Case report

Multiple juvenile xanthogranuloma: A rare case of having clinical appearance mimicking molluscum contagiosum or syringoma

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Abstract

Introduction: Juvenile xanthogranuloma (JXG) is an uncommon benign cutaneous fibrohistiocytic, self healing, class II non-Langerhan’s cell histiocytosis (NLCH). JXG was first described by Adamson in 1905 and defined as a single or multiple cutaneous nodules in infancy as congenital xanthoma multiplex. JXG accounts for 80-90% of cases of NLCH. It occurs most commonly on the head and the neck of infants and young children and resolves spontaneously without treatment. Eighty percent cases appear in the first year of life and 20-30% cases present at birth. Lesions vary in size, but children younger than six months old usually present with multiple lesions with predominance on the head and the neck. In JXG there are two main clinical forms, papular and nodular. However, sometimes in small number cases can be found mixed form, which is characterized by the simultaneous presence of both small and large nodules.

Case: A one year-old boy presented with asymptomatic, multiple yellowish, shiny, and firm nodules with telangiectasia on the surface, measuring around 1 centimeter in diameter, on the face, the trunk, the arms, and lower extremities. The first lesion appeared on the face 8 months prior to consultation then spread gradually to other areas. He was referred from another hospital and was diagnosed as molluscum contagiosum (MC) with differential diagnosis of syringoma. Enucleation had been performed but failed to produce the molluscum bodies. Initial histopathological examination provided the diagnosis of syringoma. The second histopathological examination showed dermal inflammatory cells consisting of eosinophils, lymphocytes, and histiocytes with marked foam cells and giant cells. Eccrine sweat glands were normal. Some lesions decreased in size at subsequent follow-ups; observation was advised until 3-6 years.

Discussion: JXG should be suspected in cases with multiple yellowish nodules appearing in the first year of life. MC usually presents with whitish papules, whereas syringoma is more rarely appeared, presenting with yellow-to-brownish papules. Histopathological examination can easily differentiate the suspected diagnoses; however, selection of lesion, timing and complete clinical information was crucial in reaching the final diagnosis. In this case there was a good clinicopathological correlation that the diagnosis of JXG was made with certainty. There was no eye and other organ abnormalities.

Keyword: Juvenile xanthogranuloma, clinicopathological correlation
Case

A one-year-old boy presented with multiple yellowish lumps with redness on the surface on the face, the trunk, the arms, and the legs. The first lesion appeared on the face 8 months prior to consultation then spread gradually to other areas. There were no other subjective complaints. Patient had been referred from other hospital with the diagnosis of molluscum contagiosum (MC). Enucleation of the lesion was done but failed to produce the molluscum bodies. The initial histopathological result from the referring hospital was syringoma. We re-examined the slides but could not find any abnormality.

On physical examination, there were multiple dome shaped papules and nodules, discrete, mostly 1 cm in diameter on the face, the neck, the trunk, the arms, and the legs (figure 1a, 1b, 2,3).

The nodules were yellowish, shiny, and firm with telangiectasia on the surface (figure 4). A solitary brownish patch at right back of the patient (café au lait lesion) was also noted (figure 5). Eye examination from Ophthalmology Department revealed no ocular involvement.

Figure 1a. Lesions on the face

Figure 1b. Lesions on the trunk

Figure 2. Lesions on the lower extremities

Figure 3. Lesions on the arm
Figure 4. Lesion was yellowish

Figure 5. Café au lait lesion on right back. Shiny and firm with telangiectasia on the surface.

To confirm the diagnosis another biopsy was performed, and the histopathological findings showed dermal inflammatory cells consisting of eosinophils, lymphocytes, and histiocytes with marked foam cells and giant cells (figure 6, 7). Eccrine sweat glands were normal. These features were consistent with xanthogranuloma.

Routine blood laboratory examination did not find any abnormality. No medication was given for this patient. Information about the disease (JXG), the course and the prognosis, also education to parents for long follow up and awareness of systemic involvement were given. From observation around 5 months some lesions decreased in size at subsequent follow-up.

Discussion

Xanthogranuloma should be suspected in cases with multiple yellowish nodules appearing in infant and children, for eighty percent of cases appeared in early life. Molluscum contagiosum can be one of the differential diagnoses. Unlike yellowish nodules found in JXG, molluscum contagiosum usually present with whitish papules and umbilication. Syringoma is clinically small, firm, smooth, skin-colored or slightly yellow papules on the face but rarely appeared in children.

Histopathological examination can easily differentiate the suspected diagnoses; however, selection of lesion, timing and complete clinical information was crucial in reaching the final diagnosis. In this case, a re-biopsy was performed because first histopathological examination showed no specific features. The later histopathological findings were consistent with xanthogranuloma. In this case there was a good clinicopathological correlation that the diagnosis of JXG was made with certainty.

There were no eye and other organ abnormalities in this case. But in few cases, JXG may be correlated with other organ involvement, such as eye, lung, liver, spleen, central nervous system and other, so regular monthly observation is indicated. Patient education about the disease is important.
Patient should understand no medication needed and the lesions will be spontaneously disappeared after 3-6 years.  

Reference


