Introduction

Turner syndrome was first described by Henry Turner in 1938. It is a chromosomal abnormality where all or a part of one of the X chromosomes is absent or it has other abnormalities. Besides characteristic abnormalities of short stature and infertility, women with Turner syndrome have increased risks for tumors of the central nervous system, especially meningioma and an otologic disease. Meningioma involving the middle ear is extremely rare, and this condition has never been published in association with Turner syndrome. In this paper, we present otological and radiological findings in a patient with TS and discuss necessary diagnostic procedures and possible treatments.

Case Report

A 44-year-old woman with TS was referred to the Clinic of Otorhinolaryngology with intermittent purulent, fetid and sanguineous discharge from the left ear and hearing loss lasting for more than three years. Otomicroscopy revealed a voluminous, soft mass in the left external ear canal (Figure 1). Enterobacter and Klebsiella spp. were isolated. Audiometry showed mixed hearing loss in the right ear and deafness in the left one. There were no neurological symptoms or deficits. High resolution computed tomography (CT) of temporal bones demonstrated hipodense mass (35-49 HU) in the left external ear canal, middle ear, antrum and mastoid air.

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cells without bony destruction. Magnetic resonance imaging (MRI) showed an enhanced infiltration of left middle cranial fossa dura with infiltration of tentorium and mild compression of temporobasal brain with propagation in the left middle ear and the cavernous sinus and surrounding the left internal carotid artery (Figure 2). Angiography of cerebral blood vessels showed normal vascularity and excluded glomus tumors. Imaging characteristics were considered as a meningioma of the skull base with propagation in the left middle and external ear. The biopsy was performed from the mass protruding in the external auditory canal. Pathohistological analysis revealed inflammatory granulation tissue.

This patient with TS had also hypothyroidism, microcytic hypochromic anemia and osteoporosis. Echocardiography showed persistent left vena cava superior and mild mitral and tricuspid regurgitations. During hospitalization the patient was treated with antibiotics for the infection of the middle and external ear. A multidisciplinary team consisting of otorhinolaryngologist, neurosurgeon, cardiologist and endocrinologist decided to repeat radiologic investigations after a year without any surgical intervention because she had no neurological deficits and hearing loss in the left ear was irreversible. It should also be noted that this decision was taken because of the high risk for surgery. A year later, otomicroscopic and MRI findings were the same. Again it was decided to adopt “wait and scan” policy.

Discussion

The authors presented a middle-aged woman with TS having the symptoms of chronic ear disease and unclear otomicroscopic and CT findings of a mass in the left external and middle ear without bone destruction. MRI findings were interpreted as a meningioma of the left middle skull base dura with propagation in the left, middle and external ear.

A middle ear disease, which affects 50-85% of girls and woman with TS, usually starts in early childhood and results in recurrent suppurative otitis media, serous otitis media, chronic suppurative otitis media and cholesteatoma [6]. The frequency of ear infection decreases with age [7]. Middle ear infections and hearing impairment in TS develop due to growth disturbances during development and delayed cell cycle caused by chromosomal aberrations per se and not only due to the specific X chromosome deletion [6]. A middle ear disease in TS is recurrent and leads to multiple surgical procedures [8]. Dhooge et al. [4] advocated careful follow-up during early childhood of girls with TS to prevent sequelae.

In addition, the patients with TS have increased risks for tumors of the CNS, especially for a meningioma and these risks might be associated with genetic and hormonal factors or effects of given hormonal treatments [3]. Ayache et al. [9] reported ten patients presenting with signs of serous otitis media whose imaging revealed a temporal meningioma involving the middle ear. They recommended MRI for all cases with indirect signs of meningioma on CT scan. MRI with contrast typically shows an enhancing, dural-based,
soft tissue mass, while CT shows a hyperostotic bone reaction and a hairy aspect of margins of the affected bone.

Shihada et al. [5] presented three cases of skull base meningiomas mimicking otitis media diagnosed by CT in conjunction with MRI. Histology is not necessary for the diagnosis of meningioma. Meningiomas are mostly benign tumors and the classical approach to meningioma treatment is surgical excision with good prognosis [10]. Fractionated radiotherapy or stereotactic single-dose radiosurgery is also recommended in selected cases. Hormonal therapy with progesterone antagonists and chemotherapy may be considered in cases of unresectable meningioma or where surgery or radiotherapy has failed [11]. Yano et al. [12] suggested conservative treatment with close follow-up to avoid surgery related morbidity in the patients with asymptomatic meningiomas. Ayache et al. [9] reported that conventional middle ear procedures were inefficient in treating a meningioma with otologic manifestations. Shihada et al. [5] suggested “wait and scan” policy in cases of neurologically asymptomatic patients and when treatment modalities carry a significant risk of morbidity.

In the presented case, the initial symptoms were otogenic complications due to chronic otitis media and that was the reason for the hospital treatment and urgent diagnosis. During further examination, radiological findings suggested the skull base meningioma with the middle ear propagation. However, the possibility of surgical treatment was limited because of a high risk for vital function. The multidisciplinary team concluded that there were comorbidities compatible with surgery, but there were no neurological deficits, hearing loss was irreversible and secondary infections of tumor in the external and middle ear was curable with antibiotics. The complete resection of the tumor was impossible without serious injury of neuronal and vascular structures. It has been decided to follow up this woman with TS with yearly scans.

**Conclusion**

The multidisciplinary team approach is essential in patients with Turner syndrome. The first signs of skull base meningioma were presented as chronic otitis media. Proper imaging diagnostics is necessary for the right diagnosis and treatment. Due to a high risk of surgical treatment in the patients with Turner syndrome, wait and scan policy is the most advocated.

**References**