

## WOLFRAM SYNDROME – A CASE REPORT

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Wolfram syndrome is characterized by the coexistence of diabetes mellitus and optic atrophy, but may also associate with diabetes insipidus, deafness, urinary tract dilatation, ataxia and psychiatric disorders. We report a 19 year-old female patient presenting with juvenile-onset diabetes mellitus, bilateral optic atrophy, high frequency hearing impairment, and gaze evoked nystagmus, but without diabetes insipidus, urinary tract abnormality or other obvious signs of neurodegeneration. Identification of Wolfram syndrome among juvenile-onset diabetics is important because of the distinctly different sources of morbidity and causes of mortality from those of classic insulin-dependent diabetics.

Key words: Wolfram syndrome, optic atrophy, diabetes mellitus, hearing loss.

### INTRODUCTION

Wolfram syndrome, first described by Wolfram in 1938,<sup>1</sup> is a rare hereditary disorder and is transmitted as an autosomal recessive single gene syndrome.<sup>2</sup> Although bilateral optic atrophy and diabetes mellitus (DM) are the essential features of Wolfram syndrome, the acronym DIDMOAD (Diabetes Insipidus, Diabetes Mellitus, Optic Atrophy and Deafness) indicates the high possibility of coexistent diabetes insipidus and hearing loss. Besides, there is additional co-morbidity, including urinary tract anomaly, psychiatric illness, and neurodegeneration involving central nervous system, peripheral nerves and neuroen-

doctrine tissues.<sup>2</sup> Here, we report a case of Wolfram syndrome presenting with juvenile-onset diabetes, optic atrophy, hearing impairment, and gaze evoked nystagmus, but without diabetes insipidus (DI), urinary tract abnormality or other obvious signs of neurodegeneration.

### CASE REPORT

A 19-year-old girl complained of progressive blurred vision of both eyes for 3 years and difficulty in color discrimination for six months. She had juvenile-onset diabetes since age of 7 but with poor compliance in insulin therapy and experienced several episodes of diabetic ketoacidosis. Mild cataract

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