

# Eruptive syringomas in Down's syndrome

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## ABSTRACT

Down's syndrome is a common chromosomal anomaly characterized by mental retardation and distinct physical appearance. It is associated with a large variety of dermatological disorders like anetoderma, cheilitis, cutis marmorata, elastosis perforans serpiginosa, fissured and geographic tongue, onychomycosis, palmoplantar hyperkeratosis, psoriasis, syringomas, alopecia areata and vitiligo. Syringomas are benign adnexal tumors which have been reported to occur with higher frequency among patients with Down's syndrome. We report a case of eruptive syringomas over face and hand in an 8-year old child with Down's syndrome.

**Key words:** Syringomas; Trisomy 21; Down's syndrome

## INTRODUCTION

Down's syndrome is a common chromosomal anomaly associated with multiple malformations, medical conditions, and cognitive impairment because of the presence of extra genetic material from chromosome 21. The typical physical findings include hypotonia, brachycephaly, epicanthal folds, flat nasal bridge, upward-slanting palpebral fissures, small mouth, small ears, excessive skin at the nape of the neck, single transverse palmar crease, and short fifth finger with clinodactyly and wide spacing between the first and second toes [1]. Down's syndrome is also associated with accelerated aging and an increased incidence of a variety of dermatological disorders like anetoderma, cheilitis, cutis marmorata, elastosis perforans serpiginosa, fissured and geographic tongue, onychomycosis, palmoplantar hyperkeratosis, pityriasis rubra pilaris, psoriasis, seborrheic dermatitis, syringoma, alopecia areata, vitiligo and xerosis [2,3].

Syringomas, benign adnexal neoplasia derived from eccrine ductal elements, are found in a higher frequency among patients with Down's syndrome. We report a case of eruptive syringomas over face and hand in an 8-year old child with Down's syndrome.

## CASE REPORT

An 8-year old male with Down's syndrome was brought to us with the complaints of gradually progressive asymptomatic skin lesions over the face and hands for the last six months. The lesions started over the face and gradually increased in number and over the course of time appeared over dorsa of hands too. On cutaneous examination, numerous skin colored papules were present over the cheeks, forehead, periorbital area, bridge of nose and dorsa of hands (Fig. 1). There was no associated hypohidrosis and the patient's nails, hair and teeth were normal. The patient was advised a skin biopsy which was refused by the parents owing to the asymptomatic nature of the lesions. The patient was advised topical tretinoin 0.025% gel application over the lesions but there was no improvement in the lesions after three months of application.

From the history and clinical examination, a diagnosis of eruptive syringomas was made owing to the increased prevalence of syringomas in Down's syndrome patients.

## DISCUSSION

Down's syndrome is one of the most common autosomal chromosomal disorders with an incidence

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**Figure 1:** Multiple skin colored papules over the a) face and b) dorsa of hands.

of around 1 in 800 live births. Apart from numerous physical traits, a large variety of dermatological conditions with increased prevalence or increased severity are seen in patients with Down's syndrome [1]. Various authors have reported an increased prevalence of skin conditions like anetoderma, cutis marmorata, cheilitis, palmoplantar hyperkeratosis, pityriasis rubra pilaris, xerosis and syringomas [2,3]. A higher incidence and severity of alopecia areata, vitiligo and atopic dermatitis has also been observed which has been attributed to immunological deficiency in T-cell function seen in patients with Down's syndrome. The B- and T- cell function and number is also reduced in these patients leading to a higher risk of infections and malignancies [2,4].

Syringomas are benign appendageal tumors of eccrine origin, most typically found in the periorbital region. Syringomas have been classified into four groups, according to their clinical features and associations, namely localized form, generalized/eruptive form, a form associated with trisomy 21 and familial form [5]. Clinically, they appear as small skin colored papules, rounded or flat-topped with angular margins, varying in size from 1 to 5mm. The front of the chest, face and neck are the main sites of involvement. Eruptive

syringomas may appear on the neck, chest, abdomen and buttocks. The typical histological features include multiple small ducts lined by rows of flattened epithelial cells with epithelial strands within the dermis, giving rise to the characteristic tadpole appearance.

Syringomas are more common in patients with Down's syndrome and twice as common in female patients. The incidence of syringomas in Down's syndrome has been reported to be approximately 30 times greater than in the general population and syringomas of the eyelids have been reported exclusively in Down's syndrome. Studies have reported an incidence varying from 18% to 39% in patients with Down's syndrome [2,3,5].

Clinically, syringomas resemble trichoepitheliomas but syringomas tend to be smaller, rather more flat-topped and disposed more evenly over the cheeks and eyelids, rather than favoring the nasolabial creases. The treatment is mainly for cosmesis and various treatment modalities like surgical excision, electrocautery, cryotherapy, dermabrasion, trichloroacetic acid and carbon dioxide laser ablation have been used.

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