



Case Report

Isolated Myxoma in the External Auditory Canal of a 10-Year-Old Girl

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Myxoma is a benign connective tissue tumor that is most commonly found in the heart. Because myxoma of the external ear is extremely rare, its diagnosis may be easily delayed or it may be misdiagnosed as another disease. Moreover, because it can be a part of Carney complex (autosomal dominant syndrome), its correct diagnosis is very important. We experienced a 10-year-old girl who had a mass on the posterior surface of the tragus at the entrance of the left ear canal. Fine-needle aspiration revealed mucoid content of the cystic mass, but its cytology did not confirm the diagnosis. The whole mass was surgically removed, and the diagnosis was confirmed as myxoma with a stellate spindle cell proliferation in the hypocellular matrix. Thorough examination failed to determine any presentation of Carney complex, and her final diagnosis was isolated myxoma of the external auditory canal. This is the first reported study regarding myxoma of the external auditory canal in the Korean literature.

KEYWORDS: Myxoma, external auditory canal, children

INTRODUCTION

Among all neoplasms of the head and neck region, benign neoplasms are rare in the external auditory canal. Benign tumors of the ceruminous glands, cylindroma, osteoma, exostosis, and angiolymphoid hyperplasia with eosinophilia can occur in the external ear.

Myxoma is a myxoid tumor of the primitive connective tissue, involving the heart and other soft tissues. The WHO histological classification of tumors of the ear does not include this disease entity; however, some English literatures have reported regarding the cases of myxoma involving the ear^[1-8]. Isolated myxoma rarely involved the external auditory canal and could not be grossly distinguished from other benign neoplasms or cutaneous cysts, such as epidermoid cysts. The exact diagnosis is very important because myxoma of the external auditory canal has been reported as a presentation of Carney complex, which is an autosomal dominant syndrome that is associated with spotty skin pigmentation, endocrinopathy, and endocrine and non-endocrine tumors. Therefore, otolaryngologists should be aware of this disease entity and should be aware that further diagnostic evaluation may be necessary after surgical excision. Although 10 cases of isolated myxoma have been reported in the head and neck area, such as the maxilla, larynx and neck, there has been no report regarding myxoma of the external auditory canal in Korean. We present a case of isolated myxoma in the external auditory canal.

CASE PRESENTATION

Informed consent was obtained from the patient.

A 10-year-old girl was transferred from a primary clinic for prolonged otitis media with effusion. Her medical history was not significant. She got both tympanostomy tube insertion at 7 years old. Otoloscopic examination revealed some effusion in her left middle ear cavity. Approximately half a month later, the middle ear effusion disappeared. Moreover, after approximately 1 month, she re-visited our clinic regarding a flu symptom. On this visit, otoscopic examination revealed some effusion in her left middle ear cavity, and this middle ear effusion improved approximately 1.5 months later.

Fifteen months after the initial visit, she was transferred from a primary clinic for otitis media with effusion persisting in both her ears for a month. Otoloscopic examination revealed amber-colored effusion behind both the tympanic membranes. Right and left otitis media with effusion improved after 1 and 1.5 months, respectively. After 2 months, a small mass-like lesion was incidentally found on the posterior surface of the tragus in her left ear. She stated that this mass appeared to have developed 1-2 months ago. Initial clinical diagnosis was otofuruncle, and an antibiotic-containing ointment was prescribed for half a month. Around this time, she underwent laser therapy for verruca plana on her face and arms. The size of the mass increased despite the antibiotic-containing ointment, and thus, fine-needle aspiration and cytology were performed. Thick, yellow, mucoid fluid was aspirated, and

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cytology was non-diagnostic. There was no specific finding in the remainder of the head and neck examination.

Surgical excision was performed for curative treatment together with a diagnostic intent. Intraoperatively, a 7-mm-sized ovoid mass was noted to be soft and glossy with well-demarcated and smooth borders (Figure 1). It was attached to the posterior surface of the tragus but did not involve the tragal cartilage. Dissection took off the capsule of a mass from the surrounding tissue, enabling gross total extirpation of the mass from the surrounding tissues. Pathological examination revealed bland stellate spindle cell proliferation in the hypocellular matrix, which was consistent with myxoma (Figure 2). Thorough evaluation, including dermatology, endocrinology, and cardiology, did not reveal any finding that was suggestive of Carney complex. The surgical wound uneventfully healed, and there was no recurrence until 1 year after surgery.

DISCUSSION

In 1871, Virchow used the term “myxoma” first for a tumor that histologically resembled the mucinous tissue of the umbilical cord [9]. However, in 1948, Stout established the generally accepted diagnostic



Figure 1. Gross finding of a 7-mm-sized, ovoid, soft, gelatinous mass with well-demarcated and smooth borders

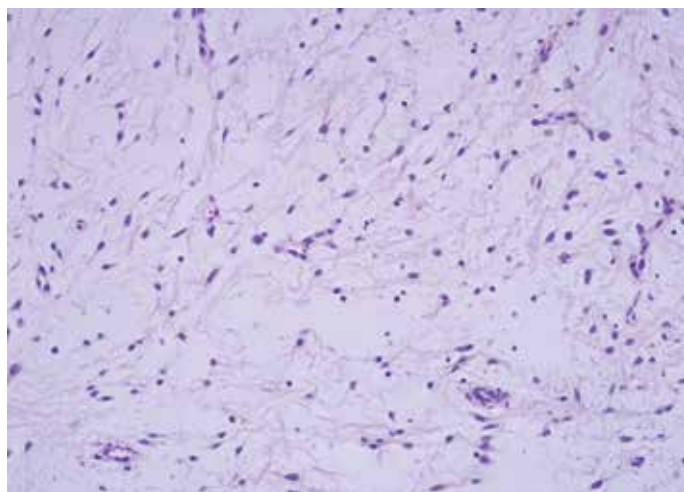


Figure 2. Histological findings at 200× magnification. It shows hypocellular myxoid matrix, scattered bland spindle cells, and sparse vascular connective tissue septa

criteria of myxoma as a true neoplasm that is composed of stellate cells that are set in a loose mucoid stroma through which course very delicate reticulin fibers in various directions [10].

Myxoma most commonly occurs in the heart and can also occur in soft tissues of other locations. In the English literature, the sites of myxoma in the head and neck area have been various, including the maxilla, mandible, and oral cavity [7, 9]. Ten cases of myxoma in the head and neck area, including the maxilla, larynx and neck, have been reported in the Korean literature.

Benign tumors in the external auditory canal are very rare, and among them, myxoma is extremely rare. In the English literature, a few cases of myxoma in the external auditory canal have been reported through a PUBMED search [1-8]. In addition, there has been no report regarding myxoma of the external auditory canal in the Korean literature.

Myxoma of the external auditory canal tends to slowly grow within its capsule without infiltrating into surrounding tissues. Most of it can be completely removed [1, 7, 8]. Moreover, our case was located at the posterior surface of the tragus, did not infiltrate the tragal cartilage, and was completely removed. To prevent recurrence, it is important to excise the entire tumor with a clear margin [7, 9].

For diagnosis, imaging studies, such as CT and MRI, are helpful to differentiate myxoma from other diseases, to evaluate extensive bony erosion or very rare intracranial extension, and to prepare a surgical plan, etc. In imaging studies, myxoma reveals hypo- to isodense with variable enhancement; however, these findings are not pathognomonic because of non-specificity [8]. Definite diagnosis of myxoma depends on the findings of the histopathological examination. Stellate or spindle cells and reticulin fibers are present in the abundant mucoid material with sparse vascularity and are absent or inconspicuous mitoses [10].

When otolaryngologists encounter a case of myxoma of the external auditory canal, it is important to check whether it is a solitary myxoma or a symptom of Carney complex. Sporadic myxoma generally affects middle-aged adults, particularly females. However, the mean age at diagnosis of Carney complex has been reported to be 10–20 years without a preference for gender. For patients diagnosed with myxoma, thorough evaluation is required, including skin pigmentation, endocrinopathy, and endocrine/non-endocrine tumors. Hormone study for thyroid and pituitary and echocardiography are also required.

Carney complex is a syndrome of autosomal dominant trait and comprises myxomas of the heart and skin, hyperpigmentation of the skin, and endocrine overactivity. It is diagnosed on the basis of the clinical diagnostic criteria and may require genetic testing for examining the PRKAR1A gene, the most common mutation [8]. Some cases of myxoma in Carney complex may be located in the cartilaginous wall of the external auditory canal, which is a component of Carney complex. Once myxoma is confirmed, the patient should be considered at risk for accompanying spotty pigmentation, endocrine tumors, or schwannomas and should be accordingly examined to rule out Carney complex. Dangerous conditions, such as cardiac myxomas and

melanotic schwannomas, can be quickly detected and treated in cases in which myxoma is diagnosed ^[7, 8].

Ethics Committee Approval: Ethics committee approval was received for this study from the ethics committee of Uijeongbu St. Mary's Hospital. / UC15ZISE0054.

Informed Consent: Written informed consent was obtained from a patient and her parents who participated in this study.

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