CASE REPORT

A CASE REPORT OF UNUSUAL PARA-MENINGEAL RHABDOMYOSARCOMA

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Abstract

Rhabdomyosarcomas arise from mesenchymal cells destined to form skeletal muscle, but they are often found at sites where skeletal muscle is typically not found. The parameningeal site has a poor prognostic risk, if it is associated with intra-cranial extensions and skull bone erosions. This is a case report of a five-year-old girl who presented with cranial nerve palsy, and later diagnosed as rhabdomyosarcoma of the mastoid and middle ear, with infiltration of the cerebellopontine angle of the brain. She did not respond to chemotherapy and rapidly deteriorated, succumbed to the disease within a short time following the diagnosis.

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Introduction

Rhabdomyosarcoma (RMS) is the most prevalent soft tissue tumour in children and adolescents, accounting for 5% of all paediatric tumours. The head and neck are the most common primary sites for rhabdomyosarcomas, followed by the genitourinary tract, limbs, thorax, and retroperitoneum. In the head and neck, the commonly involved sites are the orbit, parameningeal sites (nasopharynx, nasal cavity, paranasal sinuses, temporal bone, pterygopalatine fossa, and the infratemporal fossa), and non-parameningeal sites. RMS originates from embryonal mesenchyme with the ability to mature into skeletal and smooth muscle, adipose and fibrous tissue, bone and cartilage. They are mostly chemosensitive with 62-66% of ten-year survival [1,2]. The survival rate worsens in the presence of metastatic disease, with rates as low as zero quoted in some studies [3]. The presentation of head and neck RMS depends on the site affected, but with the presence of cranial palsies, base of the skull bony erosion and intra-cranial extension [4], it is linked with high treatment failure rate [5]. Various subtype identified histologically namely embryonic, alveolar and pleomorphic. Often RMS presents as painless mass, but symptoms and signs of

presentation are depended on the site of the tumour. Treatment for RMS is individualised for each patient and would require various disciplines involvement such as radio-oncology, surgery and oncology.
We present a rare case of a child diagnosed with localised and aggressive parameningeal embryonal rhabdomyosarcoma, and later developed several complications leading to her fatal outcome.

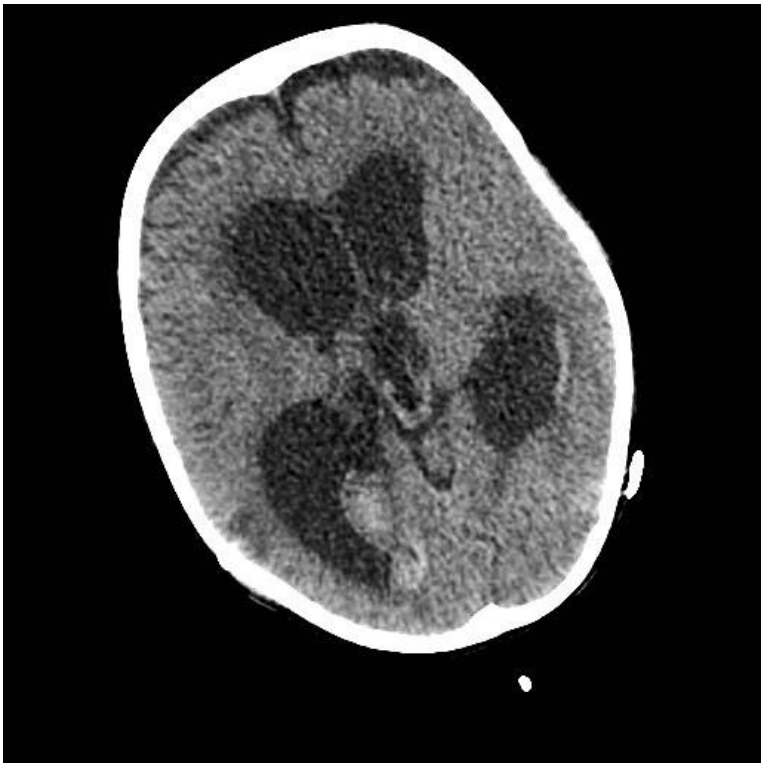
Case History

A five-year-old girl presented with a four-week history of intermittent vomiting, loss of balance, headache and right facial droop. She had a history of recurrent ear infections and reduced hearing, which required several antibiotics courses. Examination revealed a firm tissue filling the right ear canal and mastoid area, a right sided facial nerve palsy, and a broad gait. Her neurological examination showed poor coordination.

The initial blood result showed haemoglobin 107 g/L, platelet 528 x 10^9/L, white cell count 12.3 x 10^9/L, sodium 139 mmol/L, potassium 3.5 mmol/L, urea 4.1 mmol/L, creatinine 40 µ mol/L, alanine transaminase 13 U/L, alkaline phosphatase 152 U/L, bilirubin 3 µmol/L and C-reactive protein 13 mg/L.

Computerised Tomography scan of the head showed a 2.2 cm mass in the region of the right cerebellopontine angle, causing local mass effect with deviation of the brainstem. There was acute haemorrhage within the mass and acute subarachnoid haemorrhage involving the ventricles. There were also expansion and lucency of the right petrous temporal bone apex with involvement of the skull base and extension of soft tissue into the right middle ear.

Figure 1. CT brain showed mass at cerebellopontine angle with deviation of brainstem and hydrocephalus. This mass was an extension from right middle ear mass with destruction of skull base.



She underwent exploration biopsy of the right ear and mastoid. Histopathology findings revealed tumour cells comprised of nests of small round blue cells separated by thick fibrous bands with a 'strap cell' morphology. Immunohistochemistry showed the tumour cells positivity for desmin and Myogenic differentiation 1 (MYOD1) (40-50%). These features were consistent of an embryonal rhabdomyosarcoma.

Staging imaging of the chest and abdomen did not reveal any evidence of metastatic disease, and examination of the Cerebrospinal fluids did not reveal any malignant cells. There was also normal bone marrow morphology with no tumour cells infiltration. The overall findings were consistent with a localised para-meningeal embryonal RMS.
Her clinical course was punctuated by recurrent brain bleeds, necessitating several neurosurgical interventions. Despite receiving a course of chemotherapy, her neurologic function progressively deteriorated, and the tumour size continued to increase. She eventually succumbed to the disease after 9 weeks post diagnosis.

Discussion

Embryonal RMS usually affects neonates and younger children with an intermediate prognosis. Prognostication of RMS include site of the lesion, size of the tumour and extent, lymph node status and distant metastasis. The reason behind the tumour appearance in parameningeal area remained unknown. Immunohistochemistry is a useful tool for diagnostic and prognostic purposes of the underlying RMS. Recognition of basic histologic and cytological pattern separates RMS into further subtypes. The distinction between alveolar and embryonal RMS can be achieved via molecular and cytogenetic investigations. Clinical presentation such as chronic unresolved otitis media should trigger healthcare professionals towards detailing investigation to avoid a misdiagnosis. Patient with middle ear and mastoid disease is considered to have a poor prognosis [6]. Treatment modalities include surgical removal of the tumour followed by chemotherapy and radiotherapy. The basic aim of treatment is to achieve disease control locally and prevention of distant metastasis. This case report highlights the relatively aggressive nature of an embryonal parameningeal RMS, given that the onset of symptoms up to time of demise was relatively short. The presence of facial nerve palsy was well correlated with the intra-cranial lesion in the cerebellopontine angle. In the earlier studies [3,4], evidence of bony erosion and intracranial tumour extension provide a poorer prognostication with higher treatment failure rate [5]. Not only did the patient fail to respond to treatment, several catastrophic events in form of cerebral bleeds occurred, which made the overall outcome direr.

Conclusion

In conclusion, parameningeal RMS with intra-cranial extension or bone erosion may pose a management challenge for clinicians. In this patient, the disease was surgically difficult to accessand she developed recurrent tumour bleeding and poor chemoresponsiveness. Multidisciplinary team discussion would be suitable in a complex case of RMS.

Competing interest

The authors declared that they have not received any financial and material support nor having any competing interest.

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