The first description of idiopathic progressive bulbar palsy

Furukawa has recently recounted the lesser known contributions to clinical neurology of Sir Charles Bell who first described the numb chin syndrome and the phantom phenomenon in his book on the nervous system. We would like to report an account of a patient with progressive bulbar palsy (PBP) who was referred to Bell, is included in the same book and predates the writings of Duchenne de Boulogne, who is usually credited with the first descriptions.1

The patient's details are contained in a letter to Bell from Robert RW Robinson, of Preston, England, dated 21 July 1825. He asked for advice about "an unmarried lady, nearly seventy years of age, who has enjoyed uninterrupted good health up to the present illness". He states:

From the first of her complaint to the present moment she has been free from headach (sic) and from pain, numbness, or debility of the limbs. The vision and hearing are natural; the appetite good; the bowels regular, and the sleep natural. . . . Some few months ago she had some difficulty in using the tongue and in expressing particular words. This difficulty has gradually increased, and now she cannot pronude the tongue, or even move it. She has lost her speech altogether. The tongue itself is soft and pulpy; but it retains its sense of taste and feeling. The deglutition is impaired and occasionally she is distressed with a sense of suffocation, in attempting to swallow food, which she is now obliged to do with great care. She cannot hack up any thing from the throat, nor draw any thing from the posterior nares by a back draught. The features of the face are quite natural, and the skin retains its feeling. The saliva occasionally flows from the mouth."

Bell noticed the similarity between this patient's condition and the syndrome observed in a dog after section of the lower cranial nerves and concluded that the patient was suffering from "a paralytic affection of the ninth nerve" and noted that the "function of the fifth nerve was enervated." Unfortunately no follow up or necropsy results are available to tell us if the disease became more widespread, neither is there any information about the deep tendon reflexes, as this physical sign was not introduced into the neurological examination until 1875. Bell advised nauseating medicines and leeches under the mastoid, amongst other remedies.

"This description would rival any modern case report for conveying clearly the clinical signs, the important negative features for the diagnosis and the disability resulting from this tragic affliction. We would argue that this patient's illness, which combines anarthria, impaired deglutition and tongue movement with palsy, probably with mixed upper and lower motor neuron signs, is due to idiopathic PBP, one of the modes of presentation of motor neuron disease and which may occasionally remain confined to the bulbar region. Given the advanced disease, a brainstem tumour seems unlikely in the absence of sensory signs and if the bulbar palsy was due to myasthenia gravis, ocular and limb symptoms would be expected. Bell identified the condition but little is known of the referring physician. Robinson was born in Lancashire and graduated Doctor of Medicine at Edinburgh on 12 September 1800. He was president of the Royal Medical Society, founded in 1734 by a group of medical students after the successful acquisition of a fresh body for dissection. The founders initially met in taverns " . . . with the intention that a dissertation in English or Latin on some medical subject . . . should be composed and read" and later meetings were attended by Charles Darwin. There is no evidence that Robinson had an earlier interest in neurology as he obtained a doctorate based on a urological subject (Disputato medica inauguralis de vesica, urethraeque morbis). He became a Licentiate of the College of Physicians (London) in 1807 and returned to his native Lancashire where he was an influential member of the board of management of the Preston Dispensary, founded in 1809. Unfortunately 169 years after his original account, the etiology of PBP remains obscure.

ANDREW M CHANCELLOR
Department of Neurology
University Hospital of Wales,
Cardiff CF4 4XW, UK

JOHN D MITCHELL
Department of Neurology,
Royal Preston Hospital,
Share Lane, Preston, UK

ROBERT J SWINGLER
Department of Neurology,
Dundee Royal Infirmary,
Dundee, UK

Correspondence to: Dr Chancellor

3 Bell C. The nervous system of the human body. As explained in a series of papers read before the Royal Society of London. With an appendix of cases and observations on nervous diseases. London: Longman, 1836. 390-1.
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A M Chancellor, J D Mitchell and R J Swingler

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