

# Zerebelläre Symptome bei Motoneuronerkrankungen

## Spezielle Form des ALS-Plus-Syndroms

### Zusammenfassung

Die amyotrophe Lateralsklerose (ALS) ist eine neurodegenerative Erkrankung, welche durch den progressiven und selektiven Untergang von Motoneuronen des zerebralen Kortex, des Hirnstamms und des Rückenmarks gekennzeichnet ist. Eine Mitbeteiligung des kortikospinalen Systems zeigt sich in Symptomen einer spastischen Lähmung. Schlaffe Paresen mit Muskeltrophien, Muskelkrämpfe sowie Faszikulationen sind kennzeichnend für die Degeneration des 2. Motoneurons. Formale Kriterien für das Krankheitsbild der ALS wurden mit den El-Escorial-Kriterien festgelegt. Im Anhang zu diesen Kriterien werden Syndrome mit darüber hinausgehenden Symptomen wie extrapyramidal-motorischen Störungen oder Demenz aufgeführt. In der vorliegenden Arbeit werden 2 Fälle einer typischen ALS mit klinischen Zeichen einer zerebellären Schädigung vorgestellt. Die dargestellten Fälle mit Zeichen einer Schädigung des 1. und 2. Motoneurons in Kombination mit zerebellärer Ataxie lassen sich anhand der revidierten El-Escorial-Kriterien in den Symptomenkomplex von ALS-Plus-Syndromen einordnen.

### Schlüsselwörter

Amyotrophe Lateralsklerose · El-Escorial-Kriterien · ALS-Plus-Syndrom · Zerebelläre Degeneration

Die amyotrophe Lateralsklerose (ALS) ist eine neurodegenerative Erkrankung, die zum Absterben sowohl des 1. als auch des 2. Motoneurons führt. Die Degeneration der  $\alpha$ -Motoneurone bedingt die peripheren oder nukleären Zeichen mit schlaffen Paresen, Muskeltrophien und Faszikulationen. Durch die Degeneration des kortikospinalen Systems treten zentrale spastische Paresen mit enthemmten Muskelreflexen und pathologischen Reflexzeichen auf. Die Erkrankung beginnt in der Regel fokal und asymmetrisch [1], am häufigsten an einer oberen Extremität und mit distaler Betonung [1]. Primär bulbäre Symptome liegen bei Diagnosestellung in ca. 25% der Fälle vor.

International angewandte Diagnosekriterien für das Krankheitsbild der ALS wurden mit den sog. El-Escorial-Kriterien 1994 erstellt [2] und im Jahr 2000 überarbeitet [3]. In Appendix 1 der aktuellen Diagnoserichtlinien werden als verwandte Krankheitsbilder die sog. ALS-Plus-Syndrome definiert. Am bekanntesten ist der ALS-Parkinson-Demenz-Komplex. Dieser wurde erstmals um 1950 in 3 ethnisch und geographisch isolierten Populationen beschrieben [4, 5]. Daneben finden sich Symptomenkomplexe mit extrapyramidal-motorischen, kognitiven oder autonomen Defiziten [4, 6]. Es wurden bisher nur wenige Fälle beschrieben, bei denen eine zerebelläre Ataxie und eine Motoneuronerkrankung zusammen auftraten [7, 8, 9, 10, 11]. Ein Symptomenkomplex mit Motoneurondegeneration und Zeichen einer zerebellären Degeneration wurde

unter anderem auch bei Hexosaminidase-A-Mangel oder im Rahmen der Creutzfeld-Jakob-Erkrankung bei den spinocerebellären Ataxien beschrieben [1]. Weiter muss die olivo-ponto-zerebelläre Atrophie (OPCA) als Unterform der multiplen Systematrophie (MSA) in differenzialdiagnostische Überlegungen einbezogen werden. Außerdem können spastisch-ataktische Symptome auch im Rahmen einer Hashimoto-Thyreoiditis auftreten [12].

In der vorliegenden Arbeit stellen wir 2 Patienten mit ALS und darüber hinausgehend ataktischen Störungen vor.

### Fallbeschreibungen

#### Fall 1

Wir berichten über eine 70-jährige Patientin mit amyotropher Lateralsklerose (ALS) und zeitlich verzögert einsetzender zerebellärer Ataxie. Die Krankheit begann mit einer progredienten Dysarthrie und Dysphagie, d. h. überwiegend bulbären Symptomen. In den folgenden Monaten traten dann Paresen im Bereich der Extremitäten mit linksseitiger Betonung auf. Zu diesem Zeitpunkt bestanden auch Muskelkrämpfe und Faszikulationen an allen Extremitäten. Zwei

# Als A Plus Ceritification

**Sabine Zange**

A red circular graphic with a gradient, appearing as a stylized arrow or a partial circle, located to the right of the name bar.

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