  
Median nerve.

#### ABSTRACT

Neural fibrolipoma is a tumor like lipomatous process that involves principally the volar aspects of hands, wrists and forearms of young persons. It usually manifests as a soft yellow growing mass consisting of proliferating fibrofatty tissue surrounding and infiltrating major nerves and their branches. Median nerve is the most frequently involved nerve. Other less frequently involved nerves are ulnar, radial, brachial plexus, superficial peroneal nerve, inferior calcaneal nerve and medial plantar nerve. An 18 year old male presented with swelling over right lateral gluteal region. Histopathological examination of the excised specimen showed features of neural fibrolipoma. To our knowledge no reported case of neural fibrolipoma at this site has been reported so far. We report a case of neural fibrolipoma in lateral cutaneous branch of iliohypogastric nerve.

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#### INTRODUCTION

A neural fibrolipoma is a rare, benign lesion first reported in 1953 (Mason *et al.*, 1953). Also known as a fibro fatty overgrowth, perineurallipoma, intraneurallipoma, lipomatous hamartoma (Mason *et al.*, 1953; Terzis *et al.*, 1978; Mikhail *et al.*, 1964), neural lipomafibromatous hamartoma, neurolipomatosis. Although neural fibrolipoma was first described in 1953 (Mason *et al.*, 1953), less than 100 cases have been documented so far in the literature (Silverman *et al.*, 1985; Toms *et al.*, 2006; Patil *et al.*, 2009; Razzaghi *et al.*, 2005). While most commonly found in the median nerve, studies have reported the lesion at other sites such as the radial, ulnar, sciatic, and plantar nerves (Gouldesbrough *et al.*, 1989; Herrick *et al.*, 1980; Johnson *et al.*, 1969) and in the lungs (Taniyama *et al.*, 1995). Most cases of neural fibrolipoma occur within the first three decades of life (Silverman *et al.*, 1985).

#### Case Report

18 year-old male presented to the surgical department with one year history of swelling over right lateral gluteal region that was insidious in onset and gradually progressive. The swelling was extending from right lateral gluteal region to right iliac region, was painless, non-tender and there was no associated difficulty in walking. On examination, a 20×13 centimeters,

soft to firm, non-tender swelling was present in the gluteal region. The overlying skin was normal. Motor and sensory examination of the leg was unremarkable. FNAC of the swelling was non conclusive and revealed fragments of adipocytes with muscle fragments. Magnetic resonance imaging (MRI) showed a large hyperintense area along right iliac crest on T1W images in gluteal region along right iliac crest. Same intensity area was seen along right iliacus muscle same side.

The area was hyperintense on T2W images and flair images. Few hypointense foci were seen in between. The MRI impression was that of a diffuse cutaneous lipoma right iliac crest with associated neurofibroma in right iliacus muscle and adjacent skin (Fig 1). An excisional biopsy was performed under general anaesthesia via incision given over swelling in the gluteal region extending to right flank. Intraoperatively the mass was soft, consisting of fatty tissue and was infiltrating cutaneous nerves. We received a grey-white to yellow well circumscribed soft tissue mass measuring 16×10×3 centimetres in size.

The cut surface had homogenous fatty appearance (Fig 2). Microscopical examination showed mature adipose tissue interspersed with fibrous tissue surrounding the individual nerve fibres and infiltrating the epineurium and perineurium of the nerve. It was accompanied by concentric thickening of perineurium and the perivascular fibrous tissue. Some portions of the affected nerve showed a pseudo-onion bulb formation (Fig 3 a and b).

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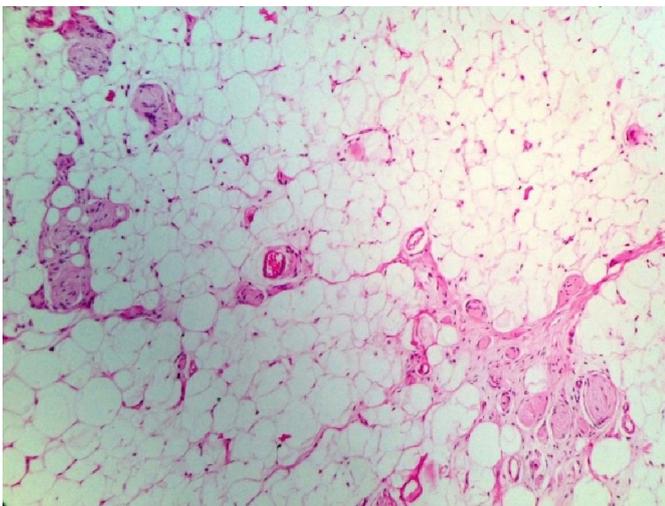
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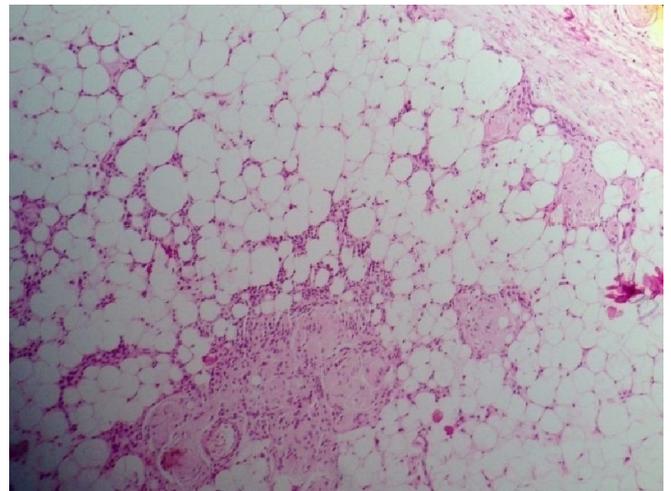
**Fig. 1.** A Coronal T2 weighed images demonstrates hyperintenselipomatous area with hypointense with interspersed hypointense areas



**Fig. 2.** Gross examination showing a grey-white to yellow well circumscribed soft tissue mass with a homogenous fatty appearance on cut section



**Fig. 3a.** Photomicrograph showing adipose tissue surrounding nerve bundles (H&E X 100)



**Fig. 3b.** Photomicrograph showing adipose tissue, fibrous tissue and nerve elements (H & E X 400)

## DISCUSSION

Neural fibrolipoma has also been called fibrolipomatous nerve enlargement, lipofibroma, fibrofatty overgrowth, neural fibrolipoma and neurilipoma (Tse *et al.*, 2011). Fibrolipomatoushamartoma (FLH) is an unusual benign tumour composed of hypertrophied fibroadipose tissue intermixed with neural tissue (Parihar *et al.*, 2014). It typically presents in childhood or early adulthood. FLH affects the median nerve in majority of the cases; this predilection remains unexplained. (Patil *et al.*, 2009; Razzaghi *et al.*, 2005). Other less frequently involved nerves are the ulnar nerve, radial nerve and brachial plexus in the upper extremity (Toms *et al.*, 2006) and rarely, the superficial peroneal nerve (Bibbo *et al.*, 1994), inferior calcaneal nerve (Zeng *et al.*, 2012), medial plantar nerve (Van *et al.*, 2003) and sural nerve (Parihar *et al.*, 2014) in the lower extremity. Our patient was a young male who presented with the involvement of lateral cutaneous branch of iliohypogastric nerve. The natural course of the lesion is a gradual increase in the size along with symptoms of compressive neuropathy (Ha *et al.*, 2012). In our patient, the swelling was asymptomatic without any neurological deficit.

In fibrolipomatoushamartoma the mature adipose tissue infiltrates the perineural and epineural compartments of the involved nerve and is admixed with fibrous tissue, which dissects between and separates individual nerve bundles. Atrophy with concentric perineural fibrosis causes thickening of the nerve fascicles. The affected nerve may also show other changes such as perineuralseptations, microfascicle formation, and pseudo-onion bulb formation (Tse *et al.*, 2011). Other intraneural tumours which need to be distinguished from FLH are intraneural lipoma, neurilemmomas and neurofibromas (Parihar *et al.*, 2014). The treatment of the fibrolipomatoushamartoma remains controversial. Surgery is not recommended in all cases, due to detrimental effects on motor and sensory functions, apart from having the potential to cause postoperative neurogenic pain. Moreover, in this infiltrative process, the optimal resection margin can be difficult to achieve. Therefore conservative treatment coupled with decompression of compromised nerves is generally

adopted (Razzaghi *et al.*, 2005; Tse *et al.*, 2011). We report this case because of the rarity of the lesion and unusual site of presentation.

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