# A Case of Juvenile Xanthogranuloma Associated with Neurofibromatosis

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**We report a case of juvenile xanthogranuloma associated with neurofibromatosis. The patient was a 10-month-old female who had multiple yellowish papules on the face and shoulders A Biopsy of these lesions revealed foamy histiocytes with an admixture of inflammatory cells and Touton giant cells. Multiple, more than 6, cafe-au-lait spots, measuring more than 1.5cm in diameter were scattered on the trunk and buttocks Retardation of physical growth and development was observed. (Ann Dermatol 5:(1) 22-24, 1993)**

*Key Words:* Juvenile xanthogranuloma, Neurofibromatosis.

There are several reports concerning an extracutaneous occurence or a combination of this lesion with other disorders. Jessen et al[1] reported that juvenile xanthogranuloma was a part of the clinical picture of von Recklinghausen's syndrome. Newell et al[2] stated that juvenile xanthogranuloma might be analogous to the pheochromocytoma which occured with increased incidence in neurofibromatosis but also occured independently.

In this report, we describe a patient in whom juvenile xanthogranuloma was associated with neurofibromatosis and retardation of physical growth and development were observed.

## REPORT OF A CASE

A 10-month-old female had multiple yellowish papules on the face and shoulders from 1 month-old (Fig. 1). Variable sized, multiple cafe-au-lait spots were present on the trunk and buttocks from birth (Fig. 2). Her body weight was 7.2Kg (average body weight for this age is 9Kg). She could not sit up, crawl, grasp objects or speak repetitive consonant sounds, mama and papa. She had one healthy sibling. Her family history was negative for xanthomatous skin lesions or neurofibromatosis. The past history was noncontributary. On physical examination, the patient had 2 or 3mm sized multiple yellowish papules on the face and shoulders. Also multiple, more than 6, at least 1.5cm diameter, cafe-au-lait spots were observed on the trunk and buttocks.

Histopathologic findings of the yellowish papules taken from the left shoulder showed poorly demarcated dermal infiltrate consisting of foamy histiocytes, Touton giant cells and various other inflammatory cells against a background of proliferating fibroblasts (Fig. 3).

Routine laboratory studies were within normal limits. The chest roentgenogram and brain CT scan were normal.

## DISCUSSION

Juvenile xanthogranuloma is a benign fibrohistiocytic proliferative disorder and usually believed to be a self healing lesion. It occurs most frequently in children but has occured in adults. Usually the lesions are single, but multiple lesions may be seen in both children and adults[3]. Gianotti and Zina[4] described two forms of the disease in children-micronodular and macronodular. The micronodular form occurs as multiple, 2 to 5 mm, red to yellow, discrete papules scat-

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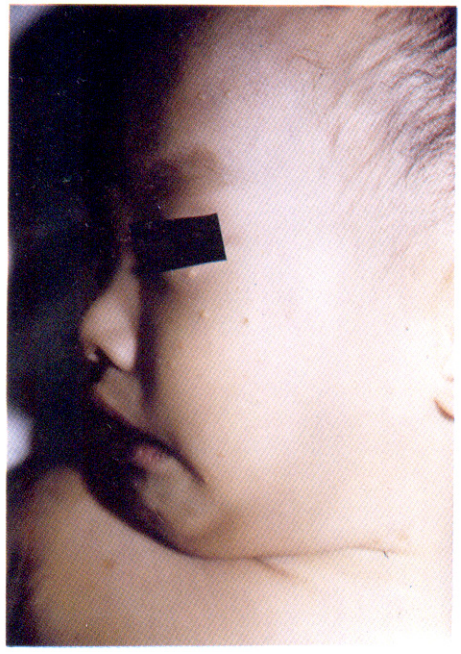

**Fig. 1.** Multiple yellowish papules on the face.

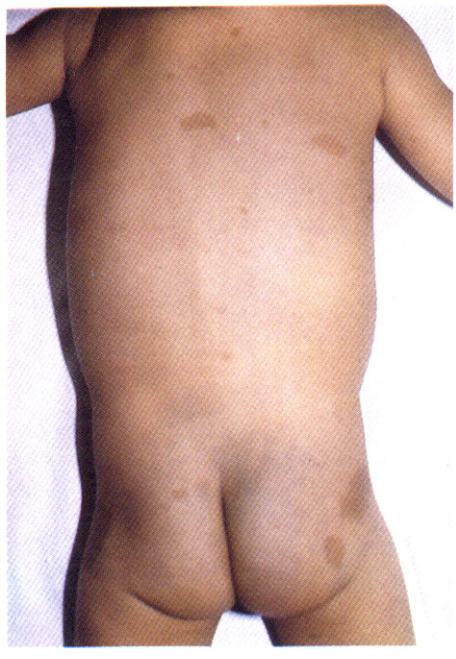

**Fig. 2.** Multiple cafe-au-lait spots on the back and buttocks.

tered on the skin. Neurofibromatosis may be associated with this disorder, and cafe-au-lait spots have been observed in patients and their relatives[2]. Mucosal lesions or lesions on the external portion of the eye may be found. The macronodular form occurs as raised, 1 to 2 cm, red

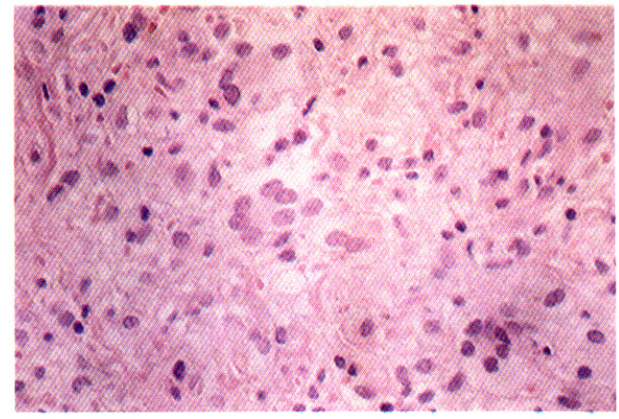

**Fig. 3.** A biopsy specimen from the yellowish papule of the left shoulder shows dermal infiltrate consisting of foamy histiocytes, Touton giant cells and various other inflammatory cells (H & E stain, x200).

or yellow lesions. The macronodular xanthogranuloma is related to the systemic lesions of lung, bone, testis, ovary, colon, kidney, and pericardium[5]. The mucous membranes also may be involved.

There are numerous reports of juvenile xanthogranuloma associated with neurofibromatosis or internal involvement. According to Jensen's report[1], there appears to be a high liability of developing juvenile xanthogranuloma in patients with neurofibromatosis. And in that case neurofibromatosis may be present in partial form only showing cafe-au-lait spots with no family history. In our case there also were no specific symptoms or signs and family history other than cafe-au-lait spots. Weidman and Schaffer[6] reported pontine tumors assocated with cutaneous xanthoma. Because pontine tumors are frequently seen in neurofibromatosis, neurologic examination should be performed in cases of juvenile xanthgranuloma. Nomland[7] and Whittle[8] have described patients with juvenile xanthogranuloma and cafe-au-lait spots. A child with congenital cafe-au-lait macular pigmentation developing cutaneous naevoxanthoendothelioma without a family history of neurofibromatosis has been reported by Pinol et al[9]. Okisaka et al[10] described a boy with cafe-au-lait pigmentation and cutaneous naevoxanthoendothelioma, involving the eye, who also has no family history of neurofibromatosis. These authors concluded that a significant association between naevoxanthoendothelioma

and cafe-au-lait macules was probable.

Flach and Winkelmann[11] reported cases that had progression and no self healing and finally developed a disabling central nervous system syndrome with cerebral space-occupying lesions. Cooper et al[12] reported a case of xanthoma and cafe-au-lait spots in a patient with juvenile chronic myeloid leukemia. The nature of the xanthoma in patients with juvenile chronic myeloid leukemia has not been completely clarified. In this case the lesions were simply termed xanthomas[13] Xanthomas may precede or follow documentation of the leukemia. In the large majority of cases, leukemia is present by the end of the second year of life.

In the Korean literature, several cases of juvenile xanthogranuloma have been reported[14-18]. Jung et al[16] reported a case associated with a coin sized well circumscribed osteolytic lesion on left parietal bone. Rho et al[18] reported a case associated with generalized brownish papular eruptions, 0.5cm sized, multiple cafe-au-lait spots, juvenile chronic myeloid leukemia and family history of neurofibromatosis.

Our case demonstrated the juvenile xanthogranuloma associated multiple, more than 6, at least 1.5cm diameter, cafe-au-lait spots and retardation of physical growth and development. In such a case close observation is considered to be important because there is the possibility of internal organ involvement or an association with other disorders.

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