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Case Report

Plexiform schwannomas of cavum conchae in a young girl: report of a rare case

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Abstract: Objective: To report an unusual case describing a plexiform Schwannoma in the ear cavity of a young child. Patient: A 3-year-old girl. Interventions: The tumor was entirely removed by surgery, the pathologic diagnosis of the isolated tissue was performed, and the surgical incision was routinely treated. Main outcome measure: Report the main clinical manifestations and rehabilitation status of patients before, during and after surgery. Results: By surgery, we completely removed the mass. Unlike previous literature reports, we found that the surface of the mass was not encapsulated, and subsequent pathologic reports confirmed that it was indeed a plexiform schwannoma. As of now, the patient did not have adverse reactions or postoperative recurrence. Conclusions: We recommend a pathologic diagnosis of isolated tissues after they are completely removed during surgery. Furthermore, if the same ear has been operated on before, it may increase the difficulty of this operation.

Keywords: Plexiform schwannomas, cavum conchae, small children

Introduction

The incidence of ear tumors is low, accounting for 8.7% of otolaryngology tumors, and the ratio of benign to malignant is 1.83:1 [1]. Schwannomas are common benign tumors in peripheral nerves, which are also known as neurilemmoma and neuronorna. Although it can occur at any age, higher risk of age in the 20 s to 50 s. Gender has no significant effect on the incidence of this disease [2]. Reports on plexiform schwannomas in the ear are rare [3-6], and the majority of patients are reported in adults. A case report on a plexiform schwannoma of cavum conchae in a small child is as follows.

Case report

On September 1, 2019, a 3-year-old girl presented to our hospital with a 1-year history of a recurrent ear neoplasm. Two years ago, her family found a lump in her right ear cavity. The operation, “ear cyst resection”, was performed in another hospital one year ago. According to the postoperative pathology report, that tumor was a cyst. Two weeks later, the girl was claimed to be cured. Six months after the operation, a soft textured, cyst-like new mass was found in the right ear cavity, and it grew faster than the previous one. Moreover, just like before, there was no redness. It completely occluded the external ear canal, and the tympanic membrane could not be seen (**Figure 1**). After a physical examination and laboratory tests, we believe that the girl had no fever, enlargement of the liver, spleen hyperthyroidism and other chronic diseases. On September 3, 2019, the girl who underwent relevant examinations and had no surgical contraindications underwent a right ear neoplastic resection under general anaesthesia.

During the operation, a huge mass was seen in the right ear cavity. There was no redness or swelling on the skin out of the mass. The texture was soft, and there was no clear boundary between the tumor and the normal tissue. We did not see a clear capsule on the surface of the mass. The skin mucosa of the posterior wall of the external auditory canal was slightly raised. We cut along the outer edge of the mass and separated the new mass under the skin. It was seen that there was no obvious boundary

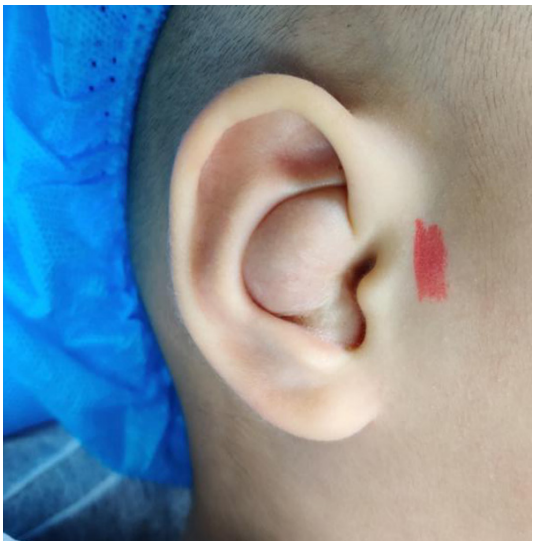

Figure 1. A 3-year-old girl presented with a recurrent ear neoplasm. A cyst-like new mass can be seen in the right ear cavity, about 3 cm sized, soft texture.

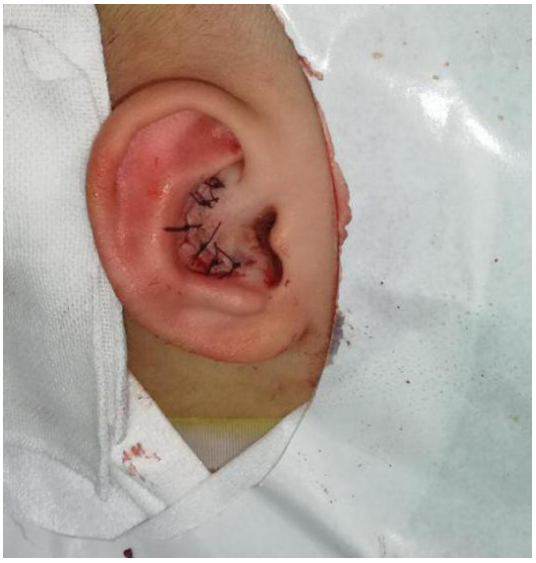

Figure 2. The child's right ear after the operation. The tumor has been completely removed, and the wound has been sutured.

between the new mass and the normal tissue, and some of the cartilage is absorbed. We removed the new organism along the edge of the mass, retained normal cartilage structure, and sutured the incision (**Figure 2**). The external auditory canal was supported by short iodoform gauze wrapped by Vaseline gauze and the incision was pressure-wrapped. The length of the isolated tissue was about 3 cm, and the surface was not covered with a clear capsule

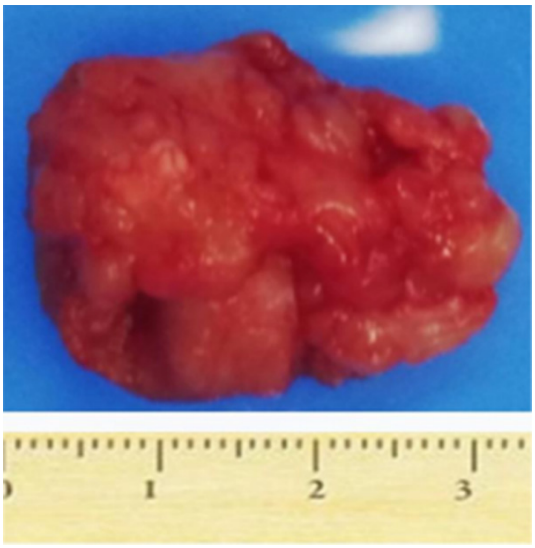

Figure 3. The isolated tissue. Soft texture, about 3 cm long, and was not covered with a clear capsule on the surface.

(**Figure 3**). Histopathologic examination of the specimen revealed a plexiform schwannoma (**Figure 4A**, **4B**). The immunohistochemical findings of the tumor also confirmed this (**Figure 5**). After the operation, the little girl did not have any bad symptoms such as fever, and the surgical incision healed well as we expected. On September 10, 2019, only a few hemorrhagic crusts and exudation could be seen. We removed the stitches (**Figure 6**).

Discussion

The pathogenesis of schwannomas may be related to an inactivating mutation of neurofibromatosis-2 (NF-2) gene [2]. According to the classification criteria of WHO nervous system tumors, schwannomas can be divided into three types: rich cell type, melanin type, and plexus. The plexiform neurinoma, first reported by Masson (1956), is multinodular or plexiform, accounting for approximately 5% of schwannomas. Plexiform schwannomas are more common in the epidermis rather than deep tissues. External auditory canal schwannomas may originate from sensory nerves such as the V, VII, IX, X cranial nerves, auricular nerves, or occipital nerves. Therefore, it is difficult to determine the neural origin of schwannomas in the external auditory canal [4]. As with typical schwannomas, plexiform schwannomas generally have a complete capsule [2]. According to previous literature [3-6], after complete resec-

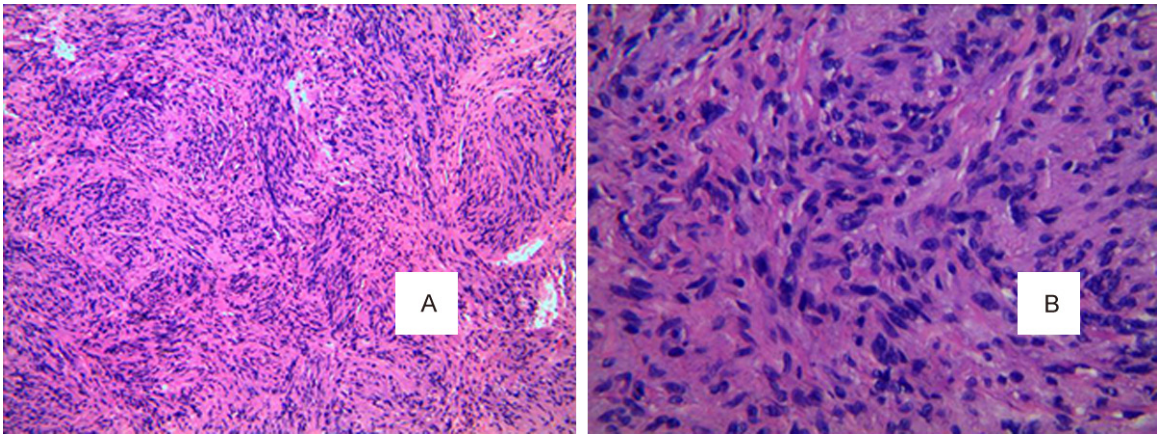

Figure 4. Histopathologic examination of the surgical specimen. A: Round and oval nodules of different sizes and different sizes can be seen under 40 X microscope. The nodules are separated by fibrous connective tissue. B: The tissue under 100 X microscope conformed to a plexiform schwannoma.

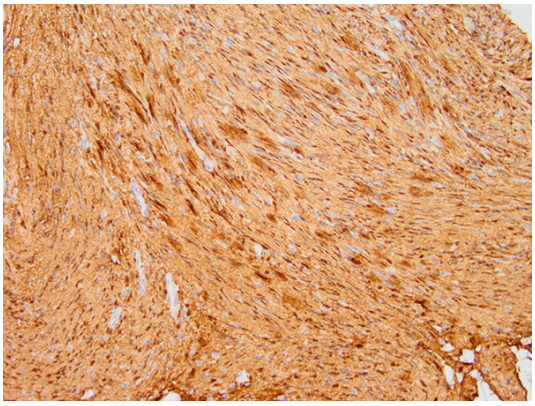

Figure 5. The immunohistochemical finding of the tumor for figures. At 400 X, the specimen is positive for S-100 immunomarker, which suggests that the tumor is a plexiform schwannoma.

tion of the diseased tissue together with the outer capsule, the recurrence rate of this disease is low. In view of the fact that we found no clear capsule during the operation, the risk of disease recurrence was not ruled out. After informing the parents of the child, it was recommended that they should return to the clinic for the test regularly. Six months after the operation, the child recovered well. A little scar formation could be seen at the surgical incision, and no tumor recurrence was observed. Schwannomas are seen on the flexor surface of the limbs most often, and then the head, neck, stomach, posterior mediastinum, and retroperitoneum, and the cranial nerve least [7]. The external ear canal schwannoma is a sporadic benign disease that is easily misdiagnosed as a foreign ear disease such as cysts in clinical

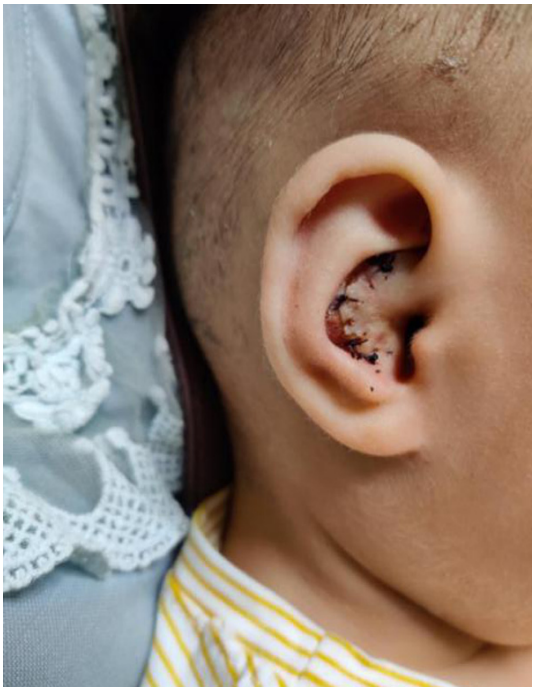

Figure 6. 7 days after the operation. The wound recovered well, only little hemorrhagiccrusts and exudation could be seen.

work. Thus, definitive diagnosis should be based on the results of the histologic and immunohistochemical examination [6]. In this case, the texture of the mass was soft, and no clear capsule could be seen in the operation, which was rare. We propose the following causes: this child had previously undergone an operation, which may cause adhesion of subcutaneous tissue so that no clear capsule could be found during the operation. The following experience

was obtained: because of the rarity of this disease, we have not found any small children in the previous literature. When the patient is a child, the disease can easily be misdiagnosed as other more common diseases. In this case, because the child had a previous history of ear cyst, we were more likely to diagnose the tumor as recurrent cyst. At present, the diagnosis mainly depends on the pathologic findings. Still, a biopsy is not recommended to confirm the diagnosis, so as not to damage the original capsule, causing tissue adhesion and increasing the difficulty operation. Since the tumor is located under the skin, it has progressive and painless growth and has no obvious specific clinical symptoms and imaging findings. Therefore, when the possibility of schwannomas is highly suspected, it is necessary to perform early surgery. The tumor should be removed as completely as possible during surgery to avoid recurrence for patients with intact preoperative mass. After the operation, the specimen should be examined by means of histopathology. Pathologic diagnosis confirmed our diagnosis [8].

Disclosure of conflict of interest

None.

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