Case Report: Eruptive Xanthoma in a 14-year-old boy

INTRODUCTION
Eruptive xanthomas (EX) are often an indicator of severe hypertriglyceridemia and can indicate undiagnosed or decompensated diabetes mellitus (DM). The triglyceride levels in these patients can be >3000-4000 mg/dl (normal <150 mg/dl). It is important to quickly diagnose and manage these patients due to the possible sequelae of diabetes mellitus and hyperlipidemia. Patients with eruptive xanthomas due to hypertriglyceridemia are also at increased risk for acute pancreatitis. The diagnosis of eruptive xanthoma is often made easily based on the clinical picture although appropriate labs and biopsies may also be necessary. While this condition is typically seen in adults it may also manifest in children. When it presents in the pediatric population the most common finding is either uncontrolled or undiagnosed type-1 DM. Incidence in this population is difficult to estimate, but a recent PUBMED search using the search terms eruptive xanthoma revealed 180 articles while a subsequent search using the terms pediatric eruptive xanthoma incidence returned only 4 articles.

CASE REPORT
An obese, 14-year-old, Native American male, presented to his pediatrician complaining of a mildly itchy rash that started on his upper extremities. Over the next 10 days it migrated onto his torso and legs. Parents admitted to starting a new laundry detergent but denied being exposed to any other new products - including foods, medications, or lotions prior to the rash. The patient applied an OTC topical corticosteroid cream for the rash which resolved and triglycerides had returned to normal levels. The patient was otherwise well and thriving. Family history was remarkable for obesity, hyperlipidemia, and DM was present on both sides of the family.

On physical examination the patient had multiple crops of 1-3 mm non-umbilicated, yellow to red papules on his arms, legs, and torso (Figure 1). Labs were ordered and serum triglyceride levels were 2,100 mg/dl, (normal <150 mg/dl) with serum cholesterol levels at 500 mg/dl (normal <200 mg/dl). Fasting serum glucose levels were 250 mg/dl (normal 79-99) with a hemoglobin A1c of 13% (normal <5.7%).

Patient was referred to dermatology for consultation at which time a punch biopsy was performed. Biopsy confirmed the diagnosis of EX and labs suggested it was most likely secondary to uncontrolled hyperlipidemia and diabetes mellitus.

In a 2008 study of 47 pediatric patients the effects of Gemfibrozil was shown to be efficacious and well tolerated. The mean decrease in triglycerides was 47% while the mean HDL increased by 20%. Our patient was subsequently started on Gemfibrozil, referred to endocrinology to manage his insulin regimen and follow up with a diet and nutrition specialist. Follow up one week later already showed improvement of the lesions as well as the pruritis. Within one month the lesions almost completely resolved and triglycerides had returned to normal levels (Figure 2).

CLINICAL PHOTOS

Figure 1

Figure 2

DISCUSSION
Xanthomas are benign collections of lipid deposits that manifest as yellow to red firm papules, nodules, or plaques. This yellowred coloration is due to the carotenoids in lipids. Xanthomas are composed of lipid filled macrophages known as “foam cells”. Xanthomas can be due to genetic disorders of lipid metabolism, such as autosomal dominant hypercholesterolemia or familial hypertriglyceridemia (familial hypertriglyceridemia). They can also be caused by severe hypertriglyceridemia and have been associated with undiagnosed diabetic dyslipidemia, hypothyroidism, cholestasis, nephrotic syndrome, biliary atresia, and Alagille syndrome. Some cases have been reported in patients with alcohol dependence or medication use. Most xanthomas erupt in adulthood, however, xanthomas due to homozygous familial hypercholesterolemia can present in childhood.

There are multiple types of xanthomas including tuberosus, tendinous, disseminated and eruptive. Eruptive xanthomas are 1.5 mm papules that often develop rapidly and can present as a papular rash. They are most common on extensor surfaces as well as the buttocks and shoulders. Eruptive xanthomas have been noted to display the Koebner phenomenon - new lesions occurring in areas of trauma. Biopsy of the xanthomatous papules will show infiltration of foam cells and multinucleate giant cells within the reticular dermis on hematoxylin-eosin staining (Figure 3-5). In early lesions histiocytes are numerous and fully developed foam cells are small in number. In an established papule, xanthoma cells have characteristic clear or foamy cytoplasm as the predominant cell type. They may also contain admixture of lymphocytes and neutrophils. Eruptive xanthomas tend to exhibit smaller and fewer foam cells than other types of xanthomas. They also tend to be predominantly triglyceride in the microcytoplasmic space while the other forms of xanthoma will have predominant amounts of cholesterol. The quantity of the lipid are also in a state of flux - this is thought to be associated with extracellular deposition - and this phenomenon is rare or absent in the other types of xanthomas. Laboratory analysis should be performed on all patients with a new diagnosis of eruptive xanthoma with unclear origin to evaluate for causes of hyperlipidemia. Labs looking for underlying metabolic disorders should include a fasting lipid panel and glucose level, HbA1c, CBC, thyroid functions, as well as liver and renal function panels.

Treatment of eruptive xanthomas is based on resolving the underlying condition. A holistic approach involving diet modification, exercise and drug therapy is necessary to manage the hypertriglyceridemia and hyperglycemia. Eruptive xanthomas typically regress quickly with treatment of the lipid abnormality and underlying cause.

REFERENCES