Tumor of the Temporal Bone Mimicking Acute Mastoiditis

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Abstract

Background: Tumors of the temporal bone in general and particular in children are extremely rare. The anatomical conditions of that special anatomic region can cause misleading symptoms. Diagnosis of rare tumor entities of the skull base in children is a challenging interdisciplinary task.

Objective: The purpose of the present article is to describe the diagnostic process of a rare pathologic condition and review the literature with current opinions of diagnostic and therapeutic methods with respect to angiosarcomas.

Methods: We report about a 13 years old boy with an angiosarcoma of the temporal bone. A review of the literature with particular emphasis on the diagnostic procedures was performed.

Results: The clinical and histological characteristics are reported in here. To date less than 20 cases of angiosarcomas of the temporal bone are published in the literature. Although a large variety of diagnostic methods is available, diagnosing and grading of such a rare tumor is still challenging.

Conclusion: Angiosarcoma of the temporal bone is extremely rare. The symptoms are unspecific, and can mimicking, like in this case, an acute mastoiditis. Diagnosis is challenging and only possible by Immunohistochemical examinations. Diagnosis of such rare pathologic conditions requires an interdisciplinary teamwork and collaboration of different institutions. Current technologies allow an easy exchange of knowledge all around the world for characterizing diseases precisely, and getting the experience of specialists together. This is crucial for a targeted and effective treatment.

INTRODUCTION

Otalgia and hearing loss are common reasons for consulting an ENT physician. In most cases, these symptoms are caused by acute ear infections. Children mostly suffer from otitis media because of anatomical conditions leading to eustachian tube dysfunction [1,2]. Acute mastoiditis is one of the most frequent complication of acute otitis media in children and adults [3]. Classical symptoms are otalgia, fever, posterior auricular swelling, erythema and mastoid tenderness [4,5]. Differential diagnoses include insect bites, severe external otitis, or rare causes like Langerhans histiocytosis and leukemia [6]. In this case report we describe a rare tumor entity, which showed symptoms similar to mastoiditis.

CASE PRESENTATION

A 13-year-old boy was admitted to the ENT department in children’s hospital with right sided otalgia, ear pressure and hearing loss. During initial examination the right retro auricular region was not swollen or hyperemic. However, otomicroscopy revealed middle-ear effusion of the right ear. Therefore the patient received a tympanic drain on the right side and was treated with parenteral cefotaxime. With this the patient’s status improved quickly.

Some weeks later the patient was readmitted with persistent complaints, now presenting a swelling of the right retro auricular region. Furthermore the mastoid was sensitive to percussion. A computed tomography of the temporal bone showed complete mastoid opacification and partial bone destruction (Figure 1). Thus, mastoidectomy was planned. During the surgical procedure the mastoid bone appeared suspicious. A sample of the bone tissue was taken and the operation aborted. A MRI scan of the head revealed skull base destruction by a large mass infiltrating the mastoid bone and the dura. The brain tissue was not affected (Figure 2).
was unimpaired. Preliminary results of histopathology revealed a kaposiform hemangioendothelioma with highly mitotic activity but without signs for invasive growth pattern, and the differential diagnosis of a sarcomatoid tumor.

The patient was hospitalized again because of a bilateral papilledema. Signs of intracranial pressure could be excluded by neurosurgeons. Examinations were completed with bone scintigraphy, computed tomography of the neck, of the thorax and of the abdomen, along with a digital subtraction angiography of the skull base region (Figure 3). Bone scintigraphy showed no further suspicious lesions. CT scan revealed no pathologic findings in the neck, thorax and abdomen. In an angiography of the head a hypervascularized tumor without an early phase venous perfusion was described. It received its main blood supply from the middle meningeal artery.

Pathologists recommended obtaining extended samples of tumor tissue, because of the rare tumor entity. For final histopathologic diagnosis consiliary examination and statements of reference centers were required. In order to take a new sample an operation together with neurosurgeons was performed. During this procedure three different samples were taken using a retro auricular approach: Soft tissue overlying the skull, destructed skull base, and tumor masses lying next to the dura mater. Microscopic examination revealed spindle cell patterns and areas with numerous erythrocytes within in the tumor, furthermore small fields with necrosis and also parts of tumor cells with endothelial growth patterns. Immunohistological findings showed high positivity for endothelial markers (CD31, CD34, ERG), rare positivity for vessel markers, especially factor VIII, signs for high proliferative activity (MiB1) and negativity for HHV8 and S100. The final results of histopathological examination in cooperation with national and international reference centers for endothelial and bone tumors (i.e. Memorial Sloan-Kettering Cancer Center New York, NY, USA; Kiel Pediatric Tumor Registry, Germany; Bone Tumor Reference Center Basel, Switzerland) revealed an highly proliferative malignant endothelial tumor of the skull base with infiltrating the soft tissue. The tumor was finally diagnosed as a high grade angiosarcoma.

Due to the tumor extension the patient first underwent chemotherapy in a local center for pediatric oncology. The regimen included cyclophosphamide, vincristine, adriamycin and paclitaxel. A response evaluation after nine weeks by MRI revealed a significant reduction of tumor mass of about 80%. However, an interdisciplinary tumor board recommended a further conservative treatment with additional radiation because complete surgical resection was not able to perform due to the tumor extension. Furthermore a tumor debulking could not be performed without extensive damage including facial palsy and hearing loss.
**DISCUSSION**

Otalgia and hearing loss are most common symptoms in ENT practice. Using an otomicroscope allows the differentiation between external otitis and acute otitis media, and shows edema and erythema of the tympanic membrane or middle ear effusion. In addition, laboratory examinations detect inflammatory response by elevated leukocyte counts in peripheral blood and increased c-reactive protein [5]. Further examinations are required in cases of a suspected complication. In this case the patient presented with symptoms of an acute otitis media with beginning mastoiditis, which were misleading and did not point to the insidious disease, which was finally diagnosed.

Tumors of the temporal bone and in children are extremely rare. Their incidence varies from 1:5,000 through 1:20,000, whereas most tumors are malignant with carcinomas being the most frequent tumor entity [7,8]. The occurrence of benign tumors of the temporal bone can only be estimated. The incidence is expected as 1:40,000 through 1:200,000 [9]. Carcinomas in this area have an estimated yearly incidence of only 6 in 1 million people [10], and are known to be ten times more frequent than sarcoma. Although it is one of the rarest tumors of the temporal bone, sarcoma occurs more frequently in children than in adults [8]. Angiosarcomas represent an amount of less than 1% of all sarcomas. The percentage of intracranial angiosarcomas is less than 2%. Reported cases involving the skull base were nearly exclusively observed in male patients with an average age of 24 years [11]. The major part of bony angiosarcomas occurs in the long bones of the extremities, the rips, pelvis and vertebra. Primary skull base angiosarcomas are extremely rare, and to best of our knowledge less than 20 cases have been reported in the literature [12,13].

Risk factors for extra-cranial angiosarcomas are prior radiation therapy, arsenic exposure, long-standing lymphedema, and history of previous trauma [14]. However, currently in primary skull base angiosarcoma no risk factors are known [15].

A variety of terms are used to describe this tumor beside angiosarcoma: hemangiendothelioma, hemangiosarcoma, malignant angiendothelioma, and lymphangiosarcoma [14].

Frequent clinical symptoms are otalgia, hearing loss, tinnitus and a swelling in the temporal region, like in the present case [11,13,16,17]. These symptoms are unspecific and can also be found i.e. in acute otitis media or in acute mastoiditis.

Radiographic findings show lytic bone lesions in conventional imaging. CT scans reveal well-defined hyperdense bone lesions with honeycomb configuration. MRI examination demonstrates tumor masses with heterogeneous enhancement. Especially T2-weighted images displays a well-defined heterogeneous mass with surrounding vasogenic edema [18]. In angiography the tumors are highly vascularized [19].

Beside benign and malignant conditions, tumors of intermediate malignant dignity have been reported to occur in the skull base. Preliminary results of histopathology in the case reported here revealed the differential diagnosis of latter entity, namely a kaposiformhemangioendothelioma.

It was Zuckerberg et al., who first defined the term “Kaposiformhemangioendothelioma” in 1993. It occurs nearly exclusively during the childhood and youth, and has features resembling to capillary hemangioma and Kaposi sarcoma, ranging from those of a benign well-differentiated hemangioma to those of an anaplastic angiosarcoma [20]. Kaposiform hemangioendotheliomas are often associated with consumption coagulopathy and thrombocytopenia, the so called Kasabach-Merrit phenomenon [21], which could be excluded in the present case.

In the year 1899 Nadoleczny already reported about the challenging differential diagnosis between endothelial tumors and sarcomas of the temporal bone by means of precisely microscopic investigation [22]. In the present case correct diagnosis is still challenging, although pathologists meanwhile are able to scrutinize surgically obtained material by using several special examinations, i.e. immunohistochemistry and molecular biologic tests. Immunohistochemical markers expressed by cells of endothelial lineage are CD31, CD34 and factor VIII, which are positive in tissue infiltrated by angiosarcoma [13,16].

However, the prognosis of angiosarcoma is generally described as poor, because of high rates of recurrence and metastasis [12,14,23]. To date, due to its rarity, no standardized recommendation for the treatment of intracranial angiosarcomas is available [24]. The best prognosis is associated with complete resection and histologically controlled clear margins. Adjuvant therapy with radiation and chemotherapy are discussed controversially [25]. New therapy concepts are based on monoclonal antibodies, i.e. inhibitors of vascular endothelial growth factor, and are focused to improve the treatment outcome of angiosarcoma [26].

Diagnostical methods of rare pathologies require an interdisciplinary teamwork. Today’s technical possibilities allow an easy exchange of knowledge all around the world to characterize diseases precisely. This is crucial for a targeted and effective treatment.

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**REFERENCES**


