

1st Place

Title:

A case of Kasabach-Merritt Syndrome

Authors:

Shilpa Reddy, MD, Barrett Zlotoff, MD, Emily DeSantis, MD

Abstract:

We report a case of a 2-month-old female who presented to our Dermatology clinic for a 4-5 week history of an enlarging violaceous plaque on the pelvic region. The patient was 2 weeks premature but otherwise healthy at birth with no medical problems and no skin lesions. The patient's mother stated that the lesion initially started off as a small linear discoloration that progressively grew larger and darker in color over a several week period. Review of system was negative and the patient had no treatment to the area prior to presentation in our clinic. A biopsy was performed which showed a kaposiform-hemangioendothelioma. Labs included a platelet count of 110,000, fibrinogen level of 174, and a D-dimer > 20. A diagnosis of Kasabach-Merritt Syndrome was made and the patient was admitted to the hospital for observation and further management. Kasabach-Merritt Syndrome is a rare thrombocytopenic consumptive coagulopathy associated with either tufted angioma or kaposiform hemangioendothelioma, first described in 1940. Consumption of platelets and coagulation factors in addition to ongoing fibrinolysis results in intra-lesional bleeding, anemia, and enlargement of the hemangioma. Other complications include congestive heart failure and compression of vital organs, which can be fatal. Mortality is 12%-24%. Ultrasound, MRI, and CT are all methods that can be used to determine the extent of the lesion. The goal of treatment includes decreasing the size of the lesion and normalization of the hematologic abnormalities (correction of thrombocytopenia and anemia). According to a literature review of current treatment options, the first line treatment is oral corticosteroids which can reverse the platelet abnormalities, but not necessarily lead to complete resolution of the tumor. Other commonly used medications include interferon alpha and vincristine. In our patient, propranolol was used initially with no significant response. She subsequently was started on oral corticosteroids and is currently undergoing treatment with that.

References :

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2nd Place

Title:

Potential exsanguination from a punch biopsy: what dermatologists need to know about lower extremity arterial bypass grafts

Authors:

Jennifer L. Pike, MD, Katherine H. Fiala, MD, David F. Butler, MD

Abstract:

A 66 year old Hispanic male presented as a new patient consult to be evaluated for nephrogenic fibrosing dermopathy secondary to a previous magnetic resonance angiography that took place 5 months prior to presentation. His past medical history included diabetes mellitus with diabetic retinopathy and nephropathy, stage IV chronic kidney disease, hypertension, anemia of chronic disease, and hypothyroidism. The patient complained of significant pruritus involving his forearms and trunk. On physical exam the patient had diffuse xerosis with mild lichenification of his forearms and trunk. It was also noted that the patient had diffuse woody induration with lichenification and hyperpigmentation involving much of his right lower extremity. Upon further questioning the patient did report having surgery for ulcers involving his right lower extremity several months earlier. To evaluate the possibility of nephrogenic fibrosing dermopathy, a 5 mm punch biopsy was planned from the right lateral thigh. The punch biopsy had not yet reached the hub when a massive arterial bleed through the end of the punch biopsy instrument occurred. Pressure and a tourniquet were attempted and emergency services were contacted. The patient was quickly transported to the emergency room where he was transfused and then taken to the operating room for repair of an arteriotomy to his right femoral to anterior tibial polytetrafluoroethylene (PTFE) bypass graft. The patient fully recovered.

We will review what dermatologists should know about the placement of lower extremity arterial bypass grafts, particularly femoral to anterior tibial bypass grafts as in the case presented, including: materials used (autogenous venous grafts vs. PTFE grafts vs. composite grafts), possible locations of bypass grafts (medial vs. lateral), and methods of tunneling bypass grafts (subcutaneous vs. interosseous vs. tibial tunneling). It is important for dermatologists to be aware that lower extremity bypass grafts can be tunneled in the superficial fat on the lateral aspect of the thigh and that prosthetic bypass graft materials, such as PTFE grafts, may not palpable or pulsatile.

3rd Place

Title:

Severe Acanthosis Nigricans and Tripe Palms without Evidence of Malignancy

Authors:

Marshall J Shuler, MD, Barrett J Zlotoff, MD, Emily DeSantis, MD

Abstract:

In 2004, a 15 year old African-American female presented to our clinic complaining of a 6 month history of progressive thickening and darkening of the skin. On exam, the periorbital face, posterior neck, presternal chest, abdomen, axillae, and antecubital fossae were noted to have an increased prominence of skin lines with a consequent velvety texture. These findings were accompanied by a palmar-plantar hyperkeratosis and fissured tongue. With the exception of the furrowed tongue, the patient claimed all of these changes were of recent onset.

At presentation she underwent punch biopsy of the posterior neck and blood tests including a complete blood count, comprehensive metabolic panel, and serum insulin. At 34uIU/ml (normal 6-27 uIU/mL), her insulin level was slightly elevated, but all other tests were within normal limits and the patient's biopsy was considered be "consistent with acanthosis nigricans."

Over the next 4.5 years the patient was lost to follow-up. She then returned to our clinic at 7 months gestation with a normal pregnancy complaining of a 2 week history of tender rapidly growing and expanding clumped nodules on the neck, with fungating masses draining malodorous purulent material in the bilateral axillae and inguinal folds, and papillated tumors in the postauricular sulci. Additionally, her entire skin was noted be severely xerotic with dramatically accentuated skin lines and exaggerated dermatoglyphics consistent with tripe palms noted on the palms and soles. Punch biopsy and wound cultures were obtained and the patient started on empiric therapy with Cephalexin. Punch biopsy results demonstrated "marked epidermal acanthosis and papillomatosis with subcorneal and intraepidermal pustules."

The patient returned to clinic 7 days later and was noted to have increased skin tenderness and general malaise. Since wound cultures were positive for *Klebsiella oxytoca*, *Serratia marcescens*, and *Staphylococcus capitis*, decision was made to admit her to the hospital and start intravenous (IV) antibiotics. After 48 hours of Zosyn 3.375g every 6 hours IV the drainage in the patient's axillae and groin decreased and the tumors began to shrink in size. At this point, cancer screening using serological markers, ultrasound, and esophagogastroduodenoscopy were undertaken to rule out occult malignancy and were all negative.

Today, she remains 8 months gestation with a viable pregnancy and persists with startling acanthosis nigricans and tripe palms of 5 years duration. Without evidence of malignancy, we present this case for suggestion and consideration of other underlying physiologic abnormalities which may be driving this process.