

Case Report

The rare case of a mastoid teratoma

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Abstract: Teratomas are quite rare in head and neck sites. We present a 5-year girl with intumescent at the back left auricle and bilateral hearing loss. Both computed tomography (CT) and magnetic resonance imaging (MRI) revealed an abnormal liquid density filling the left mastoid area and the front of petrous pyramid with bony destruction. Radical mastectomy with a microscope was performed. Pathology was consistent with a mature teratoma. Etiology and treatment are discussed and the pertinent medical literature is reviewed. This report presents the very rare case of a mature teratoma of head and neck located in the mastoid of a girl, possibly the 6th case patient documented with this type of lesion in the English literature. Prompt surgical treatment is mandatory and always brings an excellent prognosis. Routine follow-up is necessary.

Keywords: Teratoma, mastoid process, etiology, treatment

Introduction

Etymologically, the word teratoma, originated from a Greek word "teras" which literally means "a monster", highlights the aberrant growth and appearance of these tumors [1]. Teratomas are congenital tumors consisting of tissues elements of tridermal lineage: ectoderm, mesoderm, and endoderm which are derived from pluripotent cells [2]. However in recent decades, this concept has become less stringent with the acceptance of cases that are composed of only bidermal ingredients. Teratomas are classified as being either mature or immature. Mature teratomas are the most common pathological type in children and contain only mature elements, such as skin, teeth, hair, fat, cartilage, bone, and glands [1, 3].

Congenital teratomas are rare tumors, accounting for 3% of all childhood tumors [4] and they are most commonly present in the sacrococcygeal and presacral regions, accounting for 60% of all teratomas [5]. Head and neck locations are extremely rare and represent only 2% of all teratomas, with cervical teratomas being the most common [6, 7]. No sex difference for teratomas in the head and neck region has been observed [8]. To date, nearly 15 cases of

temporal teratomas have been reported in English literatures, as scattered case reports. Of these cases, we could find only five reports, involved in mastoid region [9-13].

Case report

A 5-year-old girl was noted to have an intumescent at the back of left auricle since birth. The girl, who failed to newborn hearing screening, hadn't respond to sound. There was no history of otorrhea and trauma. Her physical examination was negative for evidence of facial nerve paresis, facial asymmetry, or ocular findings. The intumescent at the back of left auricle was growing slowly without any symptom, including pain and anabrosis. Her hearing was not very good, but she had no obvious speech impediment. She saw a doctor in a local hospital before going to school, she had related examinations, the details were not provided, but she was diagnosed as "sensorineural hearing loss, congenital cholesteatoma". With the help of local government, she was sent to our hospital for preparing to accept cochlear implantation surgery. After she arrived at the hospital, additional related examinations were made. A computed tomographic (CT) scan of the temporal bones revealed normal anatomy on the right

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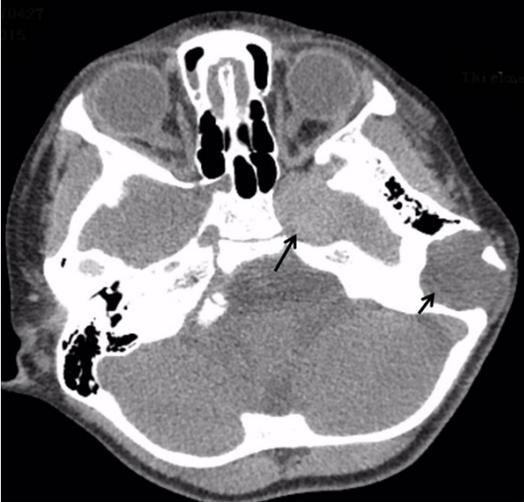


Figure 1. Temporal CT scanning shows an abnormal liquid density filling the left mastoid area following bony destruction (*short arrow*). Moreover, a similar lesion was seen in the front of petrous pyramid (*long arrow*).

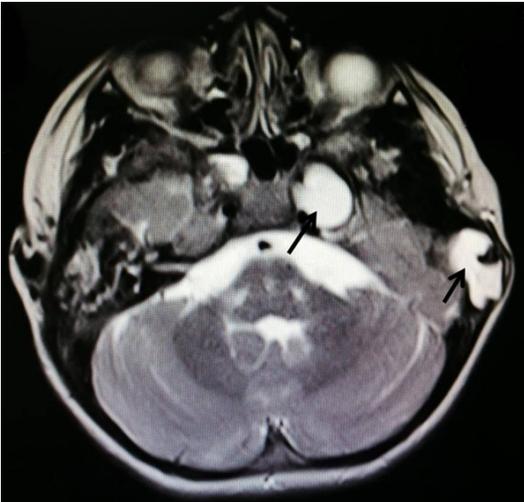


Figure 2. MRI of temporal bone shows irregular cystic masses with homogeneous high signal intensity, which were located in the mastoid area (*short arrow*) and the front of petrous pyramid (*long arrow*), respectively.

but an abnormal liquid density filling the left mastoid area with a CT value of 15H, was similar with the front of petrous pyramid (**Figure 1**). Bony destruction was evident. The structure of ossicles and cochlea were normal. Preoperative magnetic resonance imaging (MRI) of the inner ear scan revealed irregular cystic masses with homogeneous high signal intensity both in T1 and T2 weighted images, located in the mastoid area and the front of petrous pyramid,

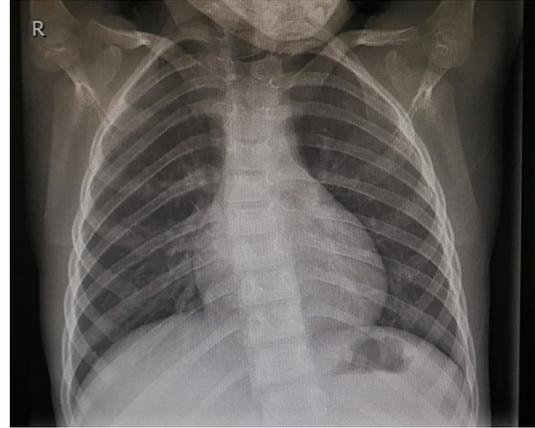


Figure 3. Chest radiography of the patient shows a scoliotic deformation of the upper vertebral column.

respectively (**Figure 2**). Considering the intracranial lesion was not our expertise, neurosurgical specialists were called in to study her case and help us to make a wise decision before surgery. After studying carefully, they thought the mass was very likely the same lesion as the one in mastoid, they suggested that we deal with the mass in the mastoid first. The second incision of the lesion located in front of petrous pyramid would be done later. Surprisingly, conventional chest x-ray of the patient showed a scoliotic deformation of the upper vertebral column (**Figure 3**).

The patient underwent radical mastectomy under the microscope. A retroauricular dermatotomy showed a solid and cystic grey mass arising from mastoid process with the lateral bony destruction. Intraoperative rupture of the cystic portion of the mass, much hair, and white keratoid mass were seen. A radical resection was performed step by step under the microscope, allowing preservation of the facial nerve continuity.

After surgery, the patient didn't show facial paralysis, tinnitus, or dizziness. Her hearing loss was unchanged compared with the preoperative level. Pathology revealed a mature teratoma with disorganized areas of hair, cutaneous adnexal epithelium, fat and fibrous tissue. In addition, epithelium and salivary gland tissue of ectodermal were included (**Figure 4**).

Discussion

We present the very rare case of a mature mastoid teratoma with bony destruction. Only 2% of

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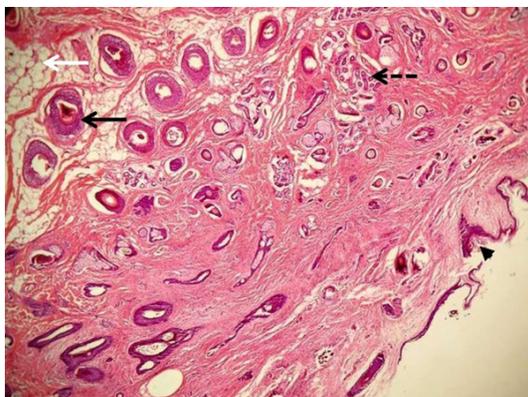


Figure 4. Histology of the tumor ($\times 100$) shows adipose tissue (white arrow), with the skin appendages with hair follicles (solid black arrow), sweat ducts (dashed black arrow), and surface of keratinizing squamous epithelium (arrowhead).

all teratomas occur in head and neck, with an incidence varying from 2.5 to 5/100000 live birth [5]. However, there have been fewer than twenty cases found in the middle ear and mastoid regions. Only a few cases of teratoma have been reported located only in mastoid region.

The etiology of teratomas remains complex and unsettled, multiple theories have been proposed. Some experts have demonstrated that the lesions derive from primordial germ cells. During the 4th and 5th week of gestation, some of these cells miss their target destination and fail to migrate appropriately during their habitual migration from the primitive yolk sac to the gonadal ridges, which induced a teratoma genesis anywhere from the brain to the coccygeal area, typically along the midline [14]. Some other reports showed that the midline cells divide into three germ layers of the embryo at the caudal end during the 3th week of gestation, and the primitive streak normally involutes in the end of week 3. A remnant of embryologic structures can produce a teratoma. The third theory has teratomas as an incomplete twinning [1]. Furthermore, there also is inconclusive evidence of a genetic link for teratomas. Examples include deletions on chromosomes 1, 6, and 12 [1, 15], trisomy 13 [16], ring X-chromosome mosaicism with inactive ring X-chromosome [17], Ring chromosome 21 [18] and so on. In our case, the irregular cystic masses with homogeneous high signal intensity both in T1 and T2, located not only in the mastoid area but also the front of petrous

pyramid (**Figure 2**). The imaging findings of anomaly in the petrous pyramid are extremely similar to the one in mastoid area. The two masses in the same side seemed to give us more inspiration to explore the etiological secret of teratoma.

Teratomas have been found to occur in isolation or in association with other anomalies. These associated comorbidities are urogenital, congenital dislocation of the hip, central nervous system lesions, congenital heart defect, and cleft lip and palate. Teratoma may be also a part of genetic syndromes, such as Proteus and Schinzel-Giedion Syndromes, Beckwith Wiedemann syndrome, Aicardi syndrome, Pierre-Robin syndrome and Klippel-Feil syndrome [1, 19, 20]. In our case, a pre-operative chest X-ray revealed a scoliotic deformation of the upper vertebral column with synostosis (**Figure 3**). We were highly skeptical that the malformation was associated with mastoid teratoma. Wu Xiaobo et al. reported a mature teratoma in temporal bone coexisted with inner ear malformation in a 18 month-year old female patient [21]. In this clinical data, the MRI also showed an abnormal facial nerve pathway. The relation between mastoid teratoma and this malformation is still unclear.

There are varied clinical manifestations in the patients of teratoma of the head and neck. The most common presenting symptom was hearing loss [22]. Other associated anomalies reported are otorrhea [23, 24], fifth nerve palsy, and swelling behind one ear [12, 25]. In our case, the patient's primary symptom was an intumescent at the back of left auricle without any symptoms. Teratomas of the head and neck are relatively rare tumors, which usually cause many diagnostic problems. Ultrasound, CT-scan and MRI are necessary to evaluate the extent of teratoma and help doctors plan surgery. Ultrasound examination of teratomas generally showed mixed echogenicity, with cystic and solid components [14]. Radiologically, the lesions in the middle ear and mastoid were heterogeneous, showed local destruction of petrous bone, displacement of ossicles, and even filling and expanding the eustachian tube [22]. Although uncommon, teratoma needs to be considered in the differential diagnosis of middle ear and mastoid lesions. A cholesteatoma is a growth of keratinizing squamous epithelium, who's clinical and radiological perfor-

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mance are similar. The possibility of a congenital cholesteatoma was included in the initial diagnosis. However, their preoperative differentiation is not critical, because treatment for both entities has been complete surgical excision.

Although sometimes a large incapacitating disfiguring tumor, prompt surgical treatment is mandatory and always brings successful results without complications and with an excellent prognosis. Recurrence may be seen when tumors are incompletely removed. Routine patient follow-up needs to be established to review the final pathology report and to monitor for postoperative complications, healing, recurrence, and transformation, despite the fact that they are often benign tumors.

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Disclosure of conflict of interest

None.

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