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CASE REPORT

Juvenile Xanthogranuloma of the External Auditory Canal in an Adult

Dong-Hee Lee, Joohwan Kim

Department of Otolaryngology-Head and Neck Surgery, The Catholic University of Korea, Uijeongbu St. Mary's Hospital, Uijeongbu City, Gyeonggi-Do, Korea.

Juvenile xanthogranuloma (JXG) is the most common non-Langerhans histiocytosis caused by the proliferation of histiocytes. These histiocytes, usually lipid-laden non-Langerhans cells, form multiple yellow-red nodules that are most often found in the skin, the eye, and sometimes in the viscera. It is predominantly a disease of infancy or early childhood, although adults may rarely be affected. In contrast to JXG, adult JXG need excisional procedures when considering its rare spontaneous regression. Herein, we report on an exceptional case of juvenile xanthogranuloma in a 43-year-old man who presented with a sensation of fullness and hearing impairment on his left ear. A peduncular soft tissue mass originated from the posterior portion of left cartilaginous external auditory canal. Under local infiltrative anesthesia, the mass was removed by simple excision. Histologically, the lesion was predominantly composed of a compact mononuclear cellular infiltrate with associated multinucleated giant cells of the Touton type, and these findings were compatible for juvenile xanthogranuloma.

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Juvenile xanthogranuloma (JXG) is a benign, usually asymptomatic, self-healing, red-to-yellowish cutaneous papule or nodule that is composed of histiocytic cells. JXG predominantly occurs in infancy and childhood.[1,2] Since adult JXG was first reported by Gartmann and Tritsch in 1963, more than 30 cases of adult JXG have been reported.Approximately 5% of all cases with adult JXG occur in extracutaneous locations.[3]

The juvenile xanthogranuloma of the external auditory canal was described in only one English literature, which reported on a case of a 13-year-old girl.[4] To the best of our knowledge, this is the first report of a case in which a juvenile xanthogranuloma developed in the external auditory canal in an adult.

Case Report

A 43-year-old male presented with a sensation of fullness and a hearing impairment on his left ear for three months. He denied tinnitus or vertigo. His medical history was unremarkable except for the use of antihypertensive drugs. A physical examination revealed that a soft tissue mass filled up his left ear canal. Its surface looked like a normal skin. While he complained a hearing impairment, pure tone audiogram showed normal hearing in both his ears. The computed tomographic scans of the temporal bone demonstrated that both temporal bones were normal; there was no abnormal finding in the mastoid, middle ear, bony ear canal, and inner ear. The scans showed a partially-enhanced, soft tissue mass that filled the cartilaginous portion of left external auditory canal (Figure 1).

Excision was performed through the transcanal approach under local infiltrative anesthesia. The mass was a peduncular and its base originated from anterior cartilaginous ear canal. Using microscissors, the mass was dissected above the plane of the cartilage of the external auditory canal. After removal of the mass, his tympanic membrane was confirmed to be normal. Because the skin defect was not large and the underlying cartilage was intact, the wound was left to secondary healing. A thin silastic sheet was placed on the wound and Merocel® ear pack (Medtronic, Minneapolis, MN, USA) was packed into the ear canal. The silastic sheet and Merocel® ear pack were removed 1 week later.

A histopathological exam showed a dense infiltrate of many mononuclear cells and several Touton giant cells with finely vacuolated cytoplasm; this was consistent with a juvenile xanthogranuloma. The mononuclear cells

Corresponding address:
Dong-Hee Lee
Department of Otolaryngology-Head and Neck Surgery, The Catholic University of Korea, Uijeongbu St. Mary's Hospital, #65-1 Geumo-Dong, Uijeongbu City, Gyeonggi-Do, 480-717, Korea.
Phone : +82-31-820-3564; Fax : +82-31-847-0038; E-mail : leedh0814@catholic.ac.kr

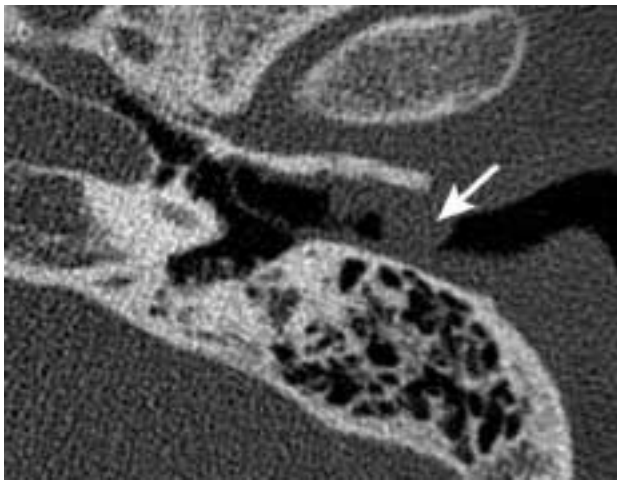

Figure 1. High-resolution CT-scan of the left temporal bone. It shows a soft tissue mass (arrow) in the left external auditory canal.

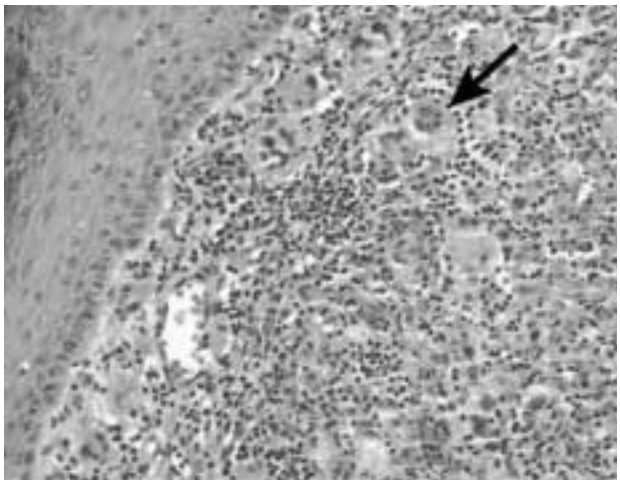

Figure 2. Histopathologic findings (H&E stain, x400). They show a compact mononuclear cellular infiltrate that extends to the basal layer of the epidermis. These cells are characterized by their bean-shaped nuclei. Also seen is multinucleated giant cell of the Touton type (arrow).

were predominantly composed of the vacuolated and oncocytic types of histiocytes (Figure 2).

Discussion

JXG predominantly occurs during infancy or early childhood; approximately 35% of all cases of JXG occur at birth, with as many as 71% of all cases occurring in the first year.[2,3] Despite the term juvenile in the disease name, JXG has been reported in 15% of adult.[3]

JXG of infancy and early childhood is predominant in males and often multiple; the average number is 4.3 per person. The lesion is usually dome-shaped rather than mass-like. It is often tiny, and its size is less than 1 cm. It is known to regress spontaneously within 3-6 years.[2,5] Adult JXG shows no gender difference. It is more often solitary, nodular and larger, and it does not show other associated systemic lesions nor spontaneous regression. However, the clinical features including the color, number or sites of the lesions, and the histologic features are not significantly different from JXG.[2,3,5]

Histological examination of JXG demonstrates a variety of findings. A time-dependent progression exists in the development of the characteristic histological features of JXG, which correlates with the age of the lesion. Early biopsy specimens reveal a dense monomorphous histiocytic infiltrate in the dermis. Older lesions show the classic features. Dehner[1] classified the cellular composition of the cutaneous lesions into three basic cell types: mononuclear histiocytes, multinucleated cells with or without Touton features, and spindle-shaped histiocytes. Because of the transient presence of Touton giant cells in JXG lesions, these classic elements may not be present in every case.[1,2]

For infants or children, anticipatory care with patient reassurance is appropriate because of the self-limiting benign nature of the lesions. Ocular and systemic lesions may respond to steroids or radiotherapy. Both the cutaneous and extracutaneous lesions resolve spontaneously within 3-6 years. However, adult JXG needs excision because of its rare spontaneous regression. Recurrence is not frequent even with a positive resection margin.[2,4,5]

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