Paul Broca's contribution to the study of peroneal muscular atrophy

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ABSTRACT

Objetive. Different biographers have underlined Broca's early contributions (1847–1852) to the study of Duchenne muscular dystrophy (1861). Our purpose is to pinpoint the origin of this claim using original sources.

Methods. We reviewed the history of muscular dystrophy according to different authors, biographies of Duchenne and Broca, and issues of the *Bulletins de la Société Anatomique de Paris* published in the specified time period and mentioning Broca's studies of muscular atrophy, foot deformities, or lower limb weakness.

Results. Between 1847 and 1852, Paul Broca described four autopsied cases of clubfoot associated with severe atrophy and fatty degeneration of the muscles of the lower extremities. Two of these individuals had experienced progressive neuromuscular disease characterised by bilateral pes cavus, marked atrophy of the peroneus muscles, and moderate impairment of the thigh muscles; there was also slight atrophy of hand muscles in one case. Broca identified the underlying cause of the foot deformity as the muscular imbalance between agonists and antagonists created by unequal degrees of muscular atrophy.

Conclusion. Broca's observations are more suggestive of peroneal muscular atrophy than of Duchenne muscular dystrophy. These early contributions to the study of this neuromuscular syndrome appeared some 40 years before Charcot, Marie, and Tooth published their classic descriptions in 1886.

KEYWORDS

Paul Broca, peroneal muscular atrophy, pes cavus, Duchenne muscular dystrophy

Introduction

Eponyms used in medicine often do not fairly identify the first researcher to describe a phenomenon, or the individual otherwise deserving of that honour.¹ Duchenne muscular dystrophy (named for Guillaume Benjamin Amand Duchenne, 1806-1875) has one of the most frequently criticised eponymous names in neurology. Different authors have suggested adding, at the very least, the names of Griesinger,^{2,3} Meryon,⁴⁻⁶ and the no less deserving Conte of Italy.⁷

Paul Broca's description of what may have been Duchenne muscular dystrophy has been overlooked by some historians of that disease⁸ and even by Broca's own

Corresponding author: Dr Santiago Giménez-Roldán E-mail: sgimenezroldan@gmail.com biographers.^{9,10} Nevertheless, it has been mentioned in books on the history of neurology and studies of early descriptions of that disorder.³ We find an important passage in the biography by Francis Schiller,¹³ which is probably the most detailed account of Broca's life available. According to that author, Paul Broca documented an anatomical study of a case of clubfoot in 1851; in doing so, he described a case of Duchenne muscular dystrophy. If this is true, Broca's contribution would have been published ten years before Duchenne de Boulogne's *Paralysie hypertrophique de l'enfance de cause cérébrale* in 1861.¹⁴ It would also have preceded Meryon's studies in 1851 and 1852,¹⁵ Little's report in 1853,¹⁶ and Griesinger's article in 1865.¹⁷ Monod-Broca, doctor and descendent of the famous French anatomist, surgeon, and

Received: 11 August 2013 / Accepted: 15 December 2013 © 2014 Sociedad Española de Neurología anthropologist, claimed that his great-grandfather had penned an early description of what he called *maladie de Duchenne*. Broca had interpreted the syndrome as a monomyelic form of myopathy.¹⁸

In light of these conflicting accounts, our purpose is to investigate Paul Broca's possible contributions to the study of Duchenne muscular dystrophy by analysing primary sources. Surprisingly, Broca's biographers^{13,18} based their conclusions on different cases; furthermore, two of Broca's cases had developed what was probably a long-term neuromuscular process suggestive of peroneal muscular atrophy, and not a myopathic disease.

Material and methods

We located references to Broca's potential contributions to the study of muscular dystrophy in books focusing on that disease,¹⁹⁻²¹ articles on the history of child neurology,²² and biographies of both Duchenne de Boulogne²³⁻²⁵ and Paul Broca.^{9,10,26} We consulted Duchenne's original studies, copies of which are kept by the Spanish Royal Academy of Medicine in its library in Madrid. The biography by Guilly,²³ which is not available in Spain, was provided by the University of Bern thanks to an interlibrary loan coordinated by the Spanish National Library. The biography written by Monod-Broca was obtained through the same channel.²⁶ Using the digital library Gallica, we were able to access the Bulletins de la Société Anatomique de Paris and consult Broca's original studies mentioning muscular atrophy, limb paralysis, or deformities of the feet between 1847 and 1852. The reason for examining that precise time period is that it was mentioned by both Schiller¹³ and Monod-Broca¹⁸ as corresponding to the dates of Broca's descriptions of muscular dystrophy.

Results

Between 1847 and 1852, Paul Broca presented anatomical findings from four cases of *pied-bot* (clubfoot) to the Société Anatomique de Paris. These abstracts were later published in that society's *Bulletins*.²⁶⁻³⁰ We should point out that the cases of muscular dystrophy according to Philippe Monod-Broca¹⁸ and Francis Schiller¹³ correspond in fact to different patients, whose respective studies were presented in 1847 and 1851.

Broca described anatomical findings from four individuals; the first (case 1) was a 20-year old male who had exhibited clubfoot since the age of 2 years. The study examined a leg amputated due to suppuration of the talus area.^{18,27} Findings included shortened foot with unilateral pes valgus. As in the other cases, this study offered purely anatomical observations and no clinical data other than age of the subject. Since we believe that this case displays a type of monomelic amyotrophy, possibly congenital in origin, we excluded the hypothesis of an early form of peroneal muscular atrophy.

Broca's second report on a case of clubfoot (case 2)^{28(p265)} seems to have gone unnoticed by his biographers even though it provided some relevant details. This autopsy was performed on a 45-year old subject provided by the Faculty of Medicine; as in earlier cases, no clinical data were given. Broca highlights the visible calluses on the sole of the foot marking the points of contact as well as the "extremely arched" shape of both feet, which were very short along their anterior-posterior axis. An excerpt from his detailed description is included here. "Toes exhibit exaggerated extension; the great toe is almost completely luxated and the tendon of its extensor muscle forms a marked prominence, raising the skin like a cord".²⁸ Fatty infiltration of the leg muscles is "particularly severe in the lateral peroneus" and also, to a lesser extent, "in the thenar and hypothenar muscles" (Figure 1).

Broca was surprised to find that "the other muscles of the thigh are affected, but not all show the same degree of involvement. The sartorius and the gracilis muscles are wasted and discoloured".²⁸ After carefully describing the state of the joints, he concluded that clubfoot in this patient was not a congenital condition; instead, it was

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M. Cruveilhier pense aussi qu'il s'agit d'un épanchement de sang, coexistant avec une maladie des ganglions lymphatiques. Cette maladie, dit M. Deville, n'est autre chose qu'une hypertrophie glandulaire.

14. — M. Broca présente deux pieds bots trouvés dans les pavillons de la Faculté sur un homme d'environ quarante-cinq ans, bien constitué du reste, et fortement musclé. Ces deux pieds bots appartiennent à l'espèce varus; mais ils ne sont pas également avancés dans leur formation.

Figure 1. Excerpt from the description of case 2, *Bulletins de la Société Anatomique de Paris*, 1849.^{28(p265)}

acquired due to muscular imbalance between agonists and antagonists, and especially because of atrophy of the lateral peroneus. While Broca does not describe the state of the spinal cord, he does mention that nerve volume was normal. In any case, the young anatomist and surgeon pointed out the absence of the fibrous tracts that deform the joints in cases of congenital clubfoot.²⁸

In 1851, Broca reflected on the pathogenesis of clubfoot in the context of a new case description (case 3),³⁰ the one summarised in the biography written by Schiller.¹³ Broca presented his findings from a study of the lower limbs of a 30-year old female whose cadaver had been provided by the École pratique. According to the summary, the deformity of the feet was characterised by marked elevation of the internal surface, abundant plantar calluses at the points of contact, traction of the extensor hallucis and the extensor digitorum, and unequal degrees of fatty transformation in the different muscles of the lower limbs. Changes were particularly noticeable in the lateral and anterior peroneus muscles, the flexor hallucis brevis, and the flexor digitorum. Atrophy and fatty transformation extended irregularly throughout the different lower limb muscle groups, appearing in any case less pronounced in the thigh muscles. According to the anatomist, the nerve thickness appeared to be normal.

In his last brief note, Broca mentioned the autopsy performed on an elderly woman named Demarques (case 4)³¹; here, foot deformity was unilateral, as in case 1 in 1847. We therefore regard case 4 as a case of monomelic amyotrophy of unknown origin.

Discussion

Between 1847 and 1852, Paul Broca presented to the Société Anatomique de Paris four autopsy cases that revealed severe deformity of one or both feet (*pied-bot* or clubfoot).²⁷⁻³¹ Broca, a very young man at the time, was working as an intern in Dr Blondin's operating theatres in prestigious Hôtel Dieu de Paris. Here, he assisted the doctor and the doctor's assistant Gerdy with daily autopsies, and he was probably most attentive to the findings they uncovered (Figure 2).

His four case studies were published one by one in the *Bulletins de la Société Anatomique de Paris*. The 1847 and 1848 articles were titled "Investigation of the pathological anatomy of clubfoot"; to his next two articles (1850 and



Figure 2. Paul Broca at the age of 18 years, soon to be admitted to Dr Blondin's operating theatres at Hospital Hôtel Dieu de Paris (from Schiller¹³ © University of California Press).

1851), he added the clarification "produced by fatty changes in the muscles". These were the first articles of Broca's career, and his output was impressive: more than 500 published studies in what was not a particularly long lifetime (1824-1880).⁹ Nevertheless, his keen deductions about the mechanism causing acquired clubfoot, which he clearly distinguished from congenital clubfoot, failed to attract the attention of his superiors –the "respectable eminences of fifty", as his great-grandson ironically described them.²⁶

We might ask ourselves why Broca might have become so fascinated with clubfoot during such a long period of

time. In the first half of the 19th century, gait disorders combined with limb deformities were commonly regarded as primarily osteoarticular processes; doctors tended not to consider neurological origins. Orthopaedic surgeon William John Little (1810-1894) was famous for his description of the spastic diplegia form of cerebral palsy and also for his book On the Treatment of the Deformities of the Human Frame. A footnote in that volume briefly mentions two brothers with severe muscle retractions. At a later date, Meryon published more extensive early observations of childhood-onset muscular dystrophy.³² Broca's reports were noteworthy because he emphasised that foot deformity was accompanied by severe atrophy in the leg muscles, also describing how the degree of atrophy varied between different impaired muscles. He also insisted that foot deformity was the result of agonists and antagonists being unevenly matched during locomotion.

We cannot clearly identify the moment when Broca's observations were erroneously interpreted descriptions of a form of myopathy or even as early reports of Duchenne muscular dystrophy. A substantial part of the error can probably be traced to comments made by Schiller, his biographer, with regard to case 4: "The young Broca was the first to observe, understand, and explain the essential pathology of muscular dystrophy".¹³ In turn, Monod-Broca also claims that his great-grandfather described muscular dystrophy based on the unilateral pes valgus in case 1: "...clubfeet are merely an anecdotal manifestation in myopathies..."¹⁸ His biographers' error may stem from confusing Duchenne muscular dystrophy with progressive muscular atrophy (Duchenne-Aran disease, described as a degenerative process of the anterior horn of the spinal cord in 1850 by François Amilcar Aran [1817-1861]). Incidentally, this is another historical example of misattributed credit.33

Although no clinical data is given for the four autopsied cases of clubfoot, Broca's exacting anatomical description invites us to speculate on the neuromuscular processes that affected cases 2 and 3, published in 1849 and 1851 respectively.^{28,30} Both displayed severe forms of bilateral pes cavus. Broca stressed that the process was acquired rather than congenital and described the distinctive distribution of atrophy and fatty degeneration of leg muscles. This degeneration was particularly marked in plantar and peroneal muscles, but it also extended to the thigh muscles where it was less severe. In case 3, mild

atrophy also affected the hands. Broca described the spinal cord in case 2 as normal, presumably basing his assessment only on its macroscopic appearance (this contrasted with findings in Aran's cases of progressive muscular atrophy). He also described the thickness of peripheral nerves as normal.

Although interpreting mere anatomical data published a century and a half ago requires a prudent approach, it seems clear that Paul Broca's detailed descriptions do not correspond to a myopathic process. Instead, it would be tempting to suggest that two of his cases could be interpreted as early descriptions of peroneal muscular atrophy. If this is true, his account is nearly 40 years older than that of Jean Martin Charcot (1825-1893) and Pierre Marie (1853-1940), whose 1886 study was published simultaneously with that of Howard Henry Tooth (1856-1925).^{34,35} It even predates the observations by Virchow (1855) and Schultze (1884), who have been mentioned by historians documenting the disorder.

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