

Abdominal Pain May Unmask Subtle Sickle Cell Traits

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A young adolescent girl was consulting for recurrent abdominal pain crisis with severe emesis, fatigue and cephalalgia.

These episodes occurred more than four times during the last few months and the patient had several biological investigations and a myriad of imaging studies; all of them being within the normal limits.

A thorough history and a complete physical examination did not reveal additional findings; but hemoglobin electrophoresis unmasked abnormal hemoglobin S at 39% of total hemoglobin.

A second blood sample confirmed this sickle cell trait; a full familial screening is ordered and the patient was counseled about preventive measures and referred to the hematology outpatient clinics.

This peculiar case is not so uncommon; and many similar reports support a routine evaluation of patient looking for such hemoglobin abnormalities, even without clear evidence of hemolysis, anemia or any other clinical or biological clues [1,2].

The term sickle cell trait is referring to a heterozygous state inherited through an autosomal-dominant gene. The prevalence of sickle cell trait is nearly 8 - 10% in African American population and up to 25 - 30% in western Africa: 2.5 million people in the United States of America and 30 million worldwide do express this heterozygous genotype of sickle cell [3].

In such sickle cell trait, red blood cells free from triggering factors are normal appearing when observed under the microscope; it is only when these cells are under oxidative stress that erythrocytes appear as drepanocytes (or sickle cells) [4,5].

Due to the benign nature of this trait, it usually does not have any clinical implications. As such, patients do not get vaso-occlusive crisis, have a good quality of life and mortality is not overexpressed [4].

However, there have been several reports of adverse conditions that occur due this trait, and sickle cell pain is unique in that it occurs as a hallmark feature in this inherited disorder as early as childhood and throughout the lifetime [5].

The cornerstone of this presentation is the occurrence during oxidative stress like in cold seasons, febrile conditions or in high altitudes; but exception do exist and searching of this rare disease is mandatory in the work up of patients with a heavy history of recurrent abdominal pain; the pain being acute, subacute, chronic, or episodic [4,5].

Evidently, the index of suspicion is higher in endemic areas with high frequency of sickle cell disease (Sub-Saharan Africa, Mediterranean countries) or in high-risk populations (i.e. Population of African ancestry in Europe and the Americas) [3-5].

As so, sickle cell trait may not be asymptomatic and patients should be managed aggressively when they develop some of these painful manifestations.

Recognizing such rare causes of recurrent abdominal crisis like sickle cell trait is crucial to promptly start appropriate management, mainly through a proactive prevention.

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Conflict of Interest

None.

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