PAEDIATRIC DERMATOLOGY

JUVENILE XANTHOGRANULOMA: ABOUT A SERIE OF 13 CASES

S Saad [1] - M Korbi [1] - Y Soua [1] - M Daldoul [1] - H Belhadjali [1] - M Youssef [1] - J Zili [1]

Fattouma Bourguiba Hospital, Department Of Dermatology, Monastir, Tunisia [1]

Introduction: Juvenile xanthogranuloma (JXG) is the most common form of non-langerhans histiocytosis. It is a rare benign condition diagnosed most often at childhood.

Objective: The purpose of this study was to describe the epidemiological and clinical features of JXG through a series of 13 cases.

Materials and methods: It was a retrospective and descriptive study of all cases of JXG diagnosed in our department between 1993 and 2018. Clinical and histological characteristics of the cases were analysed.

Results: Thirteen cases was diagnosed with JXG (10 girls and 3 boys). The mean age at diagnosis was 9.66 months. The clinical presentation was similar in all cases: a well-defined papule or nodule, firm, round or oval. The size ranged from 0.5 to 2 cm. The colour varied from pink to yellowish brown. The lesion was unique in 9 patients (69.23% of cases) and multiple in 3 patients (30.77%). They were located at the face (3 cases), the limbs (3 cases), the scalp (2 cases) and the trunk (1 case). The diagnosis was confirmed in all cases by histology showing a dermal infiltrate composed of histiocytosis, giant cells and a lymphocyte inflammatory infiltrate. Two patients had neurofibromatosis type 1 (NF1) (15.38% of cases). Only one of them progressed to the occurrence of an optic neurilemmoma complicated with blindness.

Conclusion: JXG appears early in life, rarely in adults. It is mainly a unique lesion, but multiple forms are reported. Recently, an association between NF1 and JXG was suggested. So, the clinician should be aware of this possible association especially known the risk of neurilemmoma in this category of patients. JXG was, also, reported to be associated with chronic juvenile myelomonocytic leukemia. Apart from these forms, the prognosis is generally benign with a regression at the age of 3 to 6 years.