CLINICAL IMAGES

Purple Urine: Cause for Concern?

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The patient is an 85 year old, white female presenting with purple discoloration of her urine and collecting bag (Figure 1-2). This condition is found upon routine, monthly physical exam. She is a long term resident of the nursing home and has a chronic, in-dwelling urinary catheter. She has only limited ambulation with the help of nursing staff. She has no complaints at the time of exam and no changes have been made to her care regimen. Her medical and family histories are non-contributory and the remainder of the physical exam shows a frail, somewhat weak constitution but is otherwise unremarkable.

QUESTIONS:

1. What is the diagnosis?

A. Porphyria

B. Medication side effect

C. Purple urine bag syndrome

D. Alkaptonuria

FIGURE 1:

Purple urine with purple discoloration of collecting bag and tubing

FIGURE 2:

Purple urine with red and blue undertones inside Foley bag

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ANSWERS

1. What is the diagnosis?

The correct Answer is:

C) Purple urine bag syndrome

The descriptively named Purple Urine Bag Syndrome (PUBS) is a phenomenon often encountered in chronically catheterized, female nursing home residents. In this condition, red and blue pigments are thought to combine and give the urine and its collecting system a purplish hue[1] (Figure 1-2). Porphyria can sometimes lead to a red or reddish-brown discoloration of urine. There are several types of porphyria but they are all due to defects in the heme biosynthetic pathway. Because of lacking or defective enzymes, the upstream substrates of the heme pathway can accumulate. These excess porphyrins can spill into the urine and give it the classic "port-wine" color.[2] This patient did not have a history of any subtypes of porphyria nor was she exhibiting any symptoms thereof. Also, with porphyria one would expect a more reddish color to the urine as opposed to the striking purple of our patient's sample. Medication side effects should always be considered in the differential but this patient was not on any medications known to cause urine discoloration. Alkaptonuria is a rare, autosomal recessive genetic disease resulting from a defect in the tyrosine catabolism pathway. Upon standing, the urine of individuals with alkaptonuria becomes oxidized and takes on a black color. Alkatponuria is usually associated with other symptoms such as arthritis and ochronosis (deposits of black pigment in various tissues).[3]

DISCUSSION

Purple Urine Bag Syndrome (PUBS) is a rare, albeit strange entity whereby urine and its container take on a purple hue (Figure 1). This phenomenon was first described via the literature in 1978, but descriptions of bluish urine date back to King George III.[1,4] This is primarily a condition of elderly females who have been chronically catheterized. There is also an association with constipation.5 Being a rare and usually benign condition, the epidemiology on the subject is lacking but there does seem to be an increased occurrence amongst nursing home residents. The prevalence has been reported to be as high as 9.8 % in long term residents who are chronically catheterized.[1]

Although the exact mechanism of PUBS remains somewhat elusive, it is thought that bacterial enzymes break down urinary products into visible pigments. The cycle is believed to start with tryptophan, contained in dietary components. In the gut, tryptophan is broken down into indole, which is shunted into the portal circulation. Indole is then conjugated in the liver to indoxyl sulphate. The indoxyl sulphate is then excreted into the urine in high amounts.[6,7] If the enzymes indoxyl sulphatase or indoxyl phosphatase are present in the urine, they can then break down the indoxyl sulphate into indigo and indirubin, which appear blue and red respectively. The combination of these two pigments is thought to create the purple hue (Figure 2). Certain bacteria have been shown to contain the enzymatic machinery (indoxyl sulphatase and indoxyl phosphatase) capable of carrying out these reactions. These include Providencia, Proteus, and Klebsiella species.[5,8] The association with female gender is possibly due to female genitourinary anatomy, which predisposes to UTIs and bacterial colonization. Similarly, catheterization also predisposes to UTIs. High tryptophan content in the diet is thought to provide more substrate for enzymatic degradation into indoxyl sulphate. Constipation slows gastrointestinal transit time and is thought to provide more time for tryptophan degradation. It is also posited that increased urine alkalinity facilitates indoxyl oxidation.[1] At this time, more studies are needed to further elucidate the underlying mechanisms of PUBS.

Treatment of PUBS consists of assurance to the patient and family, who can become alarmed at the drastic and sudden change of urine color. Although usually benign, doctors should be aware that this syndrome can be associated with chronic UTIs and poor urinary hygiene. Medical intervention is aimed at evaluating and treating any underlying infections and correcting any poor Foley practices.[1]

The color of urine has been analyzed by healers since the dawn of medicine. Hippocrates is generally credited with being the father of urinalysis, or uroscopy, as it was called then. However, the study of urine is thought to be much older. As early as 4000 BC, ancient Sumerians and Babylonians studied urine and recorded their findings on clay tablets.[9] Tailabindu Pariksha, an ancient form of Ayurvedic urinalysis, focused on several aspects of the urine, including detailed descriptions of its color.[10] Around 600 years ago, Paracelsus and others started to look toward a biochemical model of urine diagnostics which eventually evolved into the "modern" approach used today.[11] As Western medicine advances and methods of urinalysis become increasingly complex, physicians cannot abandon the use of their own senses in physical diagnosis. In this case, timely recognition of PUBS saved the patient considerable stress and the financial burden of an expensive and unnecessary workup.

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