

Picture Stories

Parameningeal embryonal rhabdomyosarcoma presenting as facial nerve palsy in a young girl

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Introduction

Rhabdomyosarcoma (RMS) is a fast growing, highly aggressive, malignant tumour, consisting of cells derived from the progenitor cells of myoblasts called satellite cells which exhibit a significant tendency for myogenesis¹. It comprises over half of paediatric soft tissue sarcomas². About 35% of RMS are located in the head and neck region, and are classified into para-meningeal (PM), orbital, non-orbital and non-PM subtypes. PM subtype has the worst prognosis, being associated with a high recurrence rate and generalized metastases through haematogenous and lymphatic routes^{1,3}. All patients with PM-RMS, irrespective of age, need adequate radiotherapy (RT)^{3,4}. Overall outcome depends on stage of the disease, site of the disease, age of the child and cellular type of the RMS.

Case report

A girl aged 5 years was admitted to our institution with a history of right sided facial swelling and deviation of the mouth to the left side of two weeks duration. On examination she had a right sided lower motor neurone type of facial nerve palsy (FNP) along with firm enlargement of the left parotid gland (Figure 1).

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Figure 1: Right sided facial nerve palsy and parotid swelling

*Permission given by parents to publish photograph

Her other cranial nerves were intact and the past medical history was not significant. Computer tomography of the brain and neck showed a locally invasive mass lesion with extra-cranial and intra-cranial components with the epicentre within the right parotid gland (Figure 2).



Figure 2: CT scan showing extensive local invasion of tumour

Immunohistochemistry of the tumour was indicative of RMS, as small round cells were positive for Vimentin, CD99 and Desmin, negative for LCA, PanCK and Chromogranine was equivocal. Myo D1 repeated at NCI showed strong nuclear positivity. Immunohistochemistry and radiological evaluation showed locally advanced embryonal RMS of para-meningeal origin. She had no lung metastasis. She was given chemotherapy (EpSSG sub group D – IVA) followed by RT, as the tumour was not resectable.

Discussion

RMS incidence was highest in 1–4 year old children and lowest in 15–19 year old children⁴. Our patient was 5 years old, just older than the high risk category. In RMS facial nerve involvement can indicate a more locally advanced disease stage and hence a more adverse prognosis^{3,5}. Our patient presented with FNP secondary to local invasion of the PM-RMS. RMS exhibits a fast and aggressive growth and is particularly painless in children. In our patient there was local invasion of the tumour. In our case neuroblastoma, lymphoma and Ewing's sarcoma were considered as differential diagnosis and the diagnosis was confirmed by immunohistochemistry. Treatment of RMS needs a multidisciplinary approach which includes radical surgical excision for resectable tumours, or neoadjuvant chemotherapy followed by surgery and radiotherapy if indicated, and further adjuvant chemotherapy. Our patient received neoadjuvant chemotherapy followed by radiotherapy, as the tumour was not resectable. Furthermore she is to receive adjuvant chemotherapy as well.

While uncommon, facial nerve paralysis may be the first presenting symptom in cases of childhood RMS. A careful evaluation enables early diagnosis and treatment of this highly invasive malignant tumour.

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