

replacement treatment in congenital adrenal hyperplasia.⁶ Continued use of hydrocortisone is recommended throughout childhood and puberty until statural growth is almost complete. When statural growth is complete, dexamethasone is an effective form of treatment to use in the long term. The longer biological half life allows medication to be administered only once, 2 or at the most, twice daily. Single daily medication should probably be given in the morning in a maximum dosage of 0-01 mg/kg/day.

It is essential for the infant to be seen early by a surgeon experienced in the techniques required for reconstruction of the genitalia. There are basically two structural abnormalities which require surgical treatment: reduction in the size of the enlarged clitoris and division of the fused labial folds to exteriorise the vaginal opening. The parents should understand that the female internal genitalia are normally developed. Pictures from a pelvic ultrasound examination or vaginography to show the uterus and ovaries can be most reassuring. Ultrasonography by an experienced sonographer is often employed to visualize the presence of female reproductive organs. It is important to establish the exact location of the vagina in relation to the bladder. Particular attention must be made to delineate the size of the vagina and most importantly the level of the confluence of the vagina with the USG. This information is invaluable in preoperative planning as well as for counseling parents regarding the extent of surgery required for vaginoplasty.^{7,8}

CAH poses many challenges, especially for women which include the issues of genital surgery, disclosure, informed consent, weight, and general well-being not to mention having a chronic condition that is lifethreatening. The management of patients with genital anomalies is a complex problem. The goal remains to correct the visible anatomical anomalies, creating an appearance corresponding to the gender, and a function enabling the individual to lead a normal life, including sexual function and, if possible, reproduction. Clitoral reduction,

especially in an adult, is a procedure which often leaves the glans clitoral without the capacity for tactile sensation.⁹ Recent investigations and reports on longterm results indicate that vaginal orgasm is more of an exception than the rule, so that for women, preservation of clitoral sensitivity is essential to a satisfying sexual life. The technique of clitoroplasty should be modified according to the size of phallus.

More research is now being carried out into the psychological long-term outcomes of women with CAH. Depression and stress are often reported,

particularly to do with relationship, weight problems associated with steroid replacement. These patients are reared as females, but have depression due to clitoral hypertrophy, imagining themselves neither female nor male. Multidisciplinary one stop patients centered care with endocrinology, gynaecology, and psychology expert teams are now gradually becoming the cornerstone of care for CAH.

CONCLUSION

Congenital adrenal hyperplasia is an uncommon chronic disorder which will present in the newborn nursery only about once every two years even in the busiest maternity unit. A high clinical index of suspicion should be maintained for all infants with abnormal genitalia

Management of virilizing CAH child with ambiguous genitalia demands multidisciplinary team approach. Various specialties such as pediatric endocrinologist, urologist, psychologist, gynecologist should work as a teamwork to achieve a normal physiologic, emotional and sexual development. The team major challenge is to use the tools available to make the least bad choice in order to select a gender able to match the individual identity, social identity, and behavioral identity. Patients well being can be achieved by team work, sustained follow-up and patient's compliance. Surgical option, performed by experienced surgeon or pediatric urologist, should be tailored, single stage surgery, at early age and executed according to the individual anatomy findings.

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Eruptive Xanthomas – A Study of 10 Cases

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Abstract: Cutaneous xanthomas are a manifestation of underlying lipid abnormalities. It is marked by sudden eruption of crops of small, yellow or yellowish orange papules encircled by an erythematous halo, especially on the buttocks, posterior thighs, and elbows, and caused by high concentrations of plasma triglycerides, especially that associated with uncontrolled diabetes mellitus. We selected 10 cases of eruptive xanthomas for our study. A complete lipid profile of the patients was done including serum cholesterol, triglycerides, HDL, VLDL and LDL. All the patients were subjected to skin biopsy and it was confirmed by histopathological examination. Back was the commonest site being involved in 90% patients, arms and legs were involved in 80% patients, abdomen was involved in 40% patients and face was involved in 20% patients. Family history of hyperlipidaemia was positive in two out of ten cases. Biopsy of the patients showed a small nodular granulomatous infiltrate in upper dermis with accompanying sparse perivascular lymphocytic infiltrate. The granuloma consisted of pale stained histiocyte with abundant foamy cytoplasm.

INTRODUCTION

Eruptive xanthomas are characterized by crops of yellow papules with an erythematous base, usually occurring over the pressure points on extensor surfaces and buttocks.¹ There are various types of xanthomas classified by both clinical and histopathological criteria. Eruptive

xanthomas are associated with hypertriglyceridemia and chylomicronaemia due to genetic disorder for an underlying disease process (secondary hyperlipoproteinemia), such as diabetes mellitus, hypothyroidism, nephritic syndrome and pancreatitis.² Although, eruptive xanthomas are usually associated with hyperlipoproteinemia, they can occur in normolipemic patients with local trauma.³ Treatment involves dietary restrictions and medications to control the hyperlipidemia. The aim of the study was to analyse the clinical and histopathological characteristics of 10 cases of eruptive xanthomas.

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MATERIAL AND METHODS

We selected 10 cases of eruptive xanthomas for our study. A detailed clinical history of all the patients was taken and a thorough physical examination was conducted to see the site, type and approximate number of xanthomas. All routine investigations of the patients were done. A complete lipid profile of the patients was done including serum cholesterol, serum triglycerides, HDL, VLDL and LDL. All the patients were subjected to skin biopsy and it was confirmed by histopathological examination.

RESULTS

The data was collected and the results were analyzed statistically. It was seen that 50% of xanthomas were seen in the age group of 41 to 60 years, 40% were seen in the age group of 21 to 40 years and 10% of xanthomas were seen in the age group below 20 years. Males outnumbered females and male: female was 4:1.

In majority of cases, Xanthomas were seen on the arms and leg (Fig. 1&2); a fair number seen on the surface of abdomen; and very few had xanthomas on the face (Table 1) estimation of serum cholestrol and triglycerides revealed normal levels in 5 of 10 cases; in 2 of them the levels were higher (Table 2).

Table 1: Table Showing Sites of Distribution of Xanthomas

SR NO	SITE OF DISTRIBUTION	NUMBER	PERCENTAGE
1	ARMS & LEGS	8	80%
2	ABDOMEN	4	40%
3	BACK	9	90%
4	FACE	2	20%

Table 2: Table Showing Lipid Profile of Patients

SR NO	LIPID LEVELS(CHOLESTEROL AND TRIGLYCERIDES)					
	NORMAL		HIGH		VERY HIGH	
1	NO OF PTS	%	NO OF PTS	%	NO OF PTS	%
5	5	50%	3	30%	2	20%



Fig. 1: Eruptive xanthomas over the elbows in a 40 years old male



Fig. 2: Eruptive xanthomas on the back of a 42 years old male

Family history of hyperlipidaemia was positive in two out of ten cases. Biopsy of the patients showed a small nodular granulomatous infiltrate in upper dermis with accompanying sparse perivascular lymphocytic infiltrate. The granuloma consisted of pale stained histiocyte with abundant foamy cytoplasm (Fig. 3). Some of the histiocytes appeared to form giant cells. At places the histiocytes were interstitial whereas at other foci they were surrounded by mild fibroplasia. Overlying epidermis was unaffected.

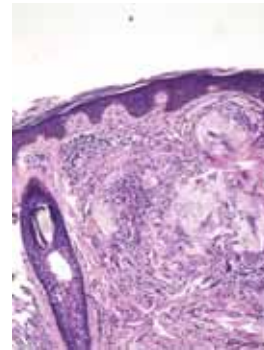


Fig. 3: Granulomatous infiltrate in upper dermis with foamy cytoplasm (H&E stain 100X)

DISCUSSION

Eruptive xanthomas is a condition affecting individuals with severe hyperlipemia in which groups of waxy yellow or yellowish-brown lesions appear suddenly, especially over extensors of the elbows and knees and on the back and buttocks. The lesions are usually 3 to 7 mm and firm to palpation. Xanthomas not always associated with underlying hyperlipidemia, but when they are, diagnosing and treating underlying lipid disorders is necessary to decrease the size of the xanthomas and to prevent the risks of atherosclerosis.⁴ Diagnosis of eruptive xanthomas includes measuring fasting blood glucose and lipid levels, including triglycerides, cholesterol, low density lipoproteins and very low density lipoproteins.^{5,6} The patients biopsy shows a dermal infiltrate of foamy macrophages with lymphocytes and neutrophils, which is suggestive of eruptive xanthomas.⁷

Treatment of the hyperlipidemia initially consists of diet and lipid-lowering agents such as statins, fibrates, bile acid-binding resins, probucol, or nicotinic acid.^{8,9} The lipid-lowering effects of these agents have been well documented, but few studies mention the efficacy of these drugs for resolving xanthomas. Eruptive xanthomas usually resolve within weeks of initiating systemic treatment. The main goal of therapy for hyperlipidemia is to reduce the risks of atherosclerotic cardiovascular disease. In patients with severe hypertriglyceridemia, the goal is to prevent pancreatitis.

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