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Prolonged peripheral facial nerve paralysis in a child – think of temporal bone rhabdomyosarcoma: case report

Case Report

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Abstract: Rhabdomyosarcoma is the most common type of soft tissue sarcoma in childhood. When localized in the middle ear and temporal bone, they are highly aggressive, whereas the outcome directly depends on an accurate and timely diagnosis, stage of the disease, and adequate multimodal therapy. Early clinical diagnosis of the temporal bone rhabdomyosarcoma is often difficult since clinical signs are not specific for this disease. We present a case of an embryonal rhabdomyosarcoma, botryoid subtype, of the middle ear in a girl aged 4 years and 4 months, diagnosed 34 days after the first symptoms – right ear pain with peripheral facial nerve palsy on the same side. The overall symptoms were poor, in no way suggesting such a serious condition. After the treatment, control follow-ups for more than 3 years showed no recurrence of the disease, while signs of the right peripheral facial palsy persist. Conclusion: In cases of prolonged peripheral facial nerve palsy in children, not responding to conservative treatment, a temporal bone rhabdomyosarcoma should be considered in a differential diagnosis.

Keywords: *Rhabdomyosarcoma • Temporal bone • Child*

1. Introduction

Rhabdomyosarcoma is the most common soft tissue sarcoma in pediatric population [1]. Approximately 77% of all rhabdomyosarcomas occur in children under the age of 12 years, and 43.5% occur in those under the age of 5 [2]. In 1854, Weber was the first to describe a rhabdomyosarcoma, whereas in 1932, 78 years later, Soderberg reported the first case of rhabdomyosarcoma of the middle ear and mastoid [3]. Rhabdomyosarcomas of the head and neck account for 41% of all rhabdomyosarcomas in childhood, while rhabdomyosarcomas of the temporal bone account for around 8% [4]. Rhabdomyosarcomas of the middle ear and temporal bone are highly aggressive with poor outcome: from 0% of survival during a 2-year period in 40 children with middle ear rhabdomyosarcoma included in several studies [5], to 41% and 81% for advanced and initial stage rhabdomyosarcomas in a 5-year period of follow-up, respectively, undergoing multimodal therapeutic procedures [6,7]. The most commonly described symptoms of the temporal bone and middle ear rhabdomyosarcomas include the clinical picture of chronic otitis media with effusion, polypoid masses in the external auditory canal, otorrhagia, ear pain with or without signs of the same sided peripheral facial nerve palsy, that all delays the diagnosis. This case report describes a middle ear rhabdomyosarcoma with a clinical picture indicating acute purulent mastoiditis [8].

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2. Case report

A female patient aged 4 years and 4 months was admitted to the Ear, Nose and Throat Clinic of the Clinical Center of Vojvodina, Novi Sad with a right peripheral facial nerve palsy. Nineteen days before admittance, the girl experienced pain in the right ear which lasted for 2 days, whereas on the second day the child developed a right peripheral facial nerve palsy. The patient was treated with the diagnosis of acute otitis, receiving peroral antibiotics and a 9-day peroral corticosteroid therapy - a Prednisone 20 mg oral tablets, one tablet daily for three days, 1/2 tablet daily for three days and 1/4 tablet daily for another three days (child`weight was 19 kilograms). After treatment, the patient underwent physical rehabilitation of the peripheral facial nerve palsy. Since therapeutic effects were absent, the child was referred to a tertiary institution. On admission, the patient was in good general condition, afebrile, with a regular pediatric status. Otomicroscopy revealed a slight edema in the pretympanic portion; an upper posterior wall edema; intact eardrum of diffuse pink color, somewhat thickened and with signs of moderate strain in the posterior quadrants, without discernable anatomic details. The otomicroscopy finding in the left ear was normal. On palpation, in the area of tragus, antitragus and bilateral planus mastoideus, no tenderness was noted. Endonasal and oropharyngeal findings were regular. There were clinical signs of right side peripheral facial nerve palsy. Functional audiological assessment revealed a type B tympanometric curve of the right side, and type A on the left side; tonal liminar audiometry showed a right sided conductive hearing loss; bone conduction threshold was 10 dBnHL at 0.5, 1, 2, and 4 kHz with AB gaps at 40 dB frequencies, whereas hearing in the left ear was normal. Shirmer's lacrimation test showed a bilateral >15 mm/5 minutes. Basic blood laboratory test findings were within the reference range (red blood cells count 4.44 ml/microlitre, hemoglobin 12.9 g/dL, hematocrit 36.9%, platelet count 287 000/microlitre, white blood cells count 8 800/microlitre with 41.55 neutrophils and 44.5% lymphocytes in differential white blood cell count, sedimentation rate after one hour was 5 mm).

Native and i.v. contrast CT scans of the temporal bones showed a hypodense mass involving the right tympanic cavity, the antrum and all the mastoid air cells, without signs of erosion or destruction of bony walls. The ossicular chain was preserved, without any morphological alterations. The inner ear structures and the pyramid were without pathological changes. The morphology of the left temporal bone was regular (Figure 1).

On the third day of hospitalization, the child underwent surgical treatment. Intraoperatively, a myringotomy

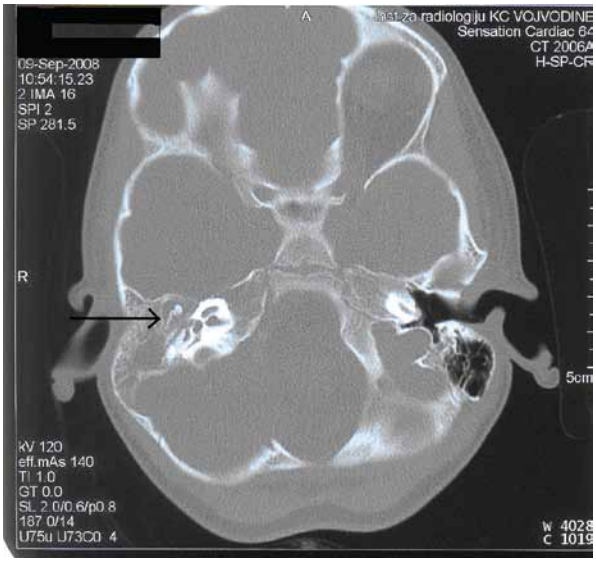

Figure 1. Temporal bone computed tomography (CT) scan of a patient.

was attempted, but at the typical location – lower anterior quadrant, it failed, so it was attempted at the lower posterior quadrant. The paracentesis revealed polypoid masses from the cavum without discharge. A wide mastoidectomy with posterior atticotomy was performed. The complete mastoid was filled with polypoid masses, as well as the aditus ad antrum and the epitympanum. Locally, yellowish mucous discharge was found. There was a slight bleeding from the bone, which appeared soft and osteitic. The dura over the antrum segment of the middle cranial cavity was denuded, 0.8 cm in length. The body of the incus and the head of the malleus were covered by polypoid masses. The tympanic membrane was completely removed due to its changes and thickening, while the posterior bony wall of the external auditory canal was lowered to the level of the Fallopian canal. The medial posterior bony wall was covered with granulations and osteitic lesions. The incus lacking one arch was identified, while the stapes suprastructure was involved with granulation. Incudomalleolar disarticulation was performed, the incus was removed, whereas the stapes was cleaned from granulations. Fallopian canal dehiscence was found in the cochleariform process area. The facial nerve was intact and pale. Carotic canal dehiscence appeared at the floor of the tympanic cavity of the Eustachian tube. Samples for pathohistological analysis were taken intraoperatively from the mastoid and from the typmanic cavity area. Canal wall down tympanoplasty was performed.

In the postoperative period, the child's general condition was good, but clinical signs of right peripheral facial nerve palsy remained unchanged regarding the preoperative status. A 5-day parenteral antibiotic therapy was

initiated using ceftriaxone, while therapy with the II generation cephalosporin was continued. Pathohistological findings revealed an embryonal rhabdomyosarcoma (Figure 2) while further immunohistological staining (tumor was positive for desmin, tropomyosin and myogenic determination factor- MyoD immunohistochemical stains) confirmed the botryoid cell subtype of embryonal rhabdomyosarcoma (Figure 3). After above mentioned pathohistological analysis was done, the child underwent revision surgery under general anesthesia. Fascia, malleus, and remains of the incus were removed, and some tissues from the typmanic Eustachian tube area and from the foramen rotundum were taken for pathohistological analysis. Finally, tamponade of the operative cavity was done. New pathohistological findings showed granulation tissue with individual osteoclast-type giant cells with multiple nuclei and small bony fragments but there were no elements of the previously removed tumor.

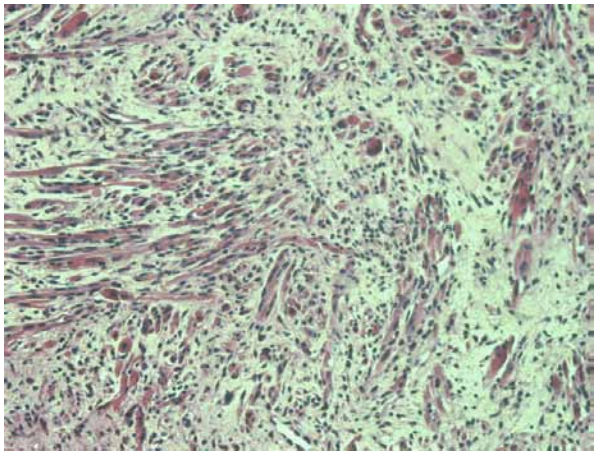

Figure 2. Typical appearance of the tumor tissue (Hematoxylin-eosin x 200).

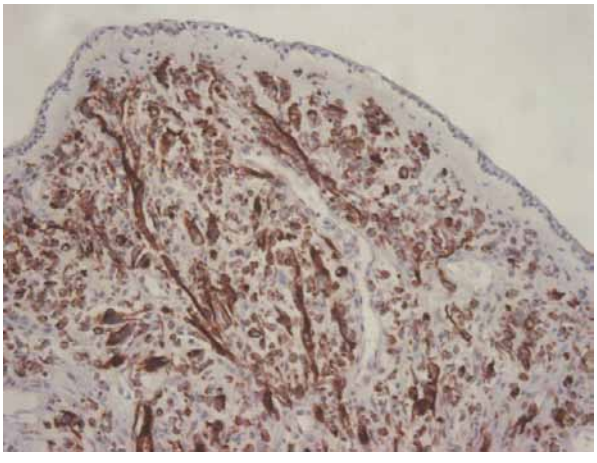

Figure 3. Botryoid subtype of embryonal rhabdomyosarcoma (desmin x 200).

Upon learning the diagnosis, the child's parents decided to continue treatment in Switzerland, where the diagnosis was confirmed, and polychemotherapy and proton radiotherapy were initiated. The child received irinotecan followed by chemo protocol ARSTO531 with VAC (vincristine, dactinomycin, cyclophosphamide) and proton radiation therapy (total dose of 50.4Gy) during six weeks. On check-ups (more than three years after the first surgery), the child is in good general condition, signs of right peripheral facial nerve palsy still persist, while findings in the right operated cavity are normal.

3. Discussion

Rhabdomyosarcoma has a bimodal age distribution: the first peak is between 2 and 5 years of age, and the second peak is between 14 and 18 years of age [9]. It is an extremely aggressive cancer, and over 50% of soft tissue sarcomas in pediatric population are rhabdomyosarcomas [10]. Four histologic subtypes of rhabdomyosarcomas have been described: embryonal, alveolar, botryoid, and pleomorphic [11]. The embryonal subtype is most common in cases of the head and neck rhabdomyosarcomas. In our case report, the child presented with a middle ear rhabdomyosarcoma, while pathohistological findings revealed an embryonal subtype.

The etiology of rhabdomyosarcomas is so far unknown. Most cases of rhabdomyosarcomas occur sporadically, without recognized predisposing factors or risk factors [12]. In the head and neck rhabdomyosarcomas, the most common sites are orbital, parameningeal and nonparameningeal. Orbital rhabdomyosarcomas have a good prognosis after chemo- and radiotherapy, whereas excisional biopsy is appropriate if surgical removal can be achieved [13]. Parameningeal rhabdomyosarcomas involve the following sites: nasopharynx, nasal cavity, paranasal sinuses, infratemporal, pterigopalatine space and the middle ear. Tumors of this localization are inadequate for complete surgical resection and are usually classified as group III tumors.

Early clinical diagnosis of temporal bone rhabdomyosarcoma is often difficult. The most common clinical symptoms include chronic otitis with polypoid masses within the external auditory canal, with or without signs of same-sided peripheral facial nerve palsy, otorrhea and otalgia. According to Pratt and Gray, who analyzed symptoms of rhabdomyosarcoma of the middle ear in 50 patients, 54% presented with polypoid masses in the external auditory canal, 40% with aural secretion, 30% with otorrhagia, 22% with otalgia, whereas 14% of patients had facial nerve palsy [14]. In a retrospective analysis of rhabdomyosarcomas of the head and neck in pediatric

patients during a 35-year period, Sbeity and associates found 39 cases of rhabdomyosarcomas, while only 6 cases accounted for temporal bone rhabdomyosarcomas. Out of the total number, only 4 cases (66%) manifested with signs of chronic otitis media not responding to drug therapy, while there were 2 cases (33%) with otalgia and 2 cases with retroauricular masses, one patient (16.6%) presented with polypoid masses in the external auditory canal, and one with peripheral facial nerve palsy. In the same study, in 2 cases (33%) with temporal bone rhabdomyosarcoma, there were clinical symptoms of multiple cranial nerve palsies involving cranial nerves V, VII, IX, XI and XII, as a consequence of intracerebral extension. Regarding their location, in one patient it was an isolated location in the mastoid, in one in the middle ear, and in one patient petrous bone was involved without intracerebral propagation [1]. In our case report the tumor was located in the middle ear cavities, with clinical manifestations of otitis media without effusion, but with signs of peripheral facial nerve palsy.

Studies of temporal bone rhabdomyosarcomas show that they are most common in children 3-5 years of age, they are more common in boys, and mostly involve the right ear [1,2,4]. In our case it was a girl aged 4 years and 4 months, with right middle ear involvement.

Parameningeal rhabdomyosarcomas are highly aggressive in regard to other head and neck sites, probably due to propagation to the meninges and brain, that makes their prognosis rather poor.

The mean time from the onset of symptoms to establishing the diagnosis of the middle ear rhabdomyosarcoma is 21 weeks [6]. In our case the first symptom was a 2-day pain in the right ear. On the second day, signs of same-sided peripheral facial nerve palsy developed. They lasted 17 days, without any response to drug therapy, so the child was referred to the Ear, Nose and Throat Clinic of the Clinical Center of Vojvodina. During the first two days of hospitalization, morphofunctional diagnostics was conducted; on the third day the child underwent surgery - wall down tympanoplasty; pathohistological diagnosis of embryonal rhabdomyosarcoma was made on the 12th day, so 34 days passes from the initial symptoms of right sided otalgia to the final diagnosis. Further diagnosis, including magnetic resonance of the endocranium, thoracic CT, bone marrow analysis, lumbar puncture, as well as therapeutic procedures of polychemotherapy and protonradiotherapy, were carried out in the country of residence – Switzerland.

Three years after the diagnosis was established and multimodal therapy conducted, today aged 7, the child is in good general health, without local relapse and with normal findings on follow-ups.

4. Conclusion

In children with signs of peripheral facial nerve palsy, complete, serious diagnostics and clinical follow-ups are necessary, especially if symptoms are prolonged and do not respond to conservative therapy. In many cases of peripheral facial nerve palsy, the cause is a malignant disease of the middle ear. Parameningeal rhabdomyosarcomas, including middle ear rhabdomyosarcomas, are significantly more aggressive compared with other sites of the head and neck, due to possible propagation to the meninges and brain, that makes their prognosis rather poor. The main therapeutic goal is locoregional disease control and prevention or treatment of systemic metastases, depending on the stage of the disease, which is directly associated with the tumor location, time between the onset of first symptoms and the diagnosis, as well as on the subtype of rhabdomyosarcoma.

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