External Ear Alveolar Subtype Rhabdomyosarcoma: A Rare Case Report

Dış Kulak Alveolar Alt Tip Rabdomyosarkom: Nadir Bir Olgu Sunumu

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ABSTRACT

External ear canal (EAC) childhood rhabdomyosarcoma is rare and aggressive due to their potential intracranial spread and meningeal involvement. Given its nonspecific presentation such as bloody otorrhoea,otalgia and aural polyps,it is commonly misdiagnosed as chronic otitis media ,thereby delaying diagnosis. Here we report a rare occurrence of EAC alveolar subtype rhabdomyosarcoma in a 7-year-old boy.

Keywords: Alveolar,Rhabdomyosarcoma ,External ear canal

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ÖZET

Dış kulak yolu (EAC) çocukluk çağı rabdomiyosarkomu, potansiyel intrakraniyal yayılımı ve meningeal tutulumu nedeniyle nadir ve agresiftir. Kanlı kulak akıntısı, kulak ağrısı ve işitsel polipler gibi nonspesifik prezentasyonları göz önüne alındığında, sıklıkla kronik otitis media olarak yanlış teşhis edilir ve bu nedenle tanıyı geciktirir. Burada 7 yaşında bir erkek çocukta nadir görülen bir EAC alveolar alt tipi rabdomyosarkom bildiriyoruz.

Anahtar Sözcükler: Alveolar, Rabdomyosarkom, Dış kulak kanalı

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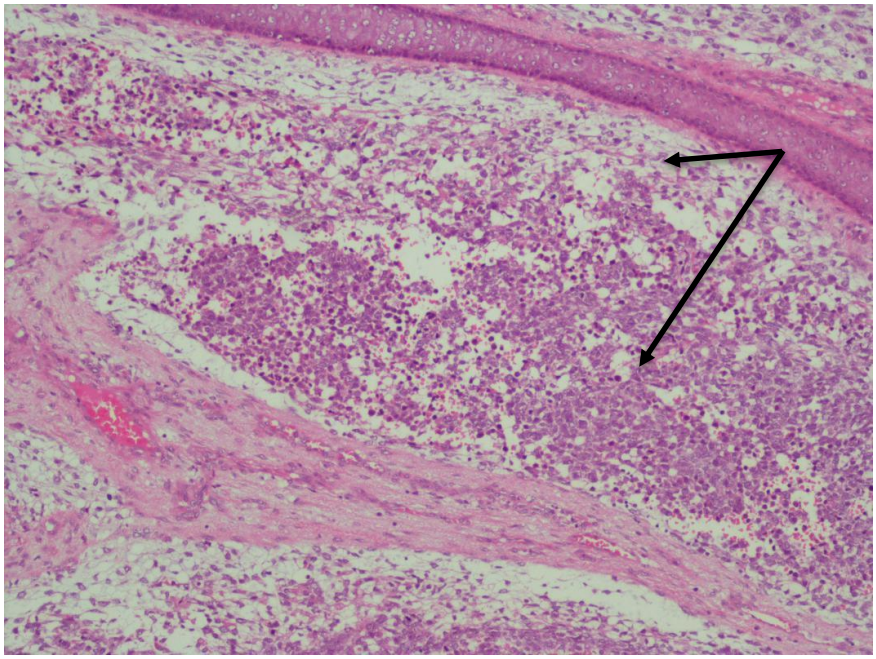
INTRODUCTION

Rhabdomyosarcoma(RMS) accounts for 41% of pediatric head and neck RMS,of which only 8% occur in the temporal bone.(1) RMS in the head and neck can be classified based on its location of the primary tumour:orbital, parameningeal and nonparameningeal.(2) Histologically RMS is classified into embryonal, alveolar,monophormous round cell,pleomorphic and botyroid type.(1,3)The alveolar subtype accounts for only 20% of cases,as rare as one case per one million.(4)It is mainly occurs in skeletal muscles of the trunk and limbs.To the best of our knowledge,there has been only one literature by Ragsdale et al(3),of ear alveolar RMS.We report a case of alveolar RMS at the external ear canal (EAC) in a 7-year-old boy.

CASE REPORT

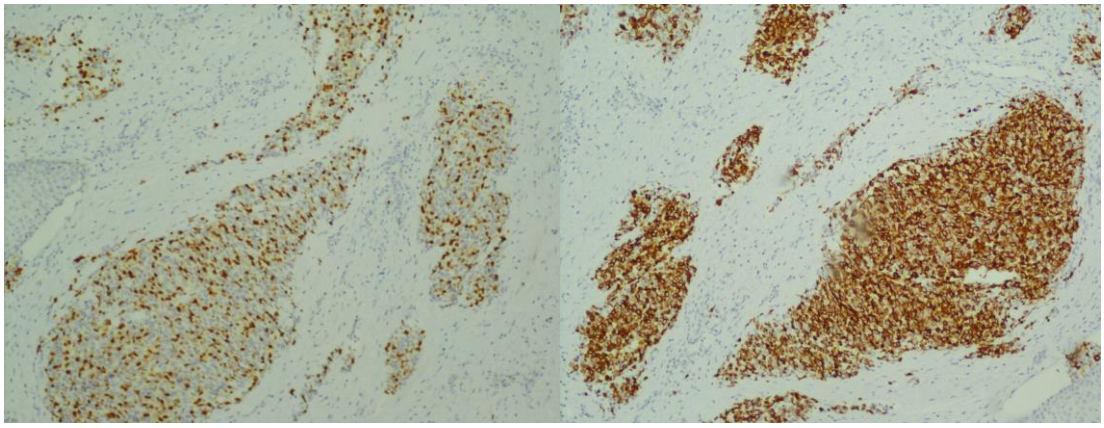
A 7-year-old boy presented with right otalgia and swelling of the external ear for two weeks.The ear canal was edematous and erythematous,with intact tympanic membrane.Patient was treated as otitis externa with topical antibiotic eardrops for two weeks. Patient however defaulted returning two months later, with complaints of serosanguinous ear discharge and disproportionate otalgia. Examination showed a polyp occupying the entire circumference of external ear.

Biopsies from right external ear canal reported as alveolar rhabdomyosarcoma with multiple nests of malignant cells.Central cellular discohesion with peripheral cells resembling pulmonary alveoli seen.The malignant cells were positive of myogenin and desmin. (Figure 1a,1b,1c)



(a)

Figure 1 (a) : Hematoxylin and eosin staning Medium power magnification (x200). Central clusters of tumour cells with discohesive periphery (black arrow)

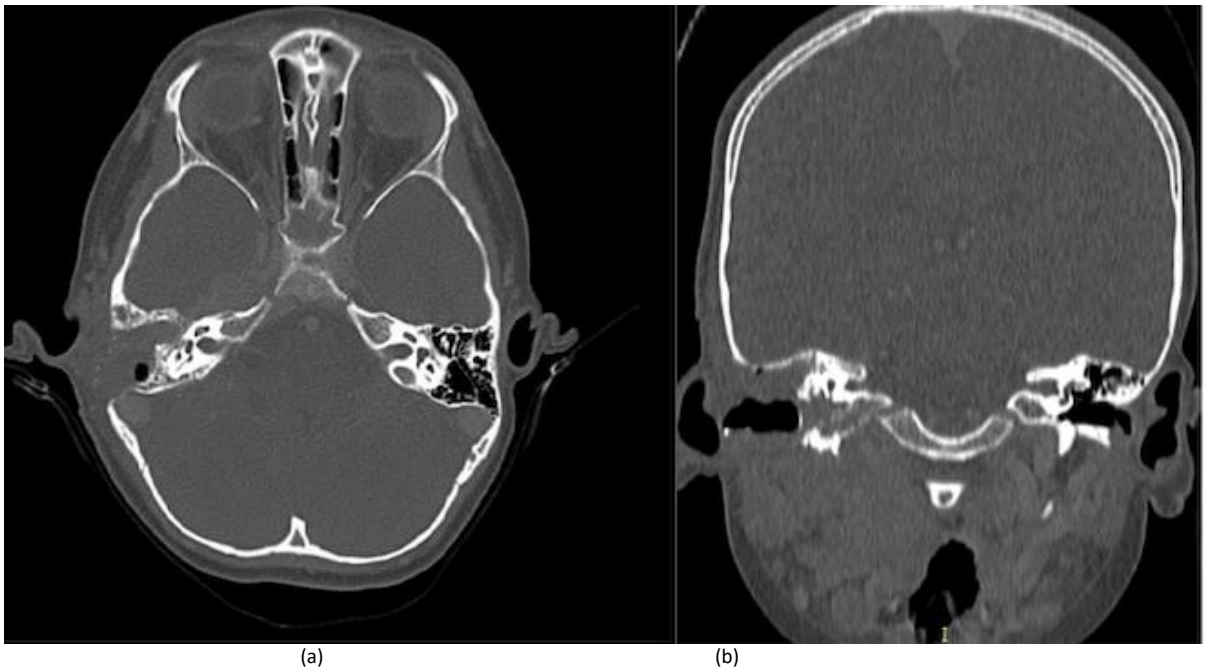


(b) (c)

Figure 1 : (b) Tumour cells positive for Myogenin **(c)** Tumour cells positive for Desmin.

High resolution computed tomography (HRCT) temporal bone and neck (Figure 2(a),(b)) revealed an enhancing soft tissue mass in the right external and middle ear with erosion of the ossicles,mastoid aircells and petrous apex,with intracranial extension. There was enlarged right cervical level V lymph node with no distant metastases.

Patient received four courses of neoadjuvant chemotherapy in accordance with the European Paediatric Soft Tissue Sarcoma (EpSSG) RMS 2005 consisting of ifosfamide, vincristine,actinomycin-D, doxorubicin(IVADo) regime.Imaging after 3months of neodjuvant chemotherapy showed substantial tumour regression in the external and middle ear with resolution of intracranial component.



(a) (b)

Figure 2 : Contrast enhanced computed tomograghy (a) axial view enhancing soft tissue mass in the right external and middle ear with erosion of the ossicles,mastoid aircells (b) soft tissue mass extending intracranially.

Patient underwent right lateral temporal bone resection with tumour clearance. (Figure 3) Patient continued to receive a total of 5 courses of chemotherapy and 28 fractions of radiotherapy 50.4Gy.Magnetic resonance imaging (MRI) performed six months after completing treatment,showed no evidence of recurrence.

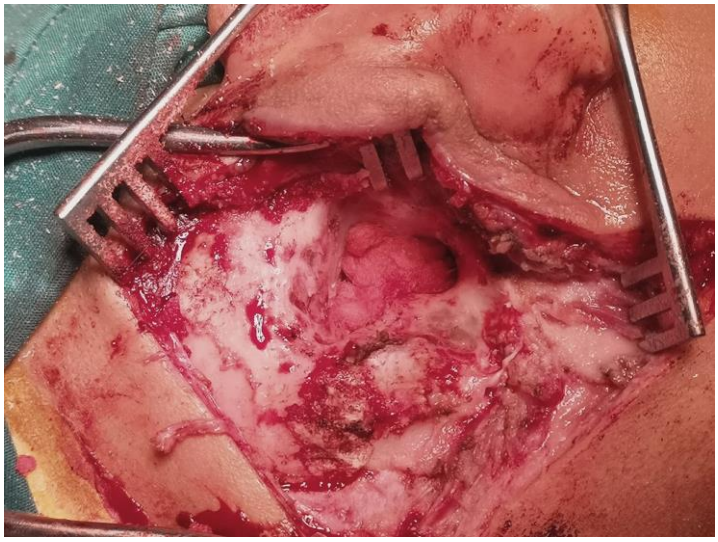


Figure 3: Soft tissue mass seen occupying the right EAC, with erosion of posterior wall , extending into the mastoid cavity.

DISCUSSION

Rhabdomyosarcoma is a malignant tumour of striated muscle with the capacity for skeletal muscle differentiation. Although there are various cases reported as RMS in the middle ear and mastoid cavity, few were from the external ear. The alveolar subtype is rare and its localization in the ear is exceptionally unusual.Features mimicking otitis externa, or otitis media, aural polyp, granulation tissue bleeding or facial palsy may occur.The former was seen in our patient,further delaying diagnosis.

Microscopically,alveolar RMS present as discohesive cells with round nuclei, separated by fibrous septa. Histology alone is insufficient as it mimics other small round cell tumours hence immunohistochemistry is done for confirmation.Immunohistochemistry shows strong and homogeneous expression to myogenin in alveolar RMS, which is less so in embryonal subtype.(3)

HRCT and MRI helps assess bony involvement and soft tissue invasion such as dural and intracranial involvement,respectively.MRI is utilized to assess residual tumour and response post treatment.CT thorax, ultrasound abdomen and bone marrow biopsy is advised,as 15 % of children with RMS will have distant metastases at the time of diagnosis.(3)

EpSSG treatment protocol has designed a multi-modality approach in management. Treatment protocol is tailored individually depending on characteristics of the primary tumour. EpSSG subcatogorizes 4 risk groups into:low,standard ,high and very high risk.It categorizes clinically confirmed lymph node into the high or very-high risk group,as seen in our patient.

Surgery remains crucial,however radical resection procedures are unwarranted and avoided owing to its high functional morbidity. With introduction of newer chemotherapeutic agents into treatment protocols and combined radiotherapy,survival rates have improved.More than 70% of children with localized disease who received combined modality treatment, survived 5 years after diagnosis (4).

CONCLUSION

A high index of suspicion of a malignant pathology in paediatric patients presenting with chronic bloody ear discharge with aural polyp,must be considered.Our case explores how an unusual presentation of RMS could be misdiagnosed for a benign cause.

Conflict of interest
No conflict of interest was declared by the authors.

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