

## Case Report

# Heterotopic Pancreas in Middle Ear: A Case Report

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Heterotopic pancreas is the congenital presence of pancreatic tissue outside its normal location in the absence of vascular and anatomical connection with the main pancreas. To our knowledge, no case of heterotopic pancreas cyst in the middle ear has been reported to date.

In this study, we report the first case of a 6-year-old boy with ectopic pancreas in the area of middle ear. The patient underwent canal wall down mastoidectomy with tympanoplasty. It was finally diagnosed as ectopic pancreas (left middle ear). During the 6-year follow-up, no evidence of recurrence or residual disease in the middle ear cleft or mastoid was found.

Heterotopic pancreas in the middle ear is an uncommon condition and may present with otorrhea or aural fullness. Diagnosis is usually straightforward on the histologic evaluation of resection specimen, complemented with immunohistochemistry. Total excision with such lesion is preferred to avoid some complications. Regular follow-up is necessary due to the potential risk of recurrence and malignant transformation.

**KEYWORDS:** Middle ear, heterotopic pancreas, canal wall down tympanoplasty

## INTRODUCTION

Heterotopic pancreas (HP) is the congenital presence of pancreatic tissue outside its normal location in the absence of vascular and anatomical connection with the main pancreas, which had been described in the previous literature in the following organs: the stomach, duodenum, jejunum, and spleen, rarely found in the ileum, liver, gall bladder, mediastinum, lungs, spleen, fallopian tubes, umbilicus, omentum bile ducts, and mesentery.<sup>1-4</sup> To our knowledge, no case of HP cyst in the middle ear has been reported to date. This study reports the first case of a 6-year-old boy with ectopic pancreas in the area of middle ear.

## CASE PRESENTATION

A boy, 6 years old, was referred to our department due to recurrent otorrhea, conductive hearing loss, and aural fullness on the left side since birth with thin yellow pus, without odor, blood, tinnitus, vertigo, or other related symptoms. The symptoms were relieved after being treated with "unknown antibiotics" for 4 days at the local clinic since birth. The above symptoms occurred repeatedly after a few days, ear pus gradually developed odor, and gradually developed hearing loss. These symptoms occur intermittently for 6 years. Six months ago, the symptoms of stuffy left ear, ear pain, and pus flow with hearing loss appeared again, and the symptoms of pus flow and hearing loss did not improve significantly after "unknown antibiotics" and topical levofloxacin ear drops treatment for 3 times a day, 2 drops at a time at the local hospital. He was referred to our hospital for further diagnosis and treatment on July 22, 2015. The child was in good health. Parents and siblings had no similar diseases. An otoendoscopy evaluation revealed thin yellow pus attached to the wall of the left ear canal and the surface of the tympanic membrane. A lesion occupied the tympanic cavity, and the tympanic membrane was swelling and granulating without pulsating (Figure 1). Pure tone audiometry showed a conductive hearing loss on the left side at all frequencies with a 30 dB gap at least and tympanometry could not be led out (Figure 2). No abnormality was found in the right ear. The rest of the physical exam was unremarkable. It is a pity that for the examination of ectopic pancreatic tissue, only computed tomography (CT) was performed, without magnetic resonance imaging or hormone levels. According to his parents, he is in good health. This is consistent with the view described in the literature that HP usually presented with non-systemic symptoms but local symptoms.

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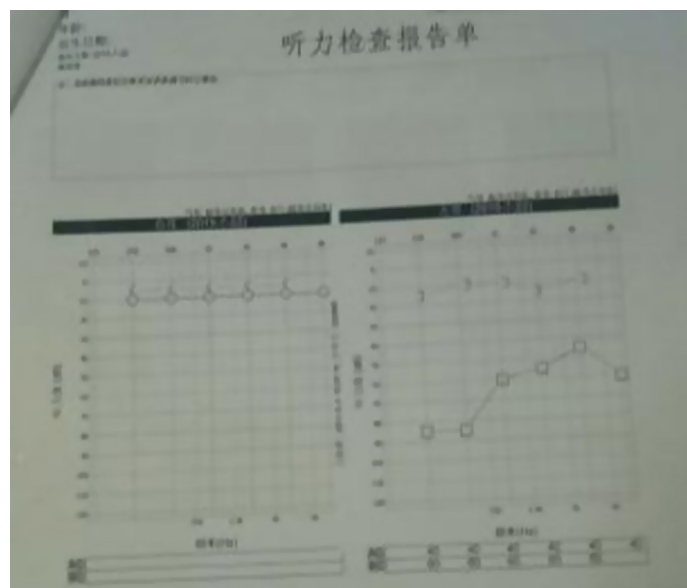
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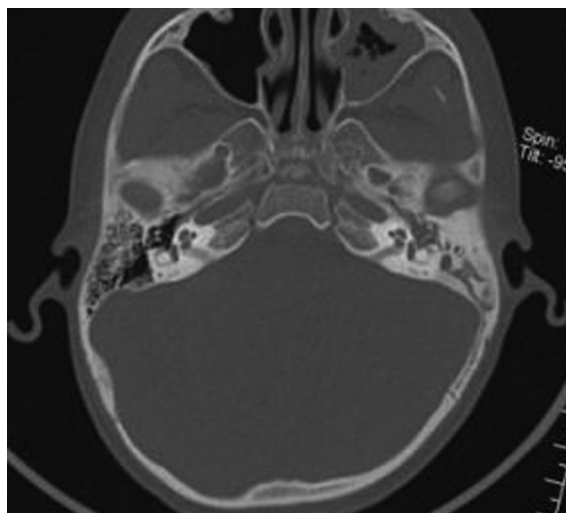
**Figure 1.** Otoendoscopy showed thin yellow pus attached to the wall of the left ear canal and the surface of the tympanic membrane. A lesion occupied the tympanic cavity, and the tympanic membrane was swelling and granulation-like.

The high-resolution CT of the temporal bone revealed an opacification in the mastoid cavity, tympanic cavity, and antrum on the left side without clear evidence of bone erosion (Figure 3). Therefore, he was initially diagnosed with left chronic otitis media.

The patient underwent canal wall down mastoidectomy with tympanoplasty. A retroauricular approach was performed and the lesion was exposed. The mass-like granulation tissues occupied the meso-tympanum and hypotympanum and mastoid cavity, completely involved the ossicular chain, and were extended medial to it, invading the bottom wall of tympanic chamber and the orifice of eustachian tube. Complete eradication of the disease was achieved microscopically after the removal of incus and the head of malleus. The stapes bone was complete, but the stapes footplate was fixed. Perforation



**Figure 2.** Pure tone audiometry showed a conductive hearing loss on the left side at all frequencies with a 30 dB gap at least.

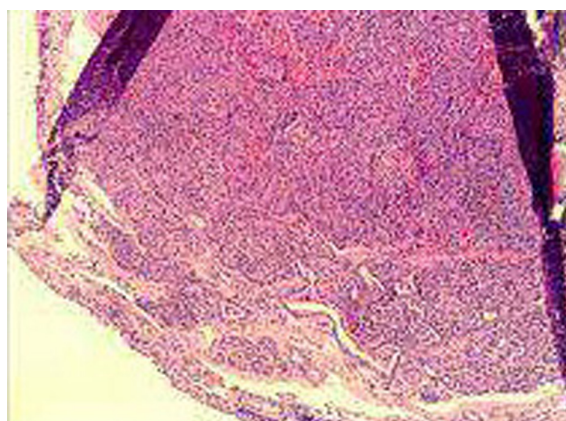


**Figure 3.** CT revealed an opacification in the mastoid cavity, tympanic cavity, and antrum on the left side without clear evidence of bone erosion. CT, computed tomography.

of the tympanic membrane was reconstructed with autologous temporal muscle fascia graft.

Histological examination of the removed tissue showed heterotopic pancreas in the middle ear (Figure 4). The mass was composed of cells with pancreatic tissue organized in multiple foci under the mucosa, and the proliferation of neuroendocrine cells can be seen locally. An immunohistochemical evaluation revealed positivity for CK, EMA, CK8/18, CD56, synaptophysin, and Ki-67(<2%) and negativity for CgA, vimentin, and TTF-1. Morphologic features along with immunohistochemical results were consistent with the diagnosis of ectopic pancreas of the middle ear. It was finally diagnosed as ectopic pancreas (left middle ear) after consultation with Henan Cancer Association (KA150717, August 3, 2015).

Postoperative recovery was uneventful, without signs of complications. He recovered well and remained free of pain and pus since then but without improvement in hearing level on operated ear. Reexamination of otoendoscopy is shown in Figures 5 and 6 and a recheck of the CT of temporal bone is shown in Figure 7. During the 6-year follow-up, no evidence of recurrence or residual disease in middle ear cleft or mastoid was found.



**Figure 4.** The mass was composed of cells with pancreatic tissue organized in multiple foci under the mucosa.

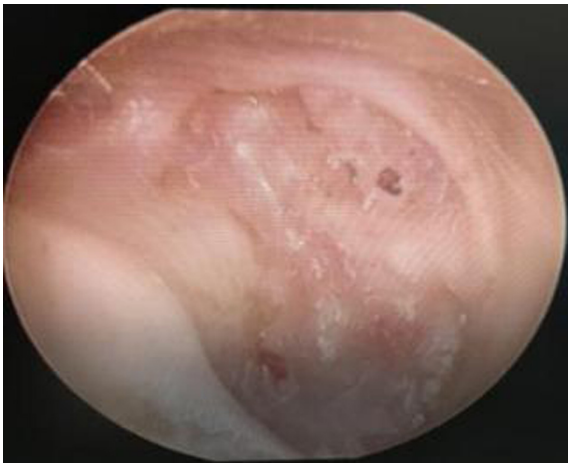


Figure 5. Reexamination of otoendoscopy after surgery.

Consent for publication was obtained from the legal guardian of the patient

### DISCUSSION

Ear discharge is common in children and is usually due to infection, cholesteatoma, and inflammatory granuloma, uncommonly due to middle ear tumor, and extremely rare due to HP. Apart from the case presented here, no report of ear HP has been published as far as we know.

The first case of HP was reported by Hunt et al in 1727,<sup>5</sup> but the first histological confirmation was described by Klob et al in 1859.<sup>6,7</sup> Heterotopic pancreas had been described in the following organs: the stomach, duodenum, jejunum, and spleen, rarely found in the ileum, liver, gall bladder, mediastinum, lungs, spleen, fallopian tubes, umbilicus, omentum bile ducts, and mesentery.<sup>1-4</sup> However, middle ear HP is extremely rare in population.<sup>8</sup> Most of the clinical symptoms caused by HP are covered by the accompanying lesions, but some have obvious clinical manifestations, such as ulcer, bleeding, and obstruction. These differences are related to the location, size, and adjacent organs.<sup>9</sup> In addition, HP can occur at any age, more often at the age of 30-50.<sup>3</sup> Ectopic children are rare, with atypical clinical manifestations, and are easily missed at diagnosis. In this case, the main symptom of the child is ear pus, which is no different from the common manifestations of otitis media. Malignant transformation

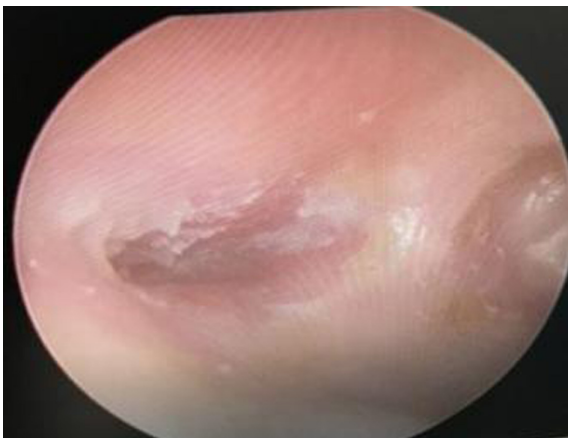


Figure 6. Reexamination of otoendoscopy after surgery.

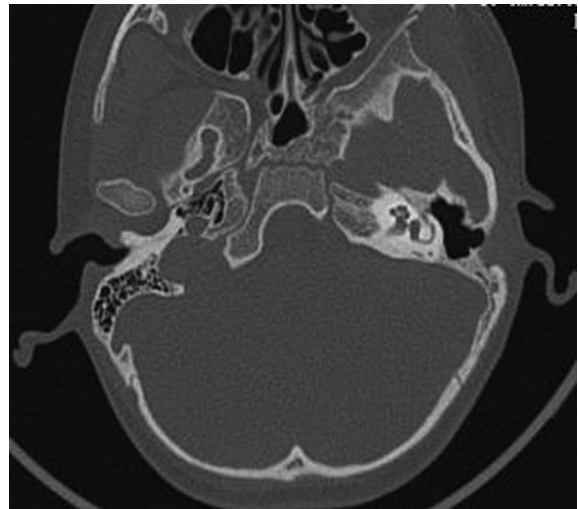


Figure 7. Rechecking the CT after surgery: no evidence of recurrence or residual disease in middle ear cleft or mastoid was found. CT, computed tomography.

of HP tissue is a rare entity with only several case reports published in the scientific literature,<sup>10</sup> most of which are in the upper digestive tract and the majority of the cases are middle-aged people.<sup>11</sup> Classification of the heterotopic tissues was modified by Fuentes in 1973<sup>12</sup>: type I: resembles the normal pancreatic tissue with the presence of ducts, acini, and endocrine islets; type II: canalicular variant with pancreatic ducts; type III or exocrine pancreas only; type IV or endocrine pancreas only. The first and most common type of heterotopic pancreatic tissue is composed of all the elements of the normal pancreas, including acini, ducts, and islet cells.<sup>13</sup> Heterotopic pancreatic tissue in the middle ear conforms to the first type I.

In addition, combined with the diagnosis and treatment process of this case, this study summarizes several experiences, hoping to provide some references for the diagnosis and treatment in the future. First of all, from the perspective of clinical symptoms, this case is mainly characterized by "ear pus," which occurs early in infancy. Therefore, clinically, for such diseases with early symptoms, we should not forget the possibility of rare diseases to avoid missed diagnosis and misdiagnosis. Early detection and treatment should be carried out. For the diagnosis of the disease, there are no characteristic changes in the imaging manifestations such as special physical examination and CT. The intraoperative lesions are mainly like granulation tissue without characteristic tissue such as the epithelioid tissue of cholesteatoma. At this time, we should also consider the possibility of rare diseases. It is hoped that the report of this case of ectopic pancreas in the middle ear will bring clinical attention.

### CONCLUSION

In conclusion, HP at the middle ear is an uncommon condition and may present with otorrhea or aural fullness. Diagnosis is usually straightforward on the histologic evaluation of resection specimen, complemented with immunohistochemistry. Total excision with such lesion is preferred to avoid some complications. Regular follow-up is necessary due to the potential risk of recurrence and malignant transformation.

**Informed Consent:** Written informed consent for publication was obtained from the guardian of the patient.

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