ABSTRACT

Tranebjaerg Svejgaard syndrome (TSS) is a X-linked intellectual disability syndrome mainly characterized by seizures and psoriasis and indicative to a specific sign, symptoms associated with a health challenge. The relationship between the inheritance to be X-linked recessive and severe mental retardation. Mental retardation (MR) is clearly caused by an inborn error of metabolism, such as in the Hunter syndrome or in Lesch-Nyhan and appears to be accessory with respect to the prevailing disorder which is in Duchenne muscular dystrophy. Most common symptoms are Generalized seizures, status epilepticus, stooping posture, ataxic gait, and contractures of fingers, hypotonic diplegia, intelligible speech, psoriasis vulgaris with pustules, Asphyxia. Physical abnormalities like Hypotonia, hyperextensible joints, large tongue and large, everted ears. Ataxic gait, thoracic dextroconvex scoliosis, convergent strabismus, big tongue, open mouth, long face, long slender fingers, curved 5th fingers, bilaterally proximal placed thumbs and bilateral simian crease. MR is mostly undiagnosed in Children and Adults which are ADHD, Alzheimer Disease, Bipolar Disorder, Schizophrenia, Epilepsy. TSS can be treated with symptomatically in case of psoriasis the possible First-line treatment will be and soap substitute for washing opical steroids are used for anti-inflammatory effect. Systemic treatments for adult include oral Prednisolone, Methotrexate, Ciclosporin and Azathioprine.

KEYWORDS: Tranebjaerg Svejgaard syndrome, Mental retardation, X-linked recessive syndrome.
INTRODUCTION
Tranebjaerg Svejgaard syndrome (TSS) is a X-linked intellectual disability syndrome mainly characterized by seizures and psoriasis and indicative to a specific sign, symptoms associated with a health challenge.\[1\] There is a relationship between the inheritance to be X-linked recessive and severe mental retardation, associated with psoriasis apparently inherited in a X-linked recessive syndrome.\[3\] Psoriasis has never been reported as a monogenic disorder.\[6\] Intellectual disability (ID), is also called as mental retardation, is characterized by below-average intelligence or mental ability and a lack of skills which are necessary for day-to-day living.\[4\]

CLINICAL SYMPTOMS
TSS is a common genetically determined X-linked intellectual disability syndrome mainly characterized by Mental retardation, seizures and psoriasis. Mental retardation (MR) is clearly caused by an inborn error of metabolism, such as in the Hunter syndrome or in Lesch-Nyhan and appears to be accessory with respect to the prevailing disorder which is in Duchenne muscular dystrophy.\[5\] MR seems to be the primary element according to our computation, 20 phenotypes such as lethal in the hemizygote, Aicardi and in the Rett syndrome which is caused by a dominant gene. Chronic, inflammatory skin disease which is characterized by rounded erythematous, dry, scaling patches and the Skin lesions seen on nails, scalp, genitalia, extensor surfaces and the lumbosacral region. Epidermopoiesis is considered to be the fundamental pathologic feature in psoriasis.\[6\]

Most common symptoms are Generalized seizures, status epilepticus, stooping posture, ataxic gait and contractures of fingers, hypotonic diplegia, intelligible speech, psoriasis vulgaris with pustules, Asphyxia.\[5\]

The facial symptoms included apparent hypertelorism, large ears, large and wick open mouth with a long upper lip, Highly arched palate, broad nasal bridge, anteverted nares, praninent lips, macrostomia and cryptorchidism.

Physical abnormalities included: Hypotonia, hyperextensible joints, large tongue and large, everted ears. Bone age was retarded. Ataxic gait, thoracic dextroconvex scoliosis, convergent strabismus, big tongue, apen mouth, long face, long slender fingers, curved 5th fingers, bilaterally proximal placed thumbs and bilateral simian crease.\[4,7\]
Ophthalmological examination includes hypertelorism, alternating esotropia and mild hypermetropia (+4.0).

Genital conditions are presented as penis was big and the testes were 25-30 ml each.

**INVESTIGATION**

A variety of tests may be done to confirm the diagnosis of MR. These include blood tests, urine tests, imaging tests to look for structural problems in the brain, or electroencephalogram (EEG) to look for evidence of seizures. Psoriasis will be confirmed by biopsy tests. Other examinations include Lysosomal enzymes in leukocytes, plasma amino acids, Metabolic screening in 111-7, urinary excretion levels, organic and uric acids will demonstrate aminoaciduria with excretion of several amino acids.

x9 Typing where the following relatives were Xg typed: II-1, II-2, II-3, I-4, III-1, III-2, III-3, and 111-5. Everything were Xg(a+) and thus, non-informative. Steroid Sulfatase Activity Assay is a method of Lykkesfeldt, where living prapositi, 111-3 and 111-5 from cultured skin fibroblasts. A standard lymphocytotoxicity tests is HLA-typed and only 111-5 has psoriasis-associated HLA antigens B13, II-2, II-3 Cw6 in a haplotype inherited. Cytogenetically analysis included a search for fra(X) and high resolution RBA banded according to standard techniques which is prometaphase analysis and Metaphase analysis.

**DIAGNOSIS**

MR is mostly undiagnosed in Children and Adults which are ADHD, Alzheimer Disease, Bipolar Disorder, Schizophrenia, Epilepsy. The diagnosis was based on a gross examination, showing guttate psoriatic plaques in the scalp and on the trunk. The palms of the hands and the soles of the feet were dry, coarse, thick and hyperkeratotic. Scaling occurred predominantly along the flexion creases of the hands; at the elbows typical psoriatic lesions were observed.

**TREATMENT**

TSS can be treated with symptomatically in case of psoriasis the possible First-line treatment treatment will be and soap substitute for washing opical steroids are used for anti-inflammatory effect. Systemic treatments for adult include oral Prednisolone, Methotrexate, Ciclosporin and Azathioprine. Antihistamines, Topical imidazoles, Coal tar preparations, Phototherapy consisting of Psoralen and long wave Ultra violet) with moderate effect.
In MR Early intervention may include speech therapy, physical therapy, occupational therapy, family counseling, training with special assistive devices, or nutrition services.

CONCLUSION
Considering the fact that relation between X-linked recessive trait, psoriasis, and considering the present family history with X-linked psoriasis invariably associated with severe mental retardation suggests segregation of a previously undescribed X-linked recessive MR/psoriasis syndrome.

REFERENCE