Ten-year-format diary of fever episodes kept over a five-year period by a patient with familial Mediterranean fever

Sirs,

The patient, a 61-year-old man, experienced his first febrile episode >38°C with chills and general fatigue on 20 June 2015. The fever spontaneously decreased within 24 hours without medication. The second attack occurred 2 months later and remitted spontaneously. Thereafter he had recurrent, irregular fevers, sometimes as frequently as three times in a month. His younger sister was suspected to have familial Mediterranean fever (FMF) based on pre-operative blood testing for renal cancer in 2020 (Creactive protein, 17.27 mg/dL and serum amyloid A, 2270 μ g/mL). He sought evaluation at our hospital because he believed his recurrent fevers were also due to FMF (1). Blood testing performed while he was afebrile revealed a slight elevation of SAA (15.1 μ g/mL). We did not perform blood testing during a febrile episode. After signing informed consent, we performed a genetic analysis and showed that he had a heterogeneous p.M694I mutation in the MEFV gene.

He maintained a diary with a 10-year-format on which he drew red circles when he had a fever (Fig. 1). In August and September 2019 and January 2020, he drew blue circles (Fig. 1) that indicated a gouty attack. The physician who diagnosed him with "gout" prescribed oral colchicine, which quickly relieved his attacks. However, because of the high dosage of 2 mg/day, he suffered from diarrhoea and consequently did not take colchicine during subsequent attacks. When we showed him an image of erysipelas-like erythema in FMF (2), he commented that the swollen joint of his "gout" was not at the base of the great toe, rather his ankles were erythematous, tender, hot, and swollen. We noted that these attacks were also due to FMF. After the diagnosis of FMF, we advised him to take colchicine, but he did not accept it because he used to suffer from diarrhoea symptoms. However, in November 2020, when febrile attacks became so frequent (nearly every week), he agreed to take colchicine at 0.5 mg/day. More than a year and a half has passed since then and no fever episodes have occurred.

The first and most important step in reaching a diagnosis of FMF is to recognise the clinical presentation with a complete medical history. However, due to the long period from disease onset to diagnosis, it is sometimes difficult to take an accurate medical

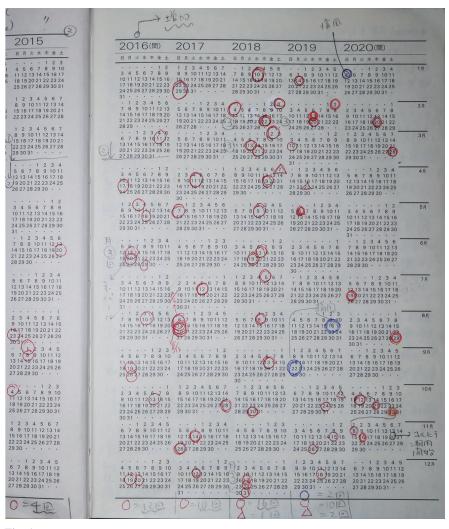


Fig. 1. A diary with a 10-year-format on which the patient drew red circles when he had a fever. Blue circles that indicated a gouty attack was noted that they were due to erysipelas-like erythema also in familial Mediterranean fever.

history from the patients. Especially, in contrast to p.M694V particularly common in the Middle East, patients with p.M694I, which is more frequent in Japanese, have milder symptoms and later age of onset of attacks (3), making the diagnosis sometimes difficult. On the other hand, Japanese FMF patients are known to respond well to low-dose colchicine (3).

Our present case recorded these fever episodes in his diary in a 10-year-format using red and blue circles, and thanks to this 10-year-format, he was able to see at a glance the fever patterns over a 5-year period. We believe this diary is impressive and educative for the awareness of this disease with the characteristic recurrent and periodic fever. His diary also told us the existence of attacks of erysipelas-like erythema. However, even though the site of painful redness and swelling was on the ankles, which is not a common site for gout, his local orthopaedic surgeon had diagnosed him with gout attacks. Although he himself suspected FMF as the cause of his periodic febrile attacks based on his sister's episodes, he did not consider the several attacks on his ankles as FMF lesions until we pointed out the possibility. This also suggests that it is still important to raise awareness of the disease to uncover undiagnosed FMF cases.

Acknowledgements

We thank the patient for granting permission to publish this information.

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EXPERIMENTAL RHEUMATOLOGY 2023.

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