PostScript

LETTERS

Psychological adjustment to locked-in syndrome

Locked-in syndrome is a severe neurological condition characterised by total or near total paralysis of motor function with preservation of vertical eye movements. Quadriplegia, lower cranial nerve paralysis and mutism are particular features of this neurological state.¹ In contrast with other neurological disorders such as akinetic mutism, coma or vegetative state, consciousness in locked-in syndrome remains intact, as do intellectual and linguistic abilities and emotional functions.^{2,3} However, communication capabilities are severely limited because the motor abilities required for self-expression are lost.

The study of psychological adjustment to this extreme condition provides the potential for insights of wider psychological and philosophical interest as well as for important clinical implications not only for patients with locked-in syndrome but also for others with extreme disabilities.

The patient described here suffers from chronic locked-in syndrome following a brainstem stroke 4 years ago. He has found an outlet for his thoughts and feelings through his poetry. These poems provide the opportunity for this study.

Case report

PK was born in Poland in 1956. He obtained his art qualifications in Poland where he worked as a satirical cartoonist. He obtained several awards for his drawings which were exhibited internationally and also won the National Award for Poetry in Poland.

On the 1 April 2001, PK suffered a brainstem stroke. After a few months, PK was able to make consistent upward movement of his eyes sufficient to establish effective communication by means of an alphabet board. PK expressed a wish to be able to dictate poetry through this medium.

The 36 poems written by PK were analysed as a source of qualitative data using Glasser and Strauss's version of grounded theory.⁴ A process of coding was carried out which involved labelling concepts from the text that were potentially relevant to the problem being studied. As coding of the sentences proceeded, eventually the concepts recurred in subsequent pieces of data (poems).

Concepts were then grouped under more abstract, higher order concepts described as "categories". A category referred to a problem, an issue or an event that was defined as being significant to the participant.

Two independent raters were involved in establishing the categories. There was 100% agreement when establishing the main themes. Minor differences were noted when referring to the subcategories.

Our identification of categories from the data has culminated in establishing 12 main themes. These are: (1) women; (2) love and beauty; (3) humour; (4) optimistic/positive thinking; (5) memories; (6) disability/discomfort; (7) unpredictability of life's events; (8)

death and finality; (9) religion; (10) rich and poor; (11) politics and history; (12) advice giving.

These categories were then grouped into three higher order themes. These were: (1) hopeful themes (women, love and beauty, humour, optimistic thinking, memories):

"You return week upon week with no
pain of which to speak
A carbon copy of carefree spring
The sunny summer feelings that you
bring
A clearing full of flowers
Lost in a forest of emerald green
towers
A burst of joy for Christmas time
A mindful eye for harder rhyme"

(2) helpless themes (disability/discomfort, unpredictability of life's events, death and finality):

"It doesn't feel great on my own...
gloomy boring sad... there's nobody
to slap on the face or nobody to get
slapped from"

(3) observant themes (religion, rich and poor, politics/history, advice giving, existential questions/reflections):

"And two and two that adds to four
Is sometimes less and often more"

The themes were also divided according to their range of applicability (eg, widespread general themes were referred to as "philosophical"). Themes that reflect on personal memories, present condition and personal enjoyment were designated as "personal" themes.

Discussion

Locked-in syndrome represents an extreme form of imprisonment. The individual is not merely locked-in a prison cell but within their own body. On reading PK's poems it becomes clear that his emotional and spiritual existence is similar to that of any able-bodied person, covering the full range of experience. There are moments of lightness, of humour, thoughts of women and his family, with bleaker moments represented too. Despite the extremity of his condition he has realistic goals for his future and believes in himself and his abilities.

It is clear PK loves life and wants to live. He makes every effort to be happy in his imprisoned existence. A former agnostic, he now acknowledges the existence of God, who gives him strength and the will to live.

For PK, being able to create and engage in his artistic endeavours is a motivating factor and a coping strategy. His 3 year creative heritage of outstanding poetry since the material incident has been acknowledged in numerous national poetry competitions.

Locked-in syndrome represents a condition where the body can no longer be "used" to act in a wilful manner. Yet in this condition cognitive processes still function and hence the person lives in their consciousness, as we all do, but is unable or less able to influence their external environment.

It is vital that all staff caring for someone with locked-in syndrome, from cleaner to consultant, are aware of the full and vital inner life that continues in the apparently inert body. While the person with locked-in syndrome can only exert choice independently in their own thoughts, they can also exert choice if others are available and prepared to act on the external environment for them. The important issue here is one of diagnosis, differentiating between those in locked-in syndrome and those where awareness and cognition are also affected. Recent studies suggest that, on average, diagnosis takes 2.5 months and in some cases it has taken 4-6 years.⁵

These clinical issues are not without cost implications. Communication is a slow, one-to-one process. Yet this is the only way in which such patients can be (partially) freed from their imprisonment.

Conclusions

The importance of providing opportunities for those with locked-in syndrome to communicate and hence articulate and elaborate their cognitive adaptation to their condition may seem obvious but can be easily overlooked, because of the precedence staff may give to physical care and because it is so difficult and requires much patience and time. However, not to enable the patient to communicate is analogous to keeping someone in solitary confinement and passing their food under the door.

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A case of posterior interosseous nerve palsy: good recovery despite diagnostic delay

It is well established that the outcome of peripheral nerve palsy due to compression by a benign mass is usually good after surgical removal, when performed early. The longest reported duration of posterior interosseous nerve (PIN) palsy prior to full recovery after surgery is 2 years.¹ We report a case of PIN lesion with a favourable post-surgical course after 4 years of progressive nerve dysfunction.

Case history

A 53-year-old man first complained of a dull deep-seated pain in his left elbow 4 years prior to examination at our facility. There was no history of direct trauma. Because of repeated elbow strain caused by sporting activities, his general practitioner suspected “tennis elbow” and prescribed analgesics, with only incomplete pain relief. Over the next years, he gradually developed weakness of the finger extensors IV and V on his left hand. This was subtle at first, but over time became more and more disabling. Wrist immobilisation with a splint was recommended. As the patient noticed an indolent subcutaneous swelling over the left proximal radius, slowly increasing in size, and his finger weakness

became almost unbearable, he was referred to our neuromuscular unit for further evaluation.

Clinical findings

There was no sensory deficit in the left upper extremity. Extensor digitorum muscle atrophy was visible, and there was complete loss of power for extension of fingers III–V (Medical Research Council for Power (MRC) grade 0), whereas relatively mild extensor weakness in fingers I and II (MRC 4) as well as ulnar wrist extension weakness (MRC 4) were present. A soft tumour was palpable in the left proximal dorsal forearm, directly below the elbow joint. (fig 1A)

Neurophysiological and diagnostic imaging findings

Nerve conduction studies of the superficial radial, median and ulnar nerves were normal. Radial motor nerve conduction studies demonstrated a reduced amplitude compound motor action potential from the M extensor digitorum communis after stimulation distal to the supinator muscle. After proximal stimulation, a complete conduction block was seen. Electromyography (EMG) of the M extensor digitorum communis showed denervation and severe loss of motor units. In the M extensor indicis, denervation was absent, but motor units were enlarged and polyphasic, with increased duration. EMG of the M brachioradialis and the muscles innervated by the median and ulnar nerves were normal. MRI of the left elbow joint revealed a T2 hyperintense tumour approximately 5×6 cm in size infiltrating the M supinator and sheathing the radius and PIN (fig 1B). The MR appearance was typical of an intramuscular lipoma.

Treatment and course of the disease

The tumour was surgically removed. A curved incision was placed over the supinator muscle

in the proximal third of the forearm; fascia and muscle were divided along a 4 cm course. Under the muscle, there was a lobulated lipoma, which was highly vascularised and well encapsulated. The nerve was found to run through the lipoma and was pressed against the distal border of the supinator muscle (fig 1C) causing a massive depression (fig 1D). This depression was 2 mm wide, involving the entire PIN. At this level, the epineurium was incised under the microscope to relieve pressure and to facilitate regeneration. The tumour was composed of mature lipocytes without malignancy, confirming the diagnosis of an intramuscular lipoma.

The postoperative course was uneventful. The patient regained partial motor function within 2 months. At the 3 year follow-up, only minor weakness (MRC 5–) of the Mm extensor carpi ulnaris and extensor digiti minimi was documented.

Discussion

PIN lesions due to nerve entrapment at the elbow result in weakness of wrist and finger extension without sensory loss. Elbow pain caused by the factors responsible for the PIN lesion is one of the most frequent presenting symptoms. PIN palsies are commonly idiopathic but may also be caused by tumours of the forearm such as gangliomas, neurofibromas, haemangiomas and, most commonly, lipomas. Lipomas are mostly parosteal or intermuscular (ie, originating from lipocytes of the radial periosteum or muscle fascias) and well delineated. In contrast, PIN palsies due to intramuscular lipomas (ie, infiltrating the muscles of the forearm adjacent to the PIN), as seen in the present patient, have rarely been described.

It is not unusual for patients with PIN palsies to experience a long course of disease prior to the correct diagnosis and treatment, despite typical neurological signs. The remarkably long diagnostic delay of 4 years in the present case is at the extreme end of the spectrum described in the literature. Outcome after surgical removal of parosteal or intermuscular lipomas is usually good when performed early,^{2–5} with 2 years reported as the longest disease duration prior to full recovery.¹ In our patient, an equally favourable post-surgical prognosis seemed unlikely taking into account the axonal damage found on EMG and the duration of disease. Furthermore, lipomas of the muscular infiltrating type are rare and information on success of surgery is sparse. In the present case, PIN function improved rapidly at first and then slowly over several months after decompression and epineurotomy. This indicates that the initial improvement resulted from resolution of the conduction block while the later gain of function was attributable to reinnervation.

This case demonstrates that surgical removal of a tumour and epineurotomy of the nerve is a promising option for PIN palsy, even after longstanding nerve dysfunction in cases with conduction block and only incomplete axonal damage.

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Figure 1 (A) Preoperative motor function in the patient with posterior interosseous nerve (PIN) palsy. The green marking outlines the indolent subcutaneous swelling in the left proximal dorsal forearm. (B) Preoperative MRI showing an intramuscular T2 hyperintense tumour approximately 5×6 cm in size infiltrating the M supinator. (C) The nerve (X) was found to run through the lipoma and was compressed by a fascial band (arrows). (D) Site of nerve compression (2 mm wide) after tumour removal. At this level, the epineurium was incised under the microscope.