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Head and neck rhabdomyosarcoma in a 4 year old girl – case report

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ABSTRACT

Introduction: Rhabdomyosarcoma is the most common soft tissue neoplasm in the pediatric population. Its diagnosis may prove to be a dauting task due to non-specific symptoms it tends to produce, which is explainable by the fact that it may arise in any part of the body. Both histology and the place of origin of the tumour significantly affect the prognosis.

Case report: A four year old girl with a history of non-painful, heamopurulent ear discharge, which was initially diagnosed as chronic granulomatous otitis media presented at the Department of Pediatric Otolaryngology upon the recurrence of these symptoms. The biopsy results showed an embryonal rhabdomyosarcoma-like infiltration and the patient was referred to the Department of Pediatric Hematooncology at the Children's Hospital. The patient underwent a resection that had been preceded by a course of neoadjuvant chemotherapy. Following the surgery, a course of chemoradiotherapy was implemented. Currently the patient remains in remission.

Conclusions: An early diagnosis is crucial in any neoplastic disease and rhabdomyosarcoma is no exception. Such diagnosis should be entertained in any instance of otolaryngologic symptoms that are refractory to standard treatment.

Key words: rhabdomyosarcoma. childhood neoplasm, otorrhea

INTRODUCTION

Among childhood soft tissue neoplasms, rhabdomyosarcoma (RMS) is the most common diagnosis, comprising 50 % of all pediatric soft tissue tumours [1]. Around 50 % of all rhabdomyosarcoma cases occur in children below the age of 10 [3]. It can arise in every part of the body and histologically resembles striated muscle cells. Head and neck (including orbit and parimeningeal location), extremities and genitourinary tract are the most common primary sites [2], with head and neck masses accounting for around 34% of cases [3]. A number of risk factors for developing RMS, such as prenatal exposure to X-rays [4], has been proposed; other factors, such as immunization with vaccines [5] and atopic diseases [5] seem to confer some degree of protection. RMS can also be associated with various genetic syndromes, like neurofibromatosis 1 [6] and Li-Fraumeni syndrome [7]. A genetic testing may thus be warranted [8]. The multitude of possible primary sites which typically entails a certain nonspecificity of symptoms can make the diagnostic process challenging.

Pediatric head and neck tumours represent around 12 % of all childhood malignancies, with rhabdomyosarcoma accounting for 8 % of the cases [9]. It needs differentiation from other malignancies that may arise in the head and neck region in children, such as lymphomas, tumours of the brain, thyroid cancer, retinoblastoma and other, non-neoplastic masses[9].

While head and neck RMS is associated with comparatively better prognosis than RMS originating from other anatomical sites, parameningeal localization has a worse treatment outcomes than other primary sites in the head and neck region [3,11] Histopathological diagnosis of the embryonal type is associated with better survival rates [3,11]. This also holds true if head and neck RMS survival rates are analyzed separately [10]. Patients between the ages of 1 to 10 show better treatment results [3,11,12].

CASE REPORT

A four year old girl who had been experiencing foul-smelling heamopurulent ear discharges since August 2016, was given a diagnosis of chronic granulomatous otitis media. In September 2016 antromastoidectomy and drainage of the right ear were performed. The results of the biopsy showed a non-specific granuloma tissue. After one month the discharge recurred and the regrowth of the granuloma tissue was observed. In December 2016 the patient underwent a second surgery at the Department of Pediatric Otolaryngology of the Children's Hospital in Lublin. The biopsy results showed an embryonal rhabdomyosarcoma-like infiltration and the patient was referred to the Department of Pediatric Hematooncology at the Children's Hospital in Lublin. The patient was classifed as standard risk (SR), subgroup D case according to the CWS 2006 (*Cooperative* Weichteilsarkom Studiengruppe). In February 2017 a 10-weeks neoadjuvant chemotherapy was

started. In April 2017 a resection of the tumour was performed. The missing tissue was replaced by adipose tissue harnessed from the abdomen region. Following the surgery an autologous adipose tissue transfer was perfromed as a means of improving aesthetic results of the resection. The patient subsequently underwent a course of chemoradiotherapy completed in August 2018. During the last visit at the clinic the patient reported no symptoms and magnetic resonanse imagining (MRI) with contrast showed complete remission. The patient has been in remission for nearly 10 months.

DISCUSSION

The middle ear is a rare primary site of head and neck rhabdomyosarcoma, accounting for less than 4 % of all cases [13]. Other malignancies that arise in the temporal bone in children include other sarcomas, lymphomas, carcinomas and infiltrating parotid gland tumours [14]. Malignancies of temporal bone typically resemble ear infections that are refractory to standard treatment [14,15]. Presenting complaints include hearing loss, otorrhea, otalgia, nerve palsies, aural polyps and or vision blurring [14,15,16]

Different treatment algorithms exist for RMS, such as Soft Tissue Sarcoma Committee of the Children's Oncology Group (COG) alogrithm or CWS 2006 (*Cooperative* Weichteilsarkom Studiengruppe). CWS 2006 staging is based on histology, primary site, tumour size, nodal involvement, age and International Rhabdomyosarcoma Study Group (IRS) Classification , thus classifying patients into three risk groups, with standard risk group having three subgroups, B, C and D. Standard risk group, subgroup D, the classification of which our patient received, is reserved for patients with IRS groups II or III (with microscopic and gross residual disease, respectively), whose tumours are located in sites classifed as unfavourable but both their age and tumour size is favourable[18].IRS classification is based on the resectability of the tumour and comprises four groups[19]. EPSSG protocol recommends adjuvant chemotherapy consisting of Ifosfamide + Vincristine + Actinomycin D (IVA) in 9 blocks for SR group, with ifosfamide being withheld after the completion of 4th block in patients with the assignment of groups B or C. Induction chemotherapy may be warranted in some cases. Radiation therapy isn't recommended for patients with completely excised tomour (group B). The remaining patients in SR group should undergo radiotherapy at doses ranging from 36 Gy to 50,4 Gy [17]

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In conclusion, non-specific symptoms affecting any part of the body that prove refractory to treatment and elude diagnosis should be thoroughly investigated as they may be caused by a neoplasmic growth. The treatment for RMS has improved significantly in recent decades, and that, combined with an early diagnosis, ensures good prognosis for most patients.

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