The coexistence of XD with hypopituitarism is a rare condition. There are various systemic treatments such as radiotherapy, cryotherapy, corticosteroids, and antiblastic chemotherapy but no single treatment is universally successful. In rare cases complete remission was obtained with low-dose oral cyclophosphamide in adults as it occurred in our case.

Succesful Systemic Treatment Of Xanthoma Disseminatum With Cyclophospamide: An Interesting Case With Endocrine And Gastrointestinal Involvement

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INTRODUCTION

- Xantoma disseminatum (XD) is a rare non-Langerhans cell histiocytosis (NLCH) which is often resistant to treatment.
- In this report, we presented a case with extensive cutaneous, hypothalamohypophysial, cerebral and gastrointestinal system involvement, which responded well to cyclophosphamide

CASE

- A sixteen-year-old female patient admitted to our hospital with the complaints of amenorrhea, weight gain, polidipsia, poliuria, yellow-brownish papular lesions on the cervical, periorbital, axillary and genital regions.
- Lesions first appeared 18 months ago and increased in amount and size in time.
- Hormonal evaluation was done including dynamic tests and secondary hypothyroidism, hypogonodothropic hypogonadism, growth hormone deficiency and central diabetes insipidus were detected.
- Pituitary MRI demonstrated a mass 15x8 mm in diameter at hypothalamohypophysial tract together with multiple cerebral lesions. Her visual and neurologic examination was normal.
- A biopsy was performed on skin and duodenal lesions and the result of pathologic analysis was coherent with NLCH.
- In the light of those findings, she was diagnosed with XD.
- Hormonal replacement therapies for hypothyroidism, hypogonadism and DI were initiated. For hypothalamic mass and skin lesions 60 mg/day methyl prednisolone was started and its dosage was gradually reduced to maintenance dose of 4 mg/d.
- MRI screening performed at 6th month of therapy didn't show any regression in mass sizes. Hence, medical therapy was changed with cyclophosphamide 100 mg/d.
- No complete remission was achieved but significant regression in cutaneous, hypothalamohypophysial, cerebral and gastrointestinal lesions was obtained in 24 months.
- No side-effects were noticed related with cyclophosphamide. The skin lesions did not relapse after discontinuation of cyclophosphamide.

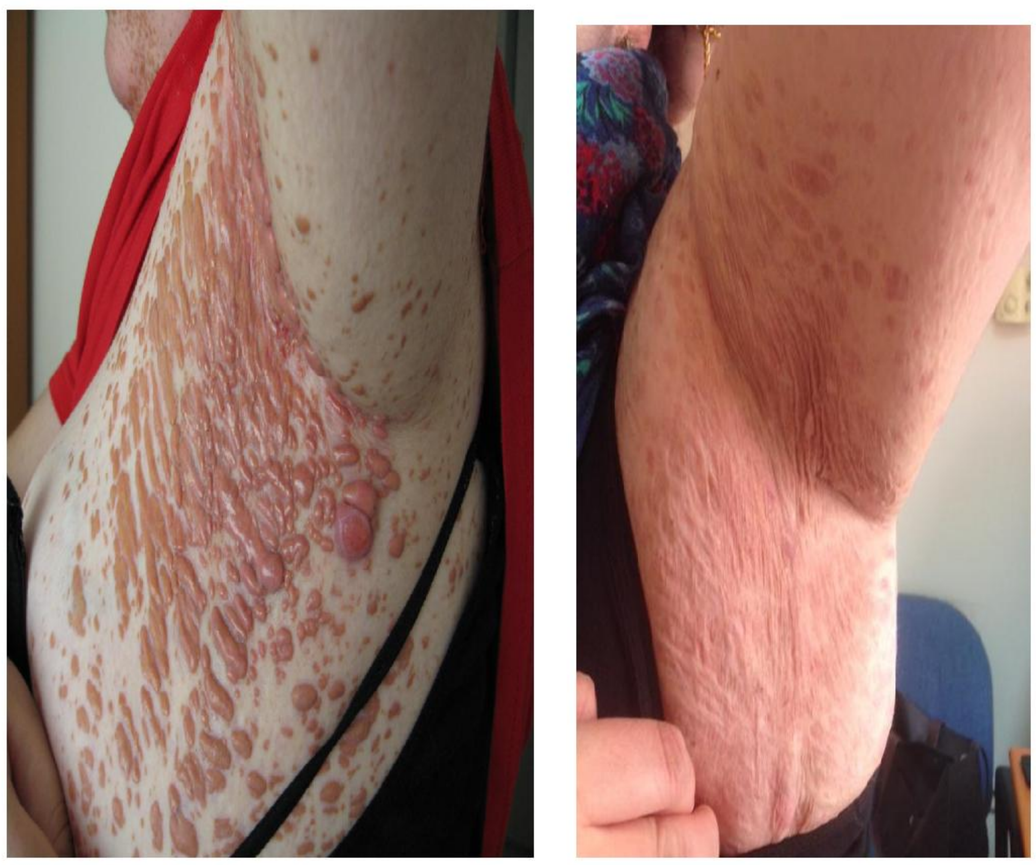

Figure : Pre and post treatment skin lesions of the patient

CONCLUSION

- The coexistence of XD with hypopituitarism is a rare condition. There are various systemic treatments such as radiotherapy, cryotherapy, corticosteroids, and antiblastic chemotherapy but no single treatment is universally successful.
- In rare cases complete remission was obtained with low-dose oral cyclophosphamide in adults as it occurred in our case.

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