

Adult-Onset Juvenile Xanthogranuloma of the External Auditory Canal: A Case Report

성인에서 발생한 외이도의 소아 황색 육아종: 증례 보고

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Juvenile xanthogranuloma (JXG) is a benign, spontaneously regressing lesion that usually occurs during the first year of life, but may also occur in adulthood. Although the most common presentation of JXG is the cutaneous lesion, it can also manifest in various visceral organs. JXG of the external auditory canal is extremely rare, and there have been only a few reports of those cases in the English literature. In this study, we present a case of pathologically proven JXG that occurred in the external auditory canal with a symptomatic clinical presentation.

Index terms

Xanthogranuloma, Juvenile
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INTRODUCTION

Juvenile xanthogranuloma (JXG) is a benign, spontaneously regressing lesion that usually occurs during the first year of life, but may also occur in adulthood. This lesion was first described in 1905 by Adamson (1), who termed the entity, “congenital xanthoma multiplex”. However, the term “juvenile xanthogranuloma” has generally been used following the histological findings, by Helwig and Hackney (2) in 1954, of lipid-laden histiocytes and giant cells in the lesion. JXG commonly presents with asymptomatic clinical features. The most frequently affected sites are the head and neck, followed by the upper torso and extremities (3). In recent years, various locations and clinical manifestations of JXG have been reported. However, there have been only a few reports describing JXG of the external auditory canal and few reports about the radiologic features in the English lit-

erature (4).

We present a case of pathologically proven JXG that occurred in the external auditory canal with a symptomatic clinical presentation.

CASE REPORT

A 30-year-old woman presented to our hospital with otalgia and blood-tinged otorrhea in the left ear, which had been present for two months after she repeatedly picked her ear. The patient had no hearing difficulty, vertigo, tinnitus, or facial weakness. She had no previous history of recurrent otitis media or trauma, and no family history of similar lesions. Otoloscopic examination demonstrated a smooth-margined, round, and pink-to-red colored soft tissue mass that was 5 mm in size in the patient's left external auditory canal (Fig. 1). A tympanic membrane

with a normal appearance was noted by the otoscopy. There was no other skin or mucosal lesions. Computed tomography (CT) scans of the temporal bone were performed with a 16-slice CT scanner (LightSpeed Pro 16, GE Medical Systems, Milwaukee, WI, USA). The CT images showed an ovoid soft tissue nodule, which was 5 × 3 mm in size, in the middle of the external auditory canal and attached to the antero-superior wall (Fig. 2). There was no evidence of infiltration to the adjacent structures. The

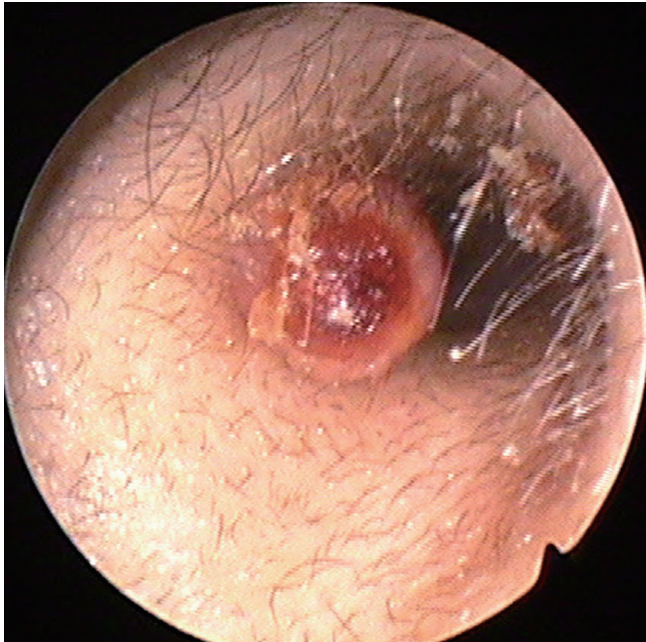


Fig. 1. An otoscopic image is showing a smooth margined, round soft tissue mass 5 mm in size in the left external auditory canal.

results of the serum chemistry tests were within the normal ranges, and the other parts of the body were found to be normal upon physical examination.

A complete mass resection was performed with the patient in the supine position and under local anesthesia; the procedure was performed using the transmeatal approach under the surgical microscope. The mass was observed in the external auditory canal, abutted to the antero-superior wall. The mass was removed using micro-scissors, whereas bleeding was controlled by bipolar cauterization. External auditory canal packing was applied, and the specimen was sent to the Department of Pathology.

The excised specimen was a soft tissue mass measuring 5 × 5 mm in size. The results of histopathological examinations indicated that the mass consisted of foamy histiocytes, Touton giant cells, lymphocytes, and a few eosinophils (Fig. 3). Immunohistochemical analysis revealed that this lesion was positive for CD68 and weakly positive for S-100. These histological findings are consistent with those for JXG. After the surgery, the patient had no complications, such as hearing difficulty or facial weakness.

DISCUSSION

JXG is a benign and rare non-Langerhans cell histiocytosis. A bimodal distribution is observed in the onset of JXG. Although it usually occurs during early childhood, about 15% of the cases involve adult patients, with the peak of onset at 20 to 40 years of

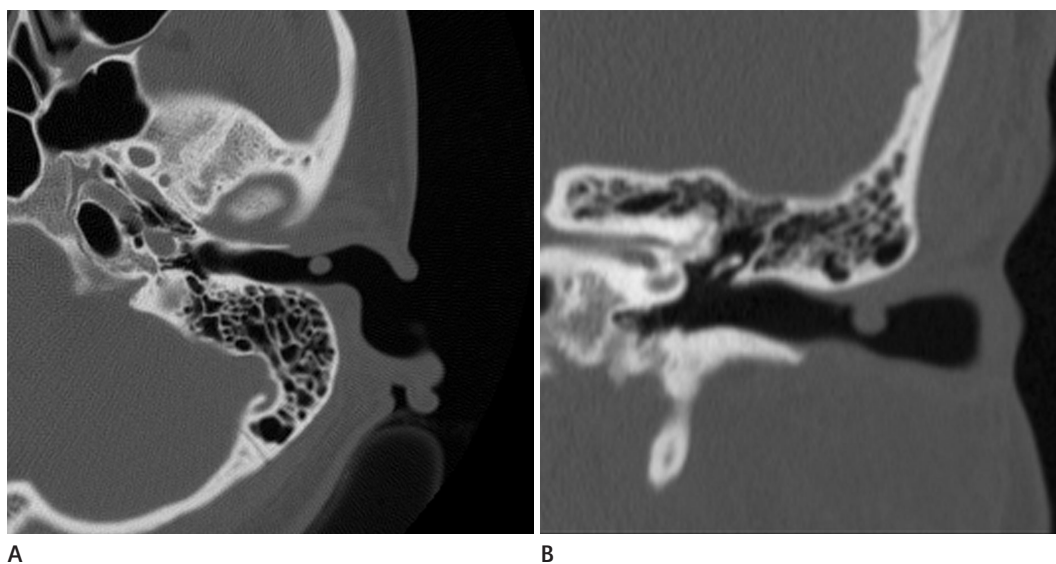


Fig. 2. Pre-contrast axial (A) and coronal (B) images of temporal bone CT demonstrating an ovoid soft tissue nodule 5 × 3 mm in size, in the middle of the left external auditory canal, attached to the antero-superior wall.

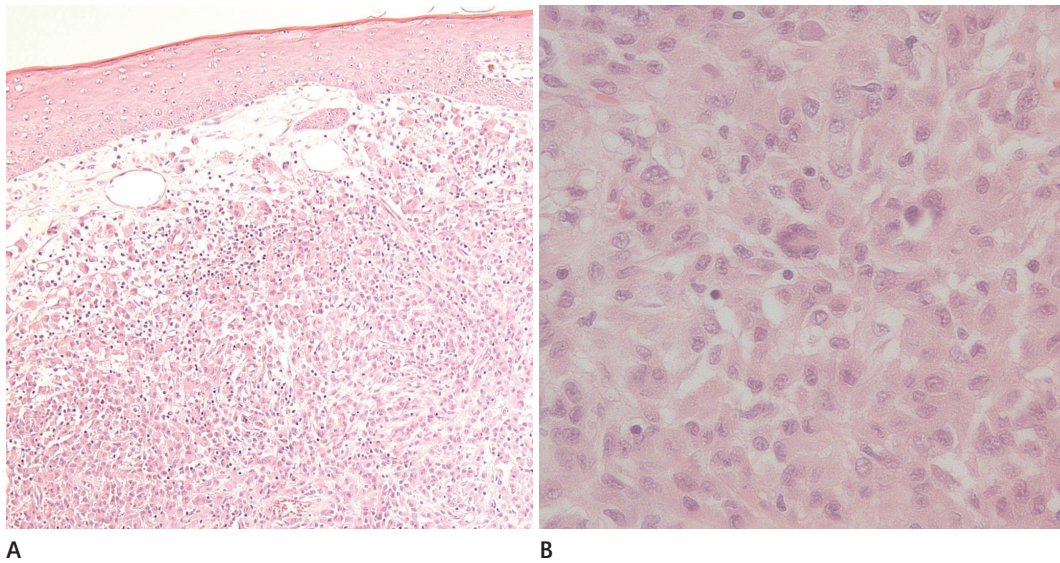


Fig. 3. Microscopic images of the specimen showing foamy histiocytes, Touton giant cells, lymphocytes, and a few eosinophils, consistent with the characteristics of xanthogranuloma; hematoxylin and eosin, $\times 100$ (A) and $\times 400$ (B).

age. Nevertheless, the term “juvenile” is used whether the lesion manifests in adolescents or adults (4). Depending on the age of the patient, the lesion appears different in nature. During early childhood, they tend to occur in multiples and spontaneously regress within one year, whereas the adult form tends to be solitary and persistent (4).

The most common presentation of JXG is the cutaneous lesion, but it can manifest in various visceral organs including the lung, bone, testis, gastrointestinal tract, heart, eye, and oral cavity (5, 6). The head, neck, and trunk are the sites most commonly affected by JXG (3). JXG of the external auditory canal is extremely rare, and it may be difficult to differentiate JXG from other lesions in the external auditory canal (4). Tumor or tumor-like lesions in external auditory canal are exostosis, osteoma, cholesteatoma, squamous cell carcinoma, keratosis obturans, and pleomorphic adenoma. Abnormal osseous proliferation, such as exostosis or osteoma, can be easily seen on CT images. Cholesteatoma can be distinguishable by bony erosion (7, 8) and squamous cell carcinoma is usually presented as invasive soft tissue mass. Keratosis obturans and pleomorphic adenoma may be distinguishable, but usually diagnosis is established on the basis of a histopathologic finding—not on the CT findings—because of their similarity (9, 10). There are few studies about the MRI finding of JXG, and homogeneous iso-signal intensity on T1- and T2-weighted images have been reported (4).

Histopathology is used to diagnose JXG, based mostly on im-

munohistochemical analysis in the clinical setting. JXG demonstrates a well-circumscribed nodule with severe infiltration of histiocytes. The Touton giant cell is observed in 85% of JXG nodules as a multinucleated cell with a peripheral ring of nuclei surrounded by a glassy, eosinophilic cytoplasm. Histiocytes and giant cells are of monocyte/macrophage origin. Therefore, JXG is strongly positive for a macrophage marker, CD68, but negative for the S-100 protein, which is a marker used for the diagnosis of Langerhans cell histiocytosis by immunohistochemistry.

Although the pathogenesis of JXG has not been established, it is believed that reactive processes in response to the initiating stimuli, possibly infectious or physical factors, may play a role in the development of JXG (4). In the current case, the patient had a habit of picking her ear. Therefore, we postulate that JXG may have occurred in response to the repeated ear picking. After complete excision of the lesion, recurrence is uncommon, although it has been reported.

In summary, we report a rare case of solitary JXG of the external auditory canal. The lesion was a well-circumscribed soft tissue mass 5 mm in size that occurred in the supero-anterior wall of the left external auditory canal, and represented adult-onset JXG. When diagnosing benign soft tissue tumors in the external auditory canal, adult-onset JXG should be considered as an alternative condition.

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성인에서 발생한 외이도의 소아 황색 육아종: 증례 보고

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소아 황색 육아종은 양성의 자발성 퇴행 병변으로 대개 1세 미만에서 나타나지만 성인에서도 발생하는 것으로 알려져 있다. 호발 부위로는 피부 병변이 가장 흔하나 다양한 내부 장기에서도 생길 수 있다. 하지만 외이도에 생긴 소아 황색 육아종은 극히 드물며, 이런 증례는 영문 문헌에서 단지 몇 개만이 보고되었다. 따라서 우리는 임상 증상을 보이던 병리학적으로 확인된 외이도에 생긴 소아 황색 육아종을 보고하고자 한다.

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