Case Report

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RHABDOMYOSARCOMA OF THE MIDDLE EAR IN AN ADULT

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ABSTRACT

The authors present a case of embryonal rhabdomyosarcoma of the middle ear in an adult patient. Computerized tomography indicated a malignant tumor and the diagnosis was confirmed by pathological analysis.

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INTRODUCTION

Rhabdomyosarcoma is the most common malignant soft-tissue tumor in children.[1] Approximately 50% of all rhabdomyosarcomas in children occur in the head and neck region [2,3]

Middle ear and mastoid involvement accounts for about 8% of head and neck rhabdomyosarcomas [4]

In a series of 91 tumors of the external and middle ear in all age groups, Chen and Dehner[5] found that the rhabdomyosarcoma constituted 7% of the total number of tumors. According to the assessment of these authors there had been only 90 cases of middle ear rhabdomyosarcoma reported in the world literature up to 1978. Nearly all rhabdomyosarcomas of the middle ear and mastoid have been reported in children's age group with the peak incidence between the ages of 2 and 5 years.[9]

The literature mainly contains case reports of rhabdomyosarcoma in the middle ear and mastoid region.[6,7,8]

The purpose of our study is to present rhabdomyosarcoma of the middle ear in an adult patient.

CASE REPORT

The patient, female, age 27, was admitted to a peripheral hospital with otalgia and a discharge from the left ear for the past four years. She also noticed a growth in the left external auditory canal three months before she came for examination. Occasionally she had bleeding from the left ear. Prior to the onset of these symptoms she was well and did not suffer from any ear disease. Throughout this time the patient was treated with medicamentous therapy as an out-patient. Because she was found to have severe headache in the left side, radical trephination of the left temporal bone was done with suspection of chronic otitis media. At surgery, bone destruction was found and granulation tissue with a "polyp" of the external auditory canal were removed. The surgeon did not suspect malignancy and histopathologic analysis was never done. Three months later a revision of the radical trephination was done and granulation tissue was found again. This tissue was removed but the pathohistologic analysis was not requested again. During the first operation a peripheral paralysis of the facial nerve developed. Immediately after operation the patient's general condition deteriorated and she was referred for computerized tomography of the cranium with contrast. These findings indicated destruction of the mastoid process due to invasion of the soft tissue tumor mass. The mass, spreading towards the pyramidal apex, destroyed the cavum tympani and partially the left temporal bone squama but did not involve the pyramidal apex. In the left temporal region, an encapsuled abscess was found. It was intensively colored by the contrast and sur-

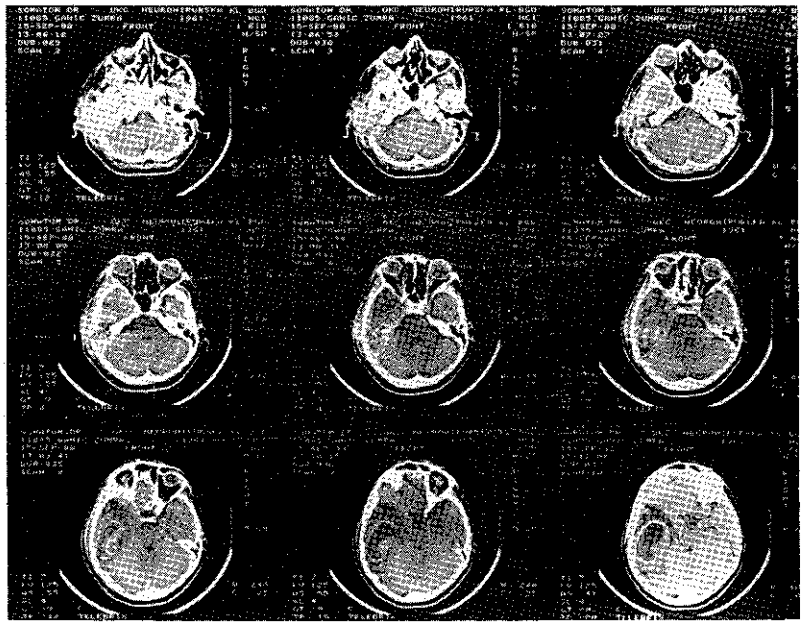

Fig.1. Computed tomography with contrast showing tumor extension towards the left pyramidal apex. Note destruction of the mastoid, cavum tympani and partially the left temporal bone squamae. An encapsuled left temporal lobe abscess is shown. The basal cisterna and the chamber system were completely shifted to the right side.

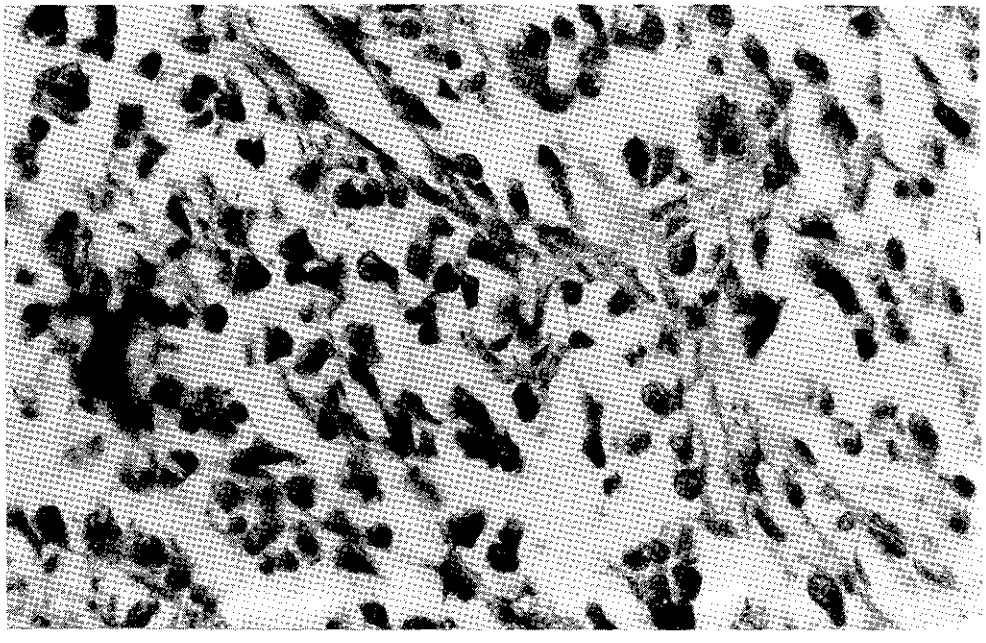

Fig.2. Embyronal rhabdomyosarcoma of the middle ear composed of spindle and giant cells forming irregular stuctures. Note also irregular shape cells in loose fibro-vascular stroma. Malignant cells are in direct contact with small blood vessels (H & E × 350).

rounded by severe edema. The basal cistern and the chamber system were completely moved to the right side (Fig. 1).

After discovering the brain abscess, the patient was immediately referred to the Clinic of Neurosurgery in Belgrade, where temporal left osteoplastic craniotomy was performed. Destruction of the pyramidal roof of the left temporal bone with granulation tissue in the

epidural space were found. Beyond this tissue an abscess of 4 × 5 cm was found in the subcortical region, which was punctured and the abscess capsule completely removed.

The general condition of the patient improved after removal of the brain abscess. However, the patient continued tohave abundant suppuration from the left ear and she was referred to our clinic. Roentgenography indicated that she most probably had a malignant tumor of the left ear. We removed a part of the tumor tissue from the left ear and requested pathohistological analysis. The patient died three days after the operation.

The histopathological result was obtained postmortem and indicated that she had an embryonal rhabdomyosarcoma. Spindle and giant cells forming irregular structures were found predominantly. Beside them, irregular-shaped cells in loose fibro-vascular stroma were also observed. Malignant cells were in direct contact with small blood vessels (Fig.2).

DISCUSSION

We presented the patient with middle ear rhabdomyosarcoma for the following reasons:

- Rhabdomyosarcoma of the middle ear is a rare tumor.

-This malignant tumor, according to the referential data, most commonly occurs in children about the age of 5, and our patient was 27.

- Clinical course had developed very similar to chronic otitis media.

- In our patient an otogenic endocranial complication, that is temporal lobe abscess, had developed.

- The patient was admitted to our clinic in the advanced stage of disease, so that no therapy was indicated. Radiological findings showed a malignant tumor and the diagnosis of rhabdomyosarcoma was confirmed with pathohistological analysis.

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Editorial Comment:

It is interesting to note that aside from the fact that rhabdomyosarcoma of the middle ear is extremely rare, it is usually seen in children, whereas this paper presents an adult case who had been receiving treatment for a chronic ear infection without relief of symptoms.

Therefore in the presence of chronic ear infection even in adults, we must consider the possibility of malignancy, even rare types such as embryonic rhabdomyosarcoma. It is mandatory then that all tissue obtained from surgery of these patients be submitted for histopathological analysis.

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