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The effects of simple trauma on patients with cervical spine neurofibromatosis: two case reports

Received: 27 May 1995
Revised: 18 December 1995
Accepted: 3 January 1996

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Abstract Von Recklinghausen's disease of the cervical spine is rare. Spinal deformities appear to occur only in peripheral neurofibromatosis as opposed to central neurofibromatosis, and such deformities include non-dystrophic and dystrophic changes. We describe two patients with neurofibromatosis of the cervical spine who were subjects of simple trauma, one of whom demonstrated dystrophic and the other non-dystrophic changes. The first presented with acute upper cervical

spine instability and subluxation, the second with cervical myelopathy. The purpose of this article is to heighten the physician's awareness that such patients have inherent pathology in the cervical spine such that simple trauma can have serious neurological consequences.

Key words Neurofibromatosis · Cervical spine trauma · Cervical spine instability · Cervical myelopathy

Introduction

The incidence of cervical spine deformities secondary to neurofibromatosis is unknown. It has been reported that 2% of patients with scoliosis will have neurofibromatosis [5]. The incidence of spinal deformities within a neurofibromatosis population ranges from 2 to 36% [5] and the cervical spine deformity present is usually a kyphosis.

The majority of patients with peripheral neurofibromatosis (NF 1) of the cervical spine are asymptomatic [1, 7]. We present two case reports of patients with NF 1 affecting the cervical spine, both of whom were symptom free prior to simple trauma, and who, between them demonstrate the two varieties of spinal deformity.

Case reports

Case 1

A 21-year-old factory worker slipped on an icy surface and sustained minor injuries to her occiput. Three days later, she com-

plained of increasing neck pains associated with right C5 root dysaesthesia and paraesthesia. On examination, she had posterior mid-line tenderness at C3/4 and objective paraesthesia along the right C5 dermatome. No other abnormal neurology was found. It was also noted that she had a 6 × 3-cm "café au lait" spot over her trunk and the possibility of neurofibromatosis was raised. This was later confirmed as neurofibromatosis type 1 (NF 1). There is a family history of neurofibromatosis: both her mother and brother have NF 1. Cervical spine radiographs showed an alarming C3/4 subluxation with a 40° kyphotic deformity (Fig. 1).

CT scans revealed abnormalities of the cervical vertebral bodies and facet joints, but no evidence of a fracture was seen. Magnetic resonance imaging (MRI) showed generalised dystrophic cervical vertebrae with dural ectasia, but no evidence of cord compression was noted (Fig. 2). At C4/5, a large intradural extramedullary tumour was present extending out through the neural foramen (Fig. 3).

The patient was put on 15 lb (6.8 kg) of halo traction. Anterior C3/4 discectomy and fusion was performed, but after the operation the cervical spine instability and pain worsened. She was immobilised in a cervico-thoracic orthosis pending further surgery. Two weeks later she underwent resection of the C4/5 neurofibroma and C2-6 posterior cervical fusion with a Hartshill rectangle and sublaminar wires (Fig. 4). Histology confirmed a benign neurofibroma. At last review, 9 months after surgery, she had no neurological deficits and radiographs showed satisfactory positioning of the instrumentation and a solid fusion.

Case 2

A 40-year-old salesman was referred with a history of transient episodic quadraparesis, quadraesthesia and urinary retention, having fallen down a flight of stairs. Seven months earlier, he had been involved in a road traffic accident and had sustained a cervical spine whiplash injury, following which he started complaining of occasional bilateral C5 root dysaesthesia. His brother has NF 1. On examination, he walked with a myelopathic gait and had several cutaneous neurofibromata on his trunk and upper limbs.

Mild pyramidal weakness of the left upper limb was noted with MRC grade 4 motor power at his elbow, wrist and hand and mild muscle wasting of the left hand. He was generally hyper-reflexic with asymmetric extensor plantar responses.

Cervical spine radiographs were unremarkable (Fig. 5), but MRI revealed intradural extramedullary masses at more than two levels (Fig. 6). At C1/2 the spinal cord was displaced posteriorly and considerably flattened by bilobed masses with evidence of myelopathy (Fig. 7). Similarly, at C5/6 the intradural component was displacing the cord to the right with marked cord compression (Fig. 8).

Subsequent cervical laminectomy with resection of multiple neurofibromata and internal fixation and stabilisation from occiput to T2 with lateral mass plates was carried out (Figs. 9, 10). The patient made an uneventful post-operative recovery and at last clinic review, 6 months after surgery, he had regained full power in all four limbs. Histology confirmed benign neurofibromata.

Discussion

Peripheral neurofibromatosis (NF 1) is notoriously variable in its manifestations, with many patients having rela-

tively few problems. However, approximately 10–20% of individuals with this condition have serious complications such as moderate mental handicap, scoliosis, pressure problems from a neurofibroma, or malignant change. Hypertension from a rare pheochromocytoma caused by multiple endocrine neoplasia can also be a problem. Symptomatic paraspinal neurofibromata are exceptional complications of NF 1, with an incidence of 1.5% reported in one study [3]. Most occur in the thoracic spine and involve the sensory nerve root [4]; excision therefore does not result in motor disturbances and sensory loss is not too disabling due to segmental overlap.

Non-dystrophic and dystrophic spinal deformities are exceptional specifically to NF 1, as opposed to central (NF 2), segmental or mixed neurofibromatosis. Intrinsic changes may be present only in the spine and associated spinal canal anomalies secondary to an abnormal dura mater [6].

It is apparent that case 1 had sustained a potentially lethal injury to her neck from minor trauma. Plain radiographs revealed dystrophic changes in the cervical spine. A high index of suspicion enabled us to diagnose neurofibromatosis and treat her accordingly. We believe that the kyphotic deformity was present in our patient prior to trauma and that simple trauma exacerbated the underlying deformity and instability. The literature contains one report of atlanto-axial subluxation following trauma affecting the odontoid peg [2]. In a series of 17 cervical spine neurofibromatosis, Yong-Hing et al. reported a case each of atlanto-axial rotatory subluxation and anterior slip of C2 on C3 [7]. Another case of atlanto-occipital subluxation has been described by Verbiest [5]. However, neither Yong-Hing's nor Verbiest's patients were subjects of trauma, and therefore it appears that these patients have inherent instability of their cervical spine, especially if the cervical spine is dystrophic. Otherwise, reports on cervical spine neurofibromatosis with instability and subluxation after simple trauma are uncommon.

The anterior C3/4 fusion in case 1 was initially chosen because we felt that the alarming degree of instability and listhesis could be corrected and stabilised with this approach. Because of the immediate progression of the instability and increased pain following surgery, we then felt that a posterior spinal fusion with the Hartshill system was required to complement the anterior procedure.

Case 2 was different in that simple trauma did not result in roentgenographic evidence of instability. This is because non-dystrophic spines confer greater mechanical stability. Intradural cervical spine tumours act as space occupying lesions such that, in an already narrowed spinal canal, cervical spine movements would, at extremes, cause repeated cord compression and cord damage. Myelo-ischaemia had therefore occurred following the fall, which explains the resulting transient quadraparesis and quadraesthesia.

◀ **Fig. 1** Lateral cervical spine radiograph demonstrating a dystrophic spine with greater than 50% C3/4 subluxation and 40° of angular kyphosis. Posterior scalloping of the C3 and C4 vertebral bodies and increased size of the neural foramina at C2/3, C3/4 and C5/6 is also shown

Fig. 2 Sagittal section magnetic resonance image (MRI) at C4/5 showing a classic "dumb-bell" tumour at C4/5 with dural ectasia

Fig. 3 Axial section MRI at C4/5 showing a large intradural extramedullary mass extending through the exit foramen

Fig. 4 Post-operative lateral radiograph showing evidence of previous anterior surgery at C3/4 and posterior cervical fusion with a Hartshill rectangle from C2 to C6

Fig. 5 Normal looking lateral cervical spine radiograph (nondystrophic spine)

Fig. 6 Post-gadolinium sagittal MRI view demonstrating two large neurofibromata at C1/2 and C5/6. The spinal cord is tightly compressed at these two levels

Fig. 7 Axial MRI view at C1/2 showing a myelopathic spinal cord that has been displaced posteriorly by the bilobed intradural masses

Fig. 8 Axial MRI views at C5/6 showing the intradural tumour displacing the markedly compressed cervical cord to the right

Fig. 9 Post-operative lateral radiograph following occiput-to-T2 posterior fusion with lateral mass plates

Fig. 10 Post-operative antero-posterior radiograph following occiput-to-T2 posterior fusion

Conclusions

We have illustrated in our two patients that simple trauma can precipitate problems in patients with neurofibromatosis type 1. Accordingly, patients with dystrophic cervical spines are susceptible to cervical spine instability with alarming subluxation following trauma. Furthermore, inherent tumours cause spinal canal narrowing and patients are at risk of transient or permanent cord damage follow-

ing trauma. These patients must be recognised early and treated expediently to avoid neurological complications. Genetic implications mean that their offspring should be alerted as well.

We felt that MRI scanning remains invaluable in the evaluation of intraspinal contents in these patients, and a combined anterior and posterior spinal fusion is recommended for severe cervical spine deformities with instability, especially if spinal dystrophism is present.

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