Childhood Seeligmüller Strümpell Philip Disease: The First Case in Iraq and a Review of the Early Historic Documentation of the Disease in the Literature

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Abstract

Seeligmüller Strümpell Philip disease is characterized by familial progressive spastic paraplegia or paraparesis associated with a progressive deterioration of gait. The disorder is genetically heterogeneous and can be inherited in an autosomal dominant, autosomal recessive or X-linked recessive manner. The mode of inheritance involved has a direct impact on the chances of inheriting the disorder. The cornerstone of treatment is the use of various muscle relaxants. The disorder has not been reported or documented in Iraq. This paper aims to report the first case of childhood Seeligmüller Strümpell Philip disease in an Iraqi girl.

Patients and Methods: A thirteen-year girl with childhood Seeligmüller Strümpell Philip disease who developed systemic lupus erythematosus is described and the early historic documentation of Seeligmüller Strümpell Philip disease in the medical literature is briefly updated.

Results: The diagnosis of Seeligmüller Strümpell Philip disease in this girl was based on clinical findings and supported by electromyography and nerve conduction studies. Brain MRI showed normal findings. Few weeks before the patient was seen, they took her outside Iraq to Medipol Mega complex of Hospitals for treatment. They performed tenotomy and tenoplasty of the hip adductors, achilloplasty, and posterior capsule relaxation. When the girl was first seen at the Children Teaching Hospital of Baghdad Medical City, the parents regretted taking her outside Iraq for treatment as she didn’t show any improvement, and was still using the wheelchair.

Conclusion: Unfortunately, the girl was seen too late, and despite consulting many doctors in Iraq and outside Iraq, she didn’t receive appropriate evidence based treatment and was treated unsatisfactorily surgically.

Keywords

Seeligmüller-Strümpell-Philip Disease, Historic Update, Iraq, Useless Surgeries
Introduction
Seeligmüller Strümpell Philip disease is characterized by familial progressive spastic paraplegia or paraparesis associated with a progressive deterioration of gait resulting from dysfunction of the upper motor neurons in the spinal cord; therefore it is not a form of cerebral palsy which results from brain damage. The condition may have early onset beginning in early childhood at about the age of two years or later onset in adulthood at about the age of 40 years. The disorder is genetically heterogeneous and can be inherited in an autosomal dominant, autosomal recessive or X-linked recessive manner. The mode of inheritance involved has a direct impact on the chances of inheriting the disorder. The cornerstone of treatment is the use of various muscle relaxants [1-13].

The disorder has not been reported or documented in Iraq [14-17]. This paper aims to report the first case of childhood Seeligmüller Strümpell Philip disease in an Iraqi girl.

Patients and methods
A thirteen-year girl with childhood Seeligmüller Strümpell Philip disease who developed systemic lupus erythematosus is described and the early historic documentation of Seeligmüller Strümpell Philip disease in the medical literature is briefly updated.

Results
A girl with progressive spasticity in the lower limbs caused by pyramidal tract dysfunction as indicated by increased muscle tone and brisk reflexes experienced progressive gait since early childhood. The parents were consanguineous. During early childhood, she was able to walk on her toes with progressive difficulty, but she had to use a wheelchair before the age of ten. She didn’t have speech abnormality or difficulty swallowing. The father confirmed that they consulted many physicians during the previous year; no one prescribed muscle relaxant to reduce spasticity. Left Achillotomy was performed at about the age of five years without experiencing any benefit.

During the previous months, she received a diagnosis of systemic lupus erythematosus after experiencing arthritis and urinary abnormalities. The ANA and Double-stranded DNA tests were both positive and renal biopsy showed mesangial lupus nephritis. The patient was treated with immune suppressive treatments.

Few weeks before the patient was seen, they took her to the Medipol Mega complex of Hospitals for treatment. She was initially admitted to the pediatric neurology department, but she was referred to the department of orthopedics and traumatology. They performed tenotomy and tenoplasty of the hip adductors, achilloplasty, and posterior capsule relaxation.

When the girl was first seen at the Children Teaching Hospital of Baghdad Medical City, the parents regretted taking her to Istanbul for treatment as she didn't show any improvement, and was still using the wheelchair (Fig-1).

![Fig-1: When the girl was first seen, she was still using the wheelchair](image)

The diagnosis of Seeligmüller Strümpell Philip disease in this girl was based on clinical findings and supported by electromyography and nerve conduction studies. Brain MRI showed normal findings.
Case Report

Discussion
In 1876, Adolph Seeligmüller (Fig-2), a German neurologist described a family of four affected children with spasticity. The patients of Seeligmüller had generalized wasting that was probably resulted from disuse atrophy [1]. In 1883, Adolph Strümpell (Fig-3), a German neurologist also reported the condition [2,3]. In 1886, Philip described the first case in the United Kingdom [4].

The cornerstone of treatment of Seeligmüller Strümpell Philip disease is the use of various muscle relaxants [11-13].

Conclusion
Unfortunately, the girl was seen too late, and despite consulting many doctors in Iraq and outside Iraq she didn’t receive appropriate evidence based treatment and was treated unsatisfactorily surgically.

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Conflict of interests
All authors have read and approved the final version of the manuscript. The authors have no conflicts of interest to declare.

References
Case Report


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