George W. Meyer, MD FACP MACG, Series Editor

Unusual Causes of Abdominal Pain



George W. Meyer

CASE

A 44 year old man presents with a complaint of episodic abdominal pain and fever beginning at age 27. He emigrated to the United States as a young adult from his home in Catania, Sicily. Typically these episodes last roughly 24 hours and occur monthly but have recently increased in frequency. Right shoulder pain, 1-2 hours before the onset of his symptoms, would occasionally precede sudden, severe subxiphoid stabbing pain radiating to the back. Progression would include generalized abdominal pain. The associated fever would last 6-12 hours; there is no vomiting but diarrhea has occurred. When evaluated in the emergency department (ED) his white blood cell count (WBC) and erythrocyte sedimentation rate (ESR) were both elevated. Although resolution of the fever and pain occur within 24 hours, these events cause exhaustion for the next 24 hours. A therapeutic trial was initiated.

See the answer and discussion on page 64.

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ANSWER AND DISCUSSION

Familial Mediterranean Fever (FMF), inherited by autosomal recessive means, affects mostly those whose families have come from around the Mediterranean Sea, such as Sephardic Jews, Armenians, Turks, North Africans and Arabs. Greeks and Italians are less often affected. Most of those who suffer with this disorder develop symptoms in their first decade and 90% are symptomatic by the age of 20. The typical patient will develop sudden onset of fever and abdominal pain (sterile peritonitis), which may last 1-3 days and resolve spontaneously. They may also have pleuritis, synovitis, pericarditis, a skin rash that looks like erysipelas, or orchitis. The abdominal exam may simulate acute peritonitis (guarding, rebound tenderness, rigidity and ileus), leading to exploratory surgery. Abnormal lab results may include elevated WBC, ESR, and CRP. Treatment with colchicine is dramatic in ~75% of patients (fewer than 1 episode in 6 months), usually 0.6 mg per day but as high as 1.8 mg may be needed daily. At least half of non responders were noncompliant patients. The major complication of untreated FMF is amyloidosis (AA type), which may develop after several decades. Differential diagnosis, to be especially considered if the patient does not respond to colchicines, should include: tumor necrosis factor receptor-1-associated periodic syndrome (TRAPS), hyper-IgD syndrome, Muckle-Wells syndrome, and familial cold autoinflammatory syndrome. Genetic testing for FMF is available but is not very sensitive.

References

- http://ghr.nlm.nih.gov/condition/familial-mediterraneanfever
- http://www.mayoclinic.com/health/familial-mediterraneanfever/DS00766

UNUSUAL CAUSES OF ABDOMINAL PAIN

We solicit our readers to submit interesting and unusual cases of abdominal pain for consideration for publication. The case should be well documented, include images (if possible), at least one reference and no more than two authors.

> Send your manuscript to Dr. George Meyer at: **geowmeyer@gmail.com**

Answers to this month's crossword puzzle:

¹E	N	² T	Е	³ R	0	⁴ N		⁵ P	Н	Р	R	⁶ O	В	⁷ E
Α		R		Н		[®] E	D	U				s		N
⁹ S	K	Α	Т	0	10 L	Е		١		¹¹ V	0	М	_	Т
Е		Ν			_			12 P	R	Е		0		Е
¹³ D	Υ	S	Е	Ν	Т	Е	14 R	Υ		15 N	16 	Т	Е	R
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18 B	Ι	L	ı	Α	R	Υ	Т	19 R	Е	Е		²⁰ C	0	G
Е		Α			S			21 N	Е	R	²² O			Α
23 D	0	Ν	0	²⁴ R		²⁵ L	Е	Α	D		26 W	0	R	M
		Т		U		U				²⁷	N			
28 W	Ι			²⁹ D	Ι	M	Е	30 R	I	С		³¹ P	³² S	33 A
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39 T	Е	N		Р		Α		С		⁴⁰ T	41 R	U	Е	
42 E	R	G		Н		43 L	Α	Т		44 Y	Е	Т		

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