Case Study:

Acute Scrotum: An Atypical Presentation of a Serious Disease

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Case Presentation

An 8-year-old previously healthy Hispanic male presented as a new patient to the primary care provider (PCP) with a 7-day history of bilateral scrotal pain with erythema and edema extending to his penis. The child noted pain in his scrotum 7 days ago with erythema and edema occurring for the past 2 days. He denied fever, abdominal pain, vomiting, and fatigue, but reported a decrease in appetite. He was able to urinate without difficulty. The symptoms were becoming progressively worse, as the erythema and swelling extended to his penis.

Family History

Family history was significant for cancer in the maternal grandfather, who was deceased at age 54 from pancreatic cancer. His paternal grandmother was diagnosed at age 54 years with abdominal cancer and died at age 45 with metastatic cancer.

Physical Examination

The child’s vital signs were as follows: temperature 37 degrees Celsius; pulse 123 beats per minute; respiratory rate 20 breaths per minute; blood pressure 124/68. His oxygen saturation was 99% on room air. He weighed 41.8 kilograms (>95th percentile), and height was 129.54 centimeters (50th-75th percentile). He was alert and oriented to person, place, and time. His skin was pink, warm, and dry; capillary refill was less than 2 seconds. There were enlarged posterior cervical lymph nodes bilaterally, with 6 palpable nodes on the left and 4 palpable nodes on the right, about 1 – 1.5 centimeters in diameter. An enlarged right submandibular lymph node was palpable. No axillary lymphadenopathy was present. Palpable inguinal nodes were present on the right side about 1 centimeter in diameter. His respiratory and cardiovascular exam was normal. He had a rounded and soft abdomen with positive bowel sounds. There was no hepatosplenomegaly. The genital exam revealed an uncircumcised male with a Tanner I sexual
maturity rating. The scrotum and penis were erythematous and edematous. The testes were descended, slightly enlarged, and firm with no hard nodules noted. The anus was patent with no abrasions or fissures.

Management

The child was referred to the emergency department at the area children’s hospital where a testicular ultrasound was normal. He was discharged and instructed to follow up with his PCP the next day. At this follow up visit, he reported decreased pain, swelling, and redness of his scrotum and penis. A complete blood count (CBC) and comprehensive metabolic profile (CMP) was drawn and antibiotics were prescribed at this time for a diagnosis of orchitis. The patient was instructed to return to the clinic within the next week. Lab results were received the next day. The CMP was normal, but the CBC revealed a white count of 22.4 cells/mm3; RBC 3.14 rbc/mm3, hemoglobin of 9.7 g/dl; hematocrit of 26.4%; MCV 84 fl; MCH 30.8 pg/cell; MCHC 36.6%; RDW 17%; platelets of 39,000 platelets/mm3; 24% lymphocytes, and 73% blasts. The findings suggested a diagnosis of leukemia.

The child was referred to a children’s hospital, where he was admitted into the pediatric intensive care unit. A chest x-ray was negative. A second CBC revealed similar results to the previous CBC: normocytic anemia, thrombocytopenia, and numerous lymphoblasts in the peripheral blood smear. The coagulation studies were normal except for the PT which was slightly elevated. The urinalysis was normal. The IgG and IgM findings confirmed that the patient had protection against the Cytomegalovirus (CMV), Epstein-Barr virus (EBV) and did not have a current infection (Corbett, 2008). The Hepatitis B surface antigen (HBsAg) was negative. After a peripherally inserted central catheter (PICC) line was placed, the patient was
transferred to the cancer hospital for an oncology consultation and treatment. His diagnosis was acute lymphocytic leukemia (ALL).

Case Study Questions

1. What are the differential diagnoses for this child’s initial presentation?
2. What are the clinical manifestations for acute lymphocytic leukemia?
3. What is the pathology of acute lymphocytic leukemia?
4. What is the overview management plan for this child with acute lymphocytic leukemia?
5. What is the involvement of the PCP after cancer treatment is complete?

Case Study Answers

1. What are the differential diagnoses for this child’s initial presentation?

Acute scrotum is defined as an acute painful swelling of the scrotum or its contents accompanied by local signs and general symptoms (Khaleghnejad-Tabari et al., 2010). Acute scrotal swelling and pain in a child should always be considered an emergency (Baldisserotto, 2009; O’Brien & Chandran; Gatti & Murphy, 2007; Motta et al., 1997). The differential diagnoses for edema and erythema of the scrotal area include testicular torsion, hydrocele, inguinal hernia, Henoch-Schonlein purpura, idiopathic scrotal edema, trauma, epididymitis, tumor, and torsion of appendix testis, lymphadenitis, and varicoceles. The clinical signs of acute epididymitis, torsion of the appendix testis, and testicular torsion overlap and distinguishing the differences can be very challenging for the health care provider (Beni-Israel, Goldman, Chaim, & Kozer, 2010; Murphy, Fletcher & Pease, 2006). The history and physical examination, timing of the presentation related to the onset of the symptoms, and the age of the child are key components to explore in order to make an accurate diagnosis and referral (Leslie & Cain, 2006).

A child with painful scrotal swelling with an enlarged testis is considered to have torsion of the testis until proven otherwise (Gatti & Murphy, 2007; Motta et al., 1997). Testicular torsion involves nearly 15-40% of all acute testicular pain (Khaleghnejad-Tabari et al., 2010). Testicular
torsion occurs in one in 160 males, with the prevalence from birth to around 60 years old; peak age is at 14 years (Lavalle & Cash, 2005). Testicular torsion is a bending or twisting of the spermatic cord causing severe pain and ischemia of the testicle. Torsion can occur intravaginally or extravaginally. The intravaginal type is the most commonly seen in young and adolescent boys; it occurs within the tunica vaginalis. Extravaginal torsion occurs mainly in neonates in the undescended testicle. The causes of torsion may be unknown or may be related to trauma, which is the cause for the majority of cases (Lavallee & Cash, 2005).

Prompt diagnosis and treatment of testicular torsion is very important because any delay will increase the risk of infarction (Gatti & Murphy, 2007; Kass & Lundak, 1997). Testicular loss can begin after twelve hours of initiation of symptoms (Khaleghnejad-Tabari et al., 2010). Classic presentation of testicular torsion is the sudden onset of unilateral testicular pain and tenderness followed by edema of the scrotum (Beni-Israel et al., 2010). Approximately 50% of patients with testicular torsion have nausea and vomiting and 25% will have fever (Lavallee & Cash, 2005). Other presenting signs can be penile discharge, frequency, and urgency. The most sensitive and specific sign of testicular torsion is absence of the cremasteric reflex on the affected side (Karmazyn et al., 2005; Lavallee & Cash, 2005). Another maneuver that may be used in assessment is to elevate the painful testicle; usually the patient with epididymitis will feel some relief, whereas there will be no relief or worsening pain in the patient who has testicular torsion (Prehn’s sign). This sign, however, is often unreliable (Lavallee & Cash, 2005).

Epididymitis is another condition that can be ruled out for this patient. This condition is very rare in prepubertal children but is the most common cause of acute scrotum in adults (Familua, 2010). It is usually caused by a microbial infection transmitted during sexual intercourse. There may be a history of dysuria, recent fever or mumps and acute onset of
incontinence or dysfunctional voiding (Familua, 2010; Leslie & Cain, 2006). The epididymis will be swollen and tender without true testicular pain. Epididymitis-Orchitis is a unilateral and non-traumatic testicle condition (Yu, Wang, Chen, & Wang, 2012; Lavelle & Cash, 2005). Infection begins at the epididymal tail and because of its rich blood supply; it can spread to the rest of the epididymis and eventually the testis (orchitis) (Wallace & Riley, 2010). The ultrasound appearance of a bacterial epididymo-orchitis, infarction, tuberculosis, and testicular cancer are similar (Pavilica & Barozzi, 2001). Elevating the scrotum takes the pressure off the spermatic cord and there may be some relief from pain. A simple urinalysis that is within normal limits and a negative culture can rule out epididymitis as the etiology for the scrotal pain and edema (Lavallee & Cash, 2005).

Another cause of scrotal pain is torsion of appendix testis. The pedunculated structure of the appendix testis, which arises from the superior portion of the testis and lies within the tunica vaginalis, may lead to torsion (Gatti & Murphy, 2007; Gilchrist & Lobe, 1992). The symptoms of torsion of the appendix testis include unilateral swelling and sudden onset of sharp pain that decreases with time (Gatti & Murphy, 2007; Gilchrist & Lobe, 1992). Torsion of the appendix testis can have a “blue-dot sign” an inflamed and ischemic appendage that can be seen as a subtle blue-colored mass through the scrotal skin (Gatti & Murphy, 2007; Rodriguez & Kaplin, 1988).

Another diagnosis to consider with scrotal swelling is a hydrocele, which is an accumulation of fluid in the scrotum without pain or tenderness. If spontaneous resolution of the hydrocele is not accomplished within the first year of life, it will be surgically repaired (Perry, Hockenberry, Lowdermilk & Wilson, 2010). Hydroceles can cause the scrotum to be tense and appear blue, but it is rarely tender (Gilchrist & Lobe, 1992).
Inguinal hernia is a protrusion of abdominal contents through the inguinal canal into the scrotum (Perry et al., 2010). Approximately 50% of inguinal hernias are observed in the first year of life (Gilchrist & Lobe, 1992). A hernia in a child is seldom a problem except when it is incarcerated or becomes trapped in the hernia sac. The symptoms are severe, sudden onset of pain and a hard, tender fixed mass in the groin (Gilchrist & Lobe, 1992). Torsion of an inguinal hernia sac is extremely rare but it has a similar clinical presentation to acute scrotal swelling in children (Motta et al., 1997). This is considered an emergency and should be evaluated by an urologist. More than 18% of these hernias progress to strangulation and require emergency surgical intervention (Motta et al., 1997).

Henoch-Schonlein purpura is a common systemic vasculitis in children of unknown origin. The clinical signs and symptoms of Henoch-Schonlein purpura (HSP) are a palpable skin rash, abdominal pain and arthralgia; and, in rare cases, acute scrotum pain with slight redness and edema (Hara, Tajiri, Matsuura & Hasegawa, 2004). Approximately 3% of patients with an acute scrotum have been found to have HSP (Clark & Kramer, 1986). External genitalia symptoms with HSP include painful swelling and ecchymosis, with the most commonly involved sites being the scrotal wall, epididymis, testis, testicular appendage and spermatic cord (Hara et al., 2004). The scrotal symptoms may be the first symptoms to appear prior to the diagnosis of HSP and usually disappear within one month; occasionally the symptoms may reappear on the opposite side. It is challenging to differentiate this condition from testicular torsion and may result in unnecessary exploratory surgery (Hara et al., 2004).

The etiology of HSP is unknown; it results from immunoglobulin-mediated inflammation with a high serum IgA level or varying C3 and C4 complement levels (Hara et al., 2004, p. 579). HSP is diagnosed based on the clinical findings of the skin, joints, and kidneys. Laboratory tests
to assess the presence of inflammation and the status of kidney function can be utilized to diagnose HSP. The treatment plan for HSP usually involves short-term administration of steroid therapy or antibiotics (Hara et al., 2004). Laboratory tests, including kidney function tests, and absent physical clinical findings ruled out HSP for this patient.

Idiopathic scrotal edema is an acute onset of edema with an unknown cause (Gatti & Murphy, 2007; Gilchrist & Lobe, 1992). The clinical symptoms are a low-grade cellulitis on one side or both sides of the scrotum in prepubertal boys. The edema occurs rapidly and may involve the groin and the penis; it subsides abruptly and it usually disappears within two days, although slight discoloration of the scrotum can persist. There is usually very little pain with this condition. Management is mainly supportive, and antihistamines or topical steroids can be given for relief of symptoms (Gatti & Murphy, 2007). Idiopathic scrotal edema was not high on the list of differential diagnoses because this patient did have pain accompanying the scrotal swelling.

Scrotal trauma is very common in active boys and it is usually due to a direct blow or a straddle injury. This type of trauma can lead to torsion and can result in a scrotal hematoma or ecchymosis (Leslie & Cain, 2006). A Doppler ultrasound can be very useful to evaluate trauma in the scrotal area. Testicular rupture can be present with a normal ultrasound. If the testicles cannot be detected upon physical examination it is urgent to refer to an urologist. The testicular rupture requires surgical repair (Leslie & Cain, 2006). Scrotal trauma was ruled out in this case because there was no history of trauma or injury.

Scrotal tumors in the pediatric population are rare, representing 1-2% of all pediatric solid tumors (Yazici, Turker & Yucel, 2010). Most pediatric testis tumors are benign, whereas in adults, approximately 95% are malignant (Yazici et al., 2010). Patients with scrotal tumors may present with pain, whereas others report a mass or swelling without pain. (Yazici et al.,
Most of the prepubertal testis tumors are painless scrotal masses. Ultrasound is used to confirm the location of the mass, and helpful in differentiating benign from malignant tumors (Leslie & Cain, 2006). This patient had a normal scrotal ultrasound, ruling out a scrotal tumor.

Lymphadenitis can be either inguinal or femoral in origin (Gilchrist & Lobe, 1992). It is a tender inflamed mass in the groin area and usually follows recent infection. Lymph nodes that are tender or inflamed and do not resolve over time or have a rubbery or firm consistency necessitate a biopsy for possible lymphoma (Gilchrist & Lobe, 1992).

A varicocele is a collection of varicose veins in the scrotum (Gilchrist & Lobe, 1992). Varicoceles are found in 15% of boys, usually around the onset of puberty, who have scrotal swelling with persistent discomfort (Lavelle & Cash, 2005). Physical examination of the cord structures with the patient standing and then supine or with and without Valsalva maneuver will allow detection of the enlarged veins along the cord structure (Leslie & Cain, 2006). Most cases of varicoceles require no treatment and in many cases the varicoceles resolve spontaneously (Gilchrist & Lobe, 1992). Acute varicocele in a prepubertal boy can necessitate an urgent referral because it may be the first sign of a retroperitoneal process such as Wilms’ tumor (Leslie & Cash, 2006). The scrotal ultrasound was normal for this patient and varicoceles were ruled out.

2. What are the clinical manifestations of acute lymphocytic leukemia?

Reported manifestations of acute lymphocytic leukemia (also known as acute lymphoblastic leukemia) include pallor, fatigue, fever, hemorrhage (petechiae) pain, hepatomegaly, splenomegaly, lymphadenopathy, severe headache, vomiting, irritability, lethargy, papilledema, pain, stiff neck and back, muscle wasting anorexia and weight loss. Fatigue (usually from anemia), low-grade fever, night sweats, and weight loss are due to the rapid productions of lymphoblasts and the hypermetabolic effect from these cells. Bleeding,
bone pain, and tenderness are due to bone marrow expansion (Porth & Matfin, 2009). The patient is at risk for infections because of the decreased number of mature white blood cells and the increased number of lymphoblasts or cancer cells. Because of the high number of lymphoblast cells, blood viscosity is increased which predisposes the patient to risk for the development of blood clots that can lead to emboli in the pulmonary and cerebral circulation (Porth & Matfin, 2009).

In this patient’s case, the oncologist discovered during the admission history that the child had a weight loss of 7 pounds over a three-week period. The parents thought it was due to their efforts to decrease calorie intake, as their previous PCP had informed them that the child was obese and needed to lose weight. There were no other presenting findings that were typical of leukemia. Pain, edema, and erythema of the scrotum are not commonly noted manifestation of leukemia.

3. What is the pathology of acute lymphocytic leukemia?

Acute lymphoblastic leukemia (ICD 204.0) (ALL) is the most common form of childhood cancer. The annual incidence is 3 to 4 cases per 100,000. It usually affects males and Caucasians with the peak onset between 2 and 5 years of age (Margolin, Steuber, & Poplack, 2006). ALL is a group of cancers that are composed of precursor B (pre-B) or T (pre-T) lymphocytes that are referred to as lymphoblasts. Leukemia is an unrestricted proliferation of immature white blood cells in the blood-forming tissues of the body. These leukemic cells depress the production of formed elements of the blood in the bone marrow and the normal cells must compete for the essential nutrients for metabolism (Perry et al., 2010).

4. What is the overview for ALL treatment plans for a child with acute lymphocytic leukemia?
Treatment of ALL consists of the use of chemotherapy. There are four phases for treating ALL: induction therapy, intensification therapy, maintenance therapy, and CNS prophylaxis (Porth & Matfin, 2009). Induction therapy eradicates almost all of the cancer cells from the body and uses various chemotherapy agents to achieve remission. Intensification therapy further reduces leukemia cells after remission has been achieved. Chemotherapy agents are administered at higher doses during intensification therapy. The primary goal of CNS prophylaxis is to ensure that no leukemic cells have crossed into the cerebrospinal fluid. This phase involves the use of high doses of chemotherapy agents that can penetrate the blood-brain barrier. Maintenance therapy maintains remission, and the chemotherapy agents are given at lower doses over a long period of time (Porth & Matfin, 2009). The chemotherapy agents used to treat acute lymphocytic leukemia can have many serious side effects that require careful monitoring and early intervention to promote remission. ALL was at one time considered a universally fatal disease; it now boasts overall cure rates ranging from 75% to 85% (Belson, Kingsley, & Holmes, 2007; Campbell, Scaduto, Van Slyke, Niarhos, Whitlock & Compas, 2009; Colby-Graham & Chordas, 2003).

5. What is the involvement of the primary care provider (PCP) after cancer treatment is complete?

The PCP role has a role of utmost importance in identifying presenting signs and symptoms associated with cancers and initiating referral to the specialist for treatment. Although the oncologist will monitor the child during the treatment phase, the child will return to the PCP for health care maintenance and management of usual childhood illnesses. Throughout the cancer treatment cycle, the PCP should maintain the relationship with the child and the family.
Haddy and Haddy (2010) suggest that there are four phases of the PCP role in the care of the patient with cancer. The first phase is the early identification of cancer in the child. The PCP should take all complaints, even those seemingly benign, seriously. Early identification of symptoms leads to better outcomes. This child who presented with a red swollen scrotum had signs and symptoms that were taken seriously and treatment was initiated without delay.

The second phase of the PCP involvement is to remain informed and involved during the child’s treatment (Haddy & Haddy, 2010). It is helpful for the oncologist to send progress updates of the child during cancer treatments to the PCP. Emotional support should be provided to the patient and family during ongoing treatment. Sometimes parents may have to make decisions about certain treatment plans and whether to accept or discontinue treatment. The PCP can offer support by discussing options and alternative choices with the parents (Haddy & Haddy, 2010). Routine medical care for siblings and proper attention to the health concerns of the other family members should also be encouraged.

The third phase follows after treatment is completed (Haddy & Haddy, 2010). The PCP continues to be involved in the child’s routine medical care and continuing to observe for any abnormal signs and symptoms related to the late effects of cancer treatment. Growth and development should be monitored during well child examinations, which should include nutritional assessment as well as screening for vision, hearing, and laboratory tests as indicated (Baggott, 2010). Children who are immunocompromised should not receive any live-virus vaccines (American Academy of Pediatrics Committee on Infectious Diseases, 2006). Inactivated vaccines administered during this time may not be effective in these immunosuppressed children. Oncologists often withhold all vaccines during cancer treatment.
and up through a year afterwards (Allen, 2007). It is best for PCPs to collaborate with the oncology team for immunization recommendations (Baggott, 2010).

Life-long surveillance and interventions are essential in the care of children with childhood cancer (Haddy & Haddy, 2010). Problems related to the late effects of childhood cancer include growth impairment, problems in cognitive functioning, and organ damage (Baggott, 2010). These children are more likely to report depression and posttraumatic stress disorder compared to the general population (Langeveld, Grootenhuis, Voute, & de Haan, 2004). These major health concerns require additional time for assessment in the cancer survivor.

The most serious of the late effects are the secondary cancers that can emerge after successful treatment of the primary cancer. Some of these secondary cancers are treated with success but others, such as brain cancer, may not be curable (Haddy, Mosher, Dinndorf, & Reaman, 2004). The cancer survivor risks can continue for 30 years (Hijiya et al., 2007).

The fourth phase occurs if treatment fails and death is inevitable. The PCP should be available for the family should they decide to receive hospice (Haddy & Haddy, 2010). It is important that the PCP remains involved with the patient and the family from the identification process through the completion of treatment (Haddy & Haddy, 2010).
References


