NAEVOXANTHOENDOTHELIOMA
(Synonym: Juvenile Xanthogranuloma)
(A case report)

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Summary

A case of Naevoxanthoendothelioma (Juvenile Xanthogranuloma) is reported with rare features like late onset of the disease, involvement of liver and diffuse cutaneous lesions including cafe au lait spots and pigmented naevus. Final diagnosis could be achieved only on histopathology report.

Naevoxanthoendothelioma was first described by Adamson in 1905[1] under the name "Congenital Xanthoma Multiplex". McDonagh in 1912[2] named it as Naevoxanthoendothelioma. Helwig and Hackney[3] laid stress on histopathology of the condition and named it as Juvenile Xanthogranuloma. Naevoxanthoendothelioma may occur in association with Von Recklinghausen's disease as reported by Jensen et al[4]. Individual lesions of Naevoxanthoendothelioma may become pigmented. Cafe' au lait spots are present in some of the cases of Naevoxanthoendothelioma as reported [3,5-10]. Naevoxanthoendothelioma usually occurs early in life but cases in adolescence and young adults can rarely occur. Systemic involvement is known in cases of naevoxanthoendothelioma. Most frequently eyes are involved but lesions in soft tissues, skeletal muscles, salivary gland, testis, stomach, periosteum, pericardium and myocardium are known[12]. Pulmonary infiltrations were seen in cases of juvenile xanthogranuloma[13-16]. Hepato-splenomegaly has been reported by Lamb[13], Nodl[14] and Lever[17]. Involvement of eyelids has been reported by Blank et al[18]. In few cases ocular involvement has been present without cutaneous lesions[19]. Plasma cholesterol and lipids are within normal limits. There is no familial predisposition. Spontaneous remissions after 3-4 years are known in this disease.

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Case Report

25 years old female patient was admitted in the skin ward on 23-9-1974 and was later diagnosed as naevoxanthoendothelioma with cafe' au lait pigmentation. History dated back to 3 years when patient developed yellowish papules on the neck. Slowly new papules developed, some coalescing to form nodules. Gradually, the surrounding skin became infiltrated, thick and folded with yellowish orange colour. Itching was absent. Similar lesions appeared on the whole of the trunk and extremities also. Scattered cafe au lait

spots appeared with the passage of time. A pigmented naevus has been present since birth on the right upper chest extending on to the right upper arm (Fig. 1). Patient was pregnant at the onset of the disease and she delivered later a full term normal female baby. There was no history suggestive of involvement of any other system.

Systemic examination revealed enlargement of liver by 4 cm below the costal margin which was smooth, sotf in consistency, and non-tender. Fundus examination revealed no abnormality. Local examination revealed that the skin of neck, eyelids, trunk, axillae and arms was yellowish orange in colour. It was thickened, infiltrated and thrown into prominent folds around neck and the axillae. Many non-tender papules and nodules were present all over the thickened skin. Nodules were non-adherent to the underlying structures. Cafe-au lait spots were seen scattered over the trunk and the extremities. A large pigmented naevus was present on the right upper chest and upper arm.

Investigations

Hb-10 gms% TLC-5500 cmm DLC P 68, L 29, M 1, E 2. Urine–NAD. Stool – NAD. ESR – 5 mm Ist hour Westergren. STS – Negative. Total proteins – 6.2 gms%, Serum Albumin – 2.7 gms% Serum Globulin 3.5 gm.% Alb/Glb ratio 0.7. Serum Cholesterol–115 mgm% Fasting blood sugar-80 mgm%. Blood, urine and stool examination did not reveal any excess of porphyrins. X-Ray skull showed no abnormality. X-Ray chest revealed a patch of pneumonitis at the base of left lung which cleared with antibiotics. Electrocardiograph did not reveal any abnormality. Histopathology of skin showed epidermal atrophy. The dermis was replaced by diffuse histiocytic infiltrate and fibroblastic cells. The histiocytic cells were oval or polygonal having abundant or vacuolated cytoplasm (Fig. 2). Touton giant cells with their characteristic peripherally arranged nuclei, having central foamy cytoplasm, were present.

Discussion

Naevoxanthoendothelioma, an old term, has been replaced by the designation of juvenile xanthogranuloma. It is generally regarded as an independent entity, as a reactive granuloma of unknown aetiology. The aetiology of naevoxanthoendothelioma is still not clear. McDonagh[2] reported this condition to be naevoid proliferation of endothelial cells. Lamb and Lain[13] considered it to be a systemic proliferative condition related to Hand Schuller Christian disease involving mainly the skin but rarely associated with systemic lesions. Thannhauser[20] opined that it was a monosymptomatic form of Hand Schuller Christian disease generally confined to skin and usually with spontaneous involution. Lever[17] suggested that it was a proliferative self healing process involving the histiocyte as in Hand Schuller Christian disease. It is related to this disease because of intracellular accumulation of lipids. Electron microscopic studies conducted by Gonzalez, Crussi and Campbell[21] had revealed the lesions to be composed of macrophages with complex pseudopodia. In mature lesions abundant lysosomal structures containing lipids in vacuoles, unlimited by membrane was seen in the macrophages. Fleishmajer[22] was of the opinion that naevoxanthoendothelioma represented a benign inflammatory granuloma, sometimes accompanied by lipid deposits, and considered this to be a local process rather than a systemic one. Helwig[3] and Nomland[6] considered it to be a reactive granuloma of unknown cause, but quite distinct from systemic reticuloendotheliosis. Webster et al[12] stressed thepathogenesis of naevoxanthoendotheliomaas a benign reactive process consisting of histiocytes with xanthomatisation. Occasionally, systemic lesions could occur in reactive

processes at any site. The frequent association of naevoxanthoendothelioma with neurofibromatosis and cafe au lait pigmented spots favoured it to be a true naevoid condition as thought by McDonagh[2] originally. The present case presented naevoxanthoendothelioma with cafe au lait pigmented spots and a naevus, but the disease manifested itself at a late age. The mother of the patient showed cafe au lait spots which might be forme fruste of neourfibromatosis.

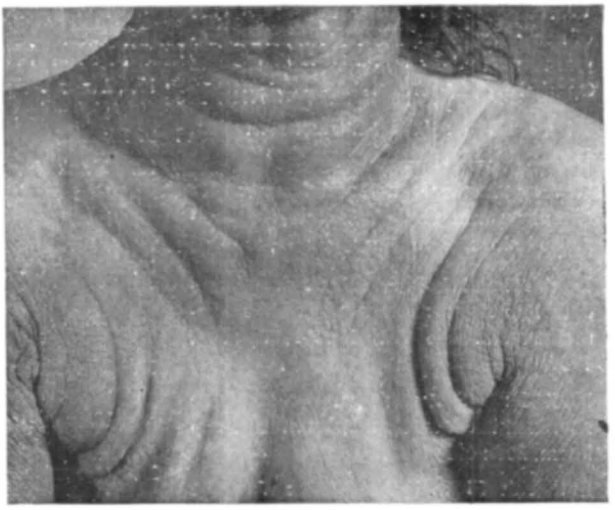

Fig. 1 Shows the cutaneous lesions including the naevus

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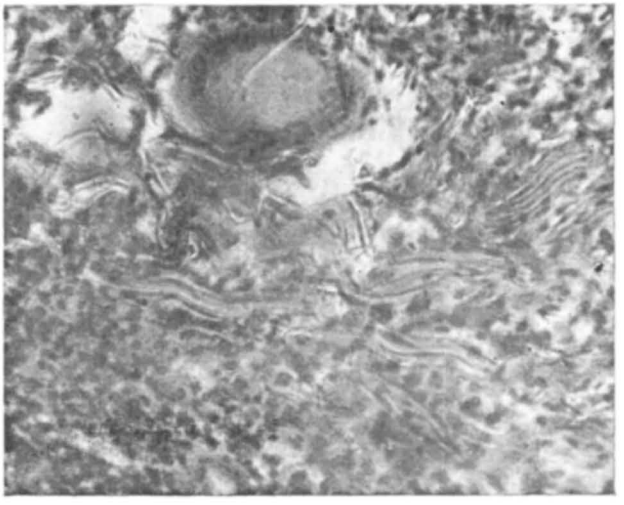

Fig. 2 Shows the diffuse histiocytic infiltrate and fibroblastic cells. A Touton giant ecll is seen in the Dermal infiltrate.

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