Secondary Polycythemia in a Patient with Repaired Cyanotic Congenital Heart Disease

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Background

- Approximately 85% of children born with cardiovascular anomalies are surviving into adulthood, making adult congenital heart disease an increasingly relevant for adult providers
- Tricuspid atresia is one of the so called “single ventricle” congenital heart defects

<table>
<thead>
<tr>
<th>Year of Birth</th>
<th>Nick Name:</th>
<th>Performance</th>
<th>Survival Rate (5 Year)</th>
<th>Survival Rate (10 Year)</th>
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<td>1956-1959</td>
<td>2</td>
<td>6.400</td>
<td>70%</td>
<td>10%</td>
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<td>39 years</td>
<td>115.000</td>
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<td>40 years</td>
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PMHx: Tricuspid Atresia s/p valvulotomy at age 1, Glenn procedure at 7 months modified Blalock-Taussig shunt at age 15. Mild LV systolic dysfunction with EF 45-50%. Secondary polycythemia. Gout. Hemorrhoids.

SHx: Married. Smokes 1 cigar daily. Occasional glass of wine.

Physical Exam: T 36.5 HR 70 BP 107/69 RR 16 O2% 90% on 4L NC. Remainder of exam significant for marked clubbing of the fingers, dark purplish discoloration of oral mucosa, soft nontender abdomen, and external hemorrhoids

- Subsequently he had an MRI of his abdomen and pelvis and was found to have nonvisualization of the IVC below the level of the renal veins, most consistent with chronic occlusion and fibrosis, given the presence of extensive venous collateralization
- Flexible sigmoidoscopy was performed and showed only external hemorrhoids
- He was started on therapeutic Lovenox which he took for 2 months before eventually transitioning to Rivaroxaban
- His creatinine and renal function remained stable throughout his hospitalization

Imaging Studies

This patient was felt to have had a renal infarct in the setting of his significant polycythemia secondary to his cyanotic congenital heart disease. Though his Hct was chronically elevated, he had no symptoms of hyperviscosity (headache, increasing fatigue, muscle weakness)

Current ACC/AHA guidelines recommend therapeutic phlebotomy for Hgb greater than 20g/dL and Hct >65% only when patients are symptomatic

This population is at risk for renal dysfunction thought to be due to a glomerulopathy and low uric acid excretion. The resultant hyperuricemia also leaves patients vulnerable to arthropathies and arthritides. Further, these patients may have pigment gallstones from increased red blood cell turnover. Health maintenance recommendations also include endocarditis prophylaxis with antibiotics prior to dental or other invasive procedures, and a flu shot each year

Case Presentation

HPI: A 45 year old male with a history of repaired tricuspid atresia presented with bright red blood per rectum and abdominal pain. The patient reported intermittent hematochezia for 3 weeks prior to the sudden onset of severe right lower quadrant pain lasting 1-2 minutes which subsequently extinguished. Review of systems was otherwise negative.

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- 32nd Bethesda Conference: Care of the adult with congenital heart disease: introduction. Gary D Webb, MD; Roberta G Williams, MD.
- PTed.org

Figure 1: CT abdomen and pelvis revealed decreased cortical enhancement of multiple portions of the right kidney consistent with infarct (see figure arrow)

References

PTed.org