



WITH FAMILIAL MEDITERRANEAN FEVER: A CASE REPORT

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BACKGROUND: Familial Mediterranean Fever (FMF) is an autosomal recessive autoinflammatory disease characterised by periodic episodes of fever, peritonitis, arthritis and may be complicated by secondary amyloidosis. FMF affects groups of people from around the Mediterranean Sea. Colchicine is the standard therapy in the prevention of both acute attacks and secondary amyloidosis but there some resistant patients. **Anakinra**, an interleukin-1 (IL-1) receptor antagonist indicated for the treatment of the signs and symptoms of rheumatoid arthritis in combination with methotrexate, is also known to affect the severity and the frequency of FMF attacks.

PURPOSE: To describe the evolution of a patient with FMF treated with anakinra as IL-1 blocker, and evaluate the efficacy and safety of this treatment.

MATERIAL AND METHODS: We describe the case of a 53-year-old colchicin-resistant woman suffering from FMF, who is treated with anakinra between April and September 2012 as second line therapy, after several episodes of recurrent fever and abdominal pain. In order to evaluate the treatment the patient's clinical history and analytical data (C-reactive protein) were reviewed.

RESULTS:

→ Anakinra was started with a daily subcutaneous dose (100 mg) associated with oral corticosteroids (metilprednisolone 8 mg). After the first cycle of treatment, the patient was fine, with no recurrent episodes of fever or abdominal pain.

→ C-reactive protein (CRP) fell down from 0,8 to <0,1 mg/dl. There were no injection site reactions.

→ The only remarkably adverse effect was neutropenia (1,4x10⁹/L). Corticosteroids and anakinra doses were reduced till zero and to 100 mg every other day respectively.

DOSAGE OF ANAKINRA AND ANALITYCAL VALUES

WEEK NUMBER	ANAKINRA Subcutaneous DOSE (mg)	CORTICOSTEROIDS (metilprednisolone) DOSE (mg)	CRP (mg/dl)	NEUTROPHILS (10 ³ /μL) [1,9 - 7,90]	LEUCOCYTES (10 ³ /μL) [4,2 -10,60]
0	---	---	0,8	7,3	8,5
1	100/24h	8/24h	---	---	---
2		4/24h	---	---	---
3		---	<0,1	2,4	4,8
4	100/48h	4/48h	---	---	---
5		---	0,2	1,6	3,6
10		4/24h	---	1,6	3,9
22		---	<0,1	1,4	3,8
31		---	---	---	2,4

CRP: PROTEIN C REACTIVE
--- : NO DATA
(*) OUT OF RANGE

CONCLUSIONS:

→ In this case of FMF, anakinra has suppressed successfully the number of attacks and the symptoms, without important adverse reactions and with improvement in quality of life.

→ Controlled trials are necessary to confirm the safety and efficacy of interleukin-1 antagonists in FMF patients.

