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CASE REPORT

Infection in a Plexiform Neurofibroma

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ABSTRACT

Plexiform neurofibroma is a typical sign of neurofibromatosis type 1, although the condition is not diagnostic. These large lesions are associated with neurological deficits, affect cosmesis, and have a predisposition for malignant change. Infection within a plexiform neurofibroma is not a common complication and has not been well described in the literature. This report describes the use of magnetic resonance imaging findings of a patient with an infected plexiform neurofibroma, the diagnosis of which was subsequently confirmed after surgery.

Key Words: *Infection; Magnetic resonance imaging; Neurofibroma, plexiform; Neurofibromatosis 1*

中文摘要

蔓狀神經纖維瘤感染

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蔓狀神經纖維瘤雖然不是用來診斷神經纖維瘤病一型的特徵，它是神經纖維瘤病一型的典型表現。這些較大的病變通常伴有神經功能缺損，不但影響外觀，還有機會惡變。蔓狀神經纖維瘤受感染的病例罕見，在文獻中極少記載。本文報告一例神經纖維瘤感染患者的磁共振成像表現，病人於術後確診。

INTRODUCTION

Plexiform neurofibromas (PNFs) are slow-growing tumours, which may be present at birth or become apparent later in life. Malignant transformation of a PNF is a well-established complication. Infection in a PNF, which has not undergone any type of intervention before, has not been well established. This report is of a middle-aged man with neurofibromatosis type 1 (NF1) and an infected PNF, without any evidence of malignant transformation. The magnetic resonance imaging (MRI) findings and the possible causative factors for the superimposed infection are discussed.

CASE REPORT

A 54-year-old man with known NF1 had multiple peripheral neurofibromas of varying sizes, including a large PNF on his right buttock. He presented in January 2010 with fever and pain in the right buttock PNF, which had increased in size during the previous 2 weeks. The right buttock swelling was warm and tender, but not fluctuant. His blood profile revealed mild leukocytosis. Urgent MRI demonstrated multiple T2-weighted hyperintense superficial nodules with contrast enhancement, in keeping with cutaneous neurofibromas. The PNF on his right buttock measured 12.2 x 7.5 cm, and had a similar T2-weighted hyperintensity

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(Figure 1) and diffuse contrast enhancement (Figure 2), with a few cystic foci within. The lesion was superficial with thickened and oedematous overlying skin (Figure 3), while the underlying muscles and bones were unremarkable. Given these appearances, the preliminary diagnosis was an infected PNF. The patient was given a course of antibiotics (amoxicillin 500 mg and clavulanate 125 mg orally 3 times a day for 10 days), which improved his condition clinically. This was followed by debulking surgery 6 weeks later at the patient's request. Histopathology revealed granulation tissue without evidence of sarcomatous transformation. The patient had an uneventful recovery and was well at follow-up 8 weeks later.

DISCUSSION

PNF, also known as 'solitary neurofibroma' or 'solitary neural sheath tumour' is a typical feature of NF1, although the condition is not diagnostic. PNF is seen in approximately 20 to 30% of patients with NF1,[1,2] but it may occasionally occur without any other signs of NF1.[3] The cells composing PNF are thought to be similar to dermal neurofibromas, but have an expanded extracellular matrix. The larger cranial nerves and exiting cervical or thoracic spinal nerves are frequently affected. PNF tends to enlarge along the length of these nerves and may show further extension into the peripheral branches.[4]

Continued tumour growth is a common cause of morbidity in PNF.[1] Chronic pain, numbness, and

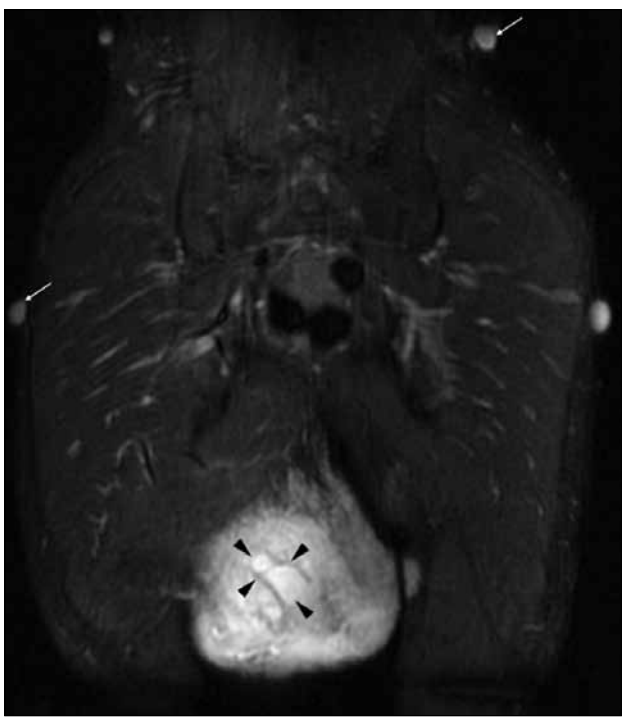

Figure 1. Coronal T2-weighted fat-saturated magnetic resonance image showing hyperintensity within the plexiform neurofibroma with a few cystic foci (arrowheads). Multiple peripheral neurofibromas are also evident (arrows).

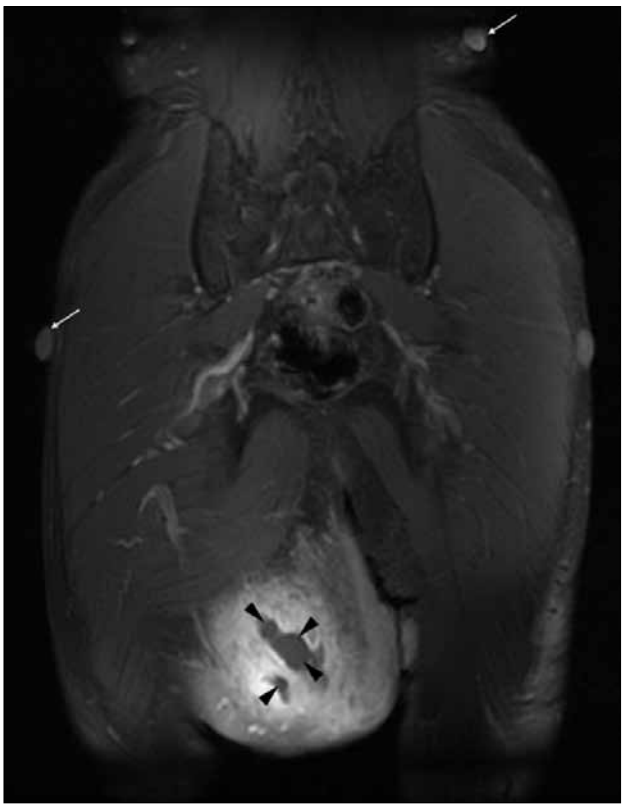

Figure 2. Coronal T1-weighted post-gadolinium fat-saturated magnetic resonance image showing avid contrast enhancement with rim-enhancing areas of central necrosis suggesting abscesses (arrowheads). Less intense enhancement is noted in the peripheral neurofibromas (arrows).

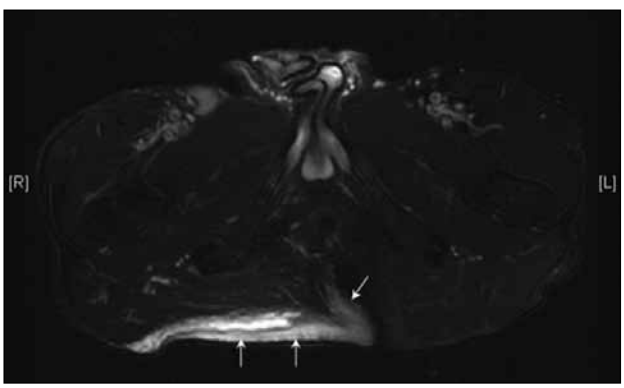

Figure 3. Axial T2-weighted fat-saturated magnetic resonance image showing thickening of the skin overlying the plexiform neurofibroma with increased signal intensity (arrows). The underlying muscles and bones appear unremarkable.

spinal cord compression with neurological deficits are well-known complications.[5,6] Intra-orbital lesions may compress the optic nerve causing loss of vision. Superficial lesions occurring at exposed sites have aesthetic implications, and can lead to social anxiety and psychological stress. In contrast, deep-seated PNF can remain silent for years, and are usually discovered incidentally during imaging for an unrelated condition.[5] Approximately 5 to 10% of affected people develop malignant peripheral nerve sheath tumours.[4,7] Mautner et al[4] proposed that the inhomogeneous appearance with patchy contrast enhancement of PNF on MRI scan is due to necrosis and haemorrhage, and is suggestive of malignant transformation. However, a relatively homogeneous appearance on T1- and T2-weighted images before and after contrast enhancement suggests a benign lesion.

Infection in a PNF that has not undergone any prior interventions has not been commonly cited as a complication. To the authors' knowledge, this is the first report of infection occurring within a PNF. It is possible that the location of PNF at the gluteal region may be a predisposing factor for infection due to repeated trauma and friction from sitting. The proximity to the anal canal may be another contributing factor. Generally, increased size or pain raises the possibility of malignant transformation. This may have prompted the referring clinician to request an MRI scan on an emergency basis. However, when these symptoms progress over a short interval and are associated with signs of sepsis, an infective aetiology is more likely than malignant transformation.

Surgery for PNF has been reserved primarily for patients with progressive enlargement and resultant morbidity. Owing to the rich vascular supply, profuse bleeding during surgery is a known complication.[8] Fortunately, surgery for this patient was not complicated by haemorrhage because of the superficial nature of the lesion and the necrosis and abscess-like contents. In view of the large dimensions of the PNF, incomplete resection with recurrence was another postoperative consideration.[8] This patient, however, remained free from recurrence 6 months after discharge from hospital. Alternative treatments using antihistamines, antiangiogenic drugs, and biological agents are largely anecdotal.[5]

In conclusion, although an increase in size or development of pain in a PNF mandates further examination to exclude malignant transformation, PNF in a susceptible location may be secondarily infected.

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