Progressive bulbar palsy with deafness (A case report)

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Progressive bulbar palsy (Brown-Vielatto-Van loere syndrome) is a rare autosomal recessive disorder that is presented with deafness usually in the first decade of life (range 1.5 - 31 year) with subsequent development of facial weakness, dysphagia and dysarthria. This disorder is slowly progressive; upper and lower limbs may become atrophic and weak over many years.

In this report a 11 years old girl from East Azarbaijan-Iran are described, that developed with deafness, gait disturbance and dysphagia. On exam she had weakness and atrophy mainly in upper limbs, with tongue fasciculation and positive romberg test. Brain MRI was normal. Electrodiagnostisit study showed normal SNAPs with low ampiitude CMAPs and sever neurogenic pattern in needle EMG.

Key Words: Progressive bulbar palsy, Deafness, weakness.