Turk J Med Sci
35 (2005) 193-193

LETTER TO THE EDITOR

Wolfram Syndrome (WS), Neither an Autosomal Nor a Sex Linked Disorder*

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Received: May 17, 2005

I have read Kabakuş and colleague's (1) short report in the recent issue of the Journal (35: 51-55, 2005). Although the authors stated "It is established that WS is a mitochondrial disease", they continue "inherited disease as an autosomal recessive trait". The authors repeatedly mentioned this kind of inheritance, which cannot be true.

I would like to emphasize that mitochondrial diseases such as WS are inherited almost always by mutant maternal mitochondrial DNA (mtDNA) and only once so far by paternal inheritance (2).

Since the authors' first two patients were from the same family, most likely their mother had the mtDNA deletion. Since recent evidence indicates that the chance of such a mother to give birth to another child with a similar mutation is 4.11% (1/24), not less than 1% as supposed previously (3). The authors used the term depression (!) anemia (megaloblastic) in the table, without bone marrow examination, folic acid and vitamin B12 determinations.

Although macrocytic anemia may occur in mitochondrial disorders, the authors did not mention MCV finding either. I believe the term depression (!) anemia, needs explanation as of "introversion pallor", "inappropriate death decision". Despite the first two patients of the authors being brothers the second case in the table was indicated as female (!)

Careful reading of the referees would be most appropriate of this kind of papers, I believe.

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2. Schwartz M, Vissing J. Paternal inheritance of mitochondrial DNA. N Engl J Med 347: 576-578, 2002.
3. Chinnery PE, Di Mauro S, Shanske S et al. Risk of developing a mitochondrial DNA deletion disorder. Lancet 364: 592-596, 2004.

* This "Letter to the Editor" was forwarded to the authors of the original case report but they declined to provide a reply.