

Case Report

A CASE REPORT ON PRIMARY RHABDOMYOSARCOMA OF SCALP WITH REVIEW OF LITERATURE

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ABSTRACT

Neoplasm's of the head and neck region account for approximately 5% of all childhood malignancies (Dickson *et al.*, 2006). Rhabdomyosarcomas are second most common neoplasm of head and neck in paediatric population second on to lymphomas (JIAPS oct-dec 2009). Rhabdomyosarcomas are the most common soft tissue tumour in children and account for 5-8% of childhood cancers (Schepper, 2006). These tumours arise from the mesenchymal tissue and are fast growing tumours, they cause localised pressure effects on neurovascular structures, and have a predilection for growing into bones (Schepper, 2006). This is a case report on scalp Rhabdomyosarcoma of a paediatric patient aged 1 year. After proper clinical and radiological evaluation, the diagnosis was confirmed by biopsy followed by histopathology by pathology department of our institution. Survival varies dependant on primary location, histological type, local invasion and metastases. Overall 5 year survival is in the region of 75% (kransdorf *et al.*, 2006).

Keywords: *Rhabdomyosarcoma, Paediatric Soft Tissue Tumour*

INTRODUCTION

This is a case report on a rare case of primary scalp rhabdomyosarcoma. While soft tissue sarcoma in paediatric population is most commonly rhabdomyosarcoma its presentation in scalp is as such very rare. Here the clinical feature/ presentation, imaging and pathological findings of this particular case are discussed.

CASES

The patient was referred to us by the paediatric surgery department. The patient presented with a fast growing non painful cystic swelling in the scalp region. Plain radiographs were obtained as first radiological investigation. MRI of head and neck region was then performed using Philips 1.5 tesla machine. Images were obtained in axial, coronal and sagittal plane. T1 post contrast images were also obtained after injecting Gadolinium.

History was given by the patient's mother. Patient presented with soft tissue swelling in the left parieto-occipital region which was gradually increasing in size since 5months. The swelling was not painful. There was no history of trauma, headache, fever, seizures, no signs of meningeal irritation or other neurological symptoms.

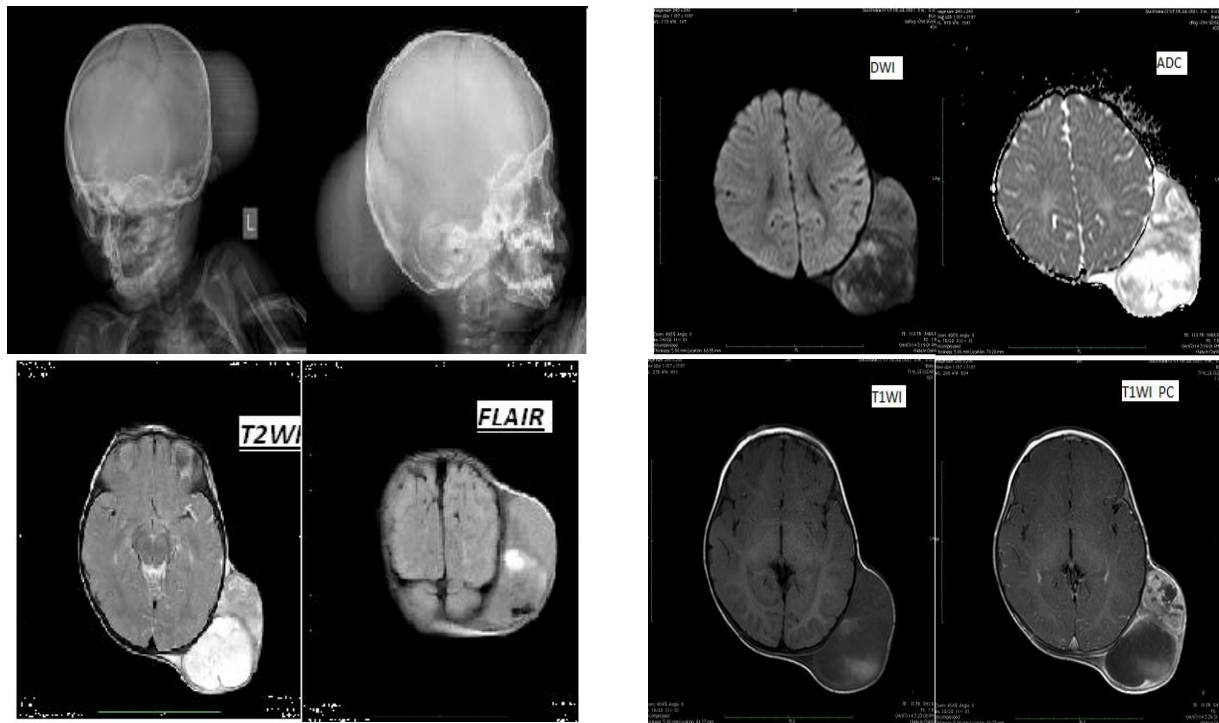
There was no history of any major illness. No history of any maternal illness. Birth history was normal. Child showed normal development of milestones.

On examination a large cystic consistency lesion was noted on left parieto-occipital region. Lesion was nontender, fixed. After proper clinical examination the radiological investigation were requested.

X-ray images show soft tissue scalp swelling involving left parieto-occipital region. No evidence of calcification within it. No evidence of adjacent bony involvement.

T2WI show well defined heterogenous predominantly hyperintense lesion with areas of solid and cystic components. T1WI show iso to hypointense signal in the lesion and shows heterogenous post contrast enhancement with areas of necrosis within. No evidence of any intracranial extension noted.

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Conclusion

After clinical and radiological examination the patient was diagnosed by biopsy and histopathology. CT scan delineates the bony involvement better and soft tissue involvement of this lesion very well depicted by MRI and also it is helpful for proper tumour staging. It is necessary before a surgery to know the complete extent of tumour involvement which is best depicted by CT scan or MRI images.

DISCUSSION

Rhabdomyosarcoma originates from mesenchymal tissue and consists of rhabdomyoblasts.

It is a common tumour of the soft tissues in paediatrics, representing 13% of all pediatric malignant neoplasms (weiss *et al.*, 2001).

Its incidence is approximately four new cases per million in children under age 15 (Raney *et al.*, 1989).

About one half of patients are less than 5 years old and 70% less than 10 (Marer, 1978).

It has a 3:1 ratio of white to black children, and approximately 60% of cases arise in males (Marer, 1978).

Four types are seen histologically, arising from rhabdomyoblasts: embryonal, botryoides, pleomorphic and alveolar.

Forty percent of rhabdomyosarcomas arise in the head and neck, although they can arise in any part of the body (Cunningham *et al.*, 1987). The sites where tumour originates from head are orbit, nasopharynx, middle ear, paranasal sinuses, and auditory canal (external). As the tumor is highly aggressive, distant metastases is common as well as spread to bony structures is quite common. In head neck region, these tumors cause bone destruction locally and extensively as the mass infiltrates and expands.

The scalp rhabdomyosarcoma can be mis-diagnosed by the clinician as hemangioma, lymphangioma, lymphoma, and lymphadenopathy.

Site Distribution

Rhabdomyosarcomas are found essentially anywhere in the body (weiss *et al.*, 2001).

- head and neck:
 - orbit
 - oro/nasopharynx and palate
 - sinuses, middle ear, mastoid

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- genito-urinary:
 - paratesticular
 - bladder
- extremities:
- other:
 - trunk and thorax
 - gastrointestinal tract

Pathological Classification

Rhabdomyosarcoma is divided into 3 sub types (Schepper, 2006; Kransdorf *et al.*, 2006):

- embryonal rhabdomyosarcoma
 - spindle cell rhabdomyosarcoma
 - botryoid rhabdomyosarcoma
 - anaplastic rhabdomyosarcoma
- alveolar rhabdomyosarcoma
- pleomorphic rhabdomyosarcoma

Radiographic Features

The appearances are non-specific and difficult to distinguish from other types of sarcomas. The site and clinical presentation of patient is of much value in getting to a specific diagnosis.

Plain film

Plain X-rays are the first investigation, although these are non specific but may provide useful information about the site, calcification within the lesion rough extent adjacent bony involvement

Ultrasound

Well defined, heterogeneously echoic, irregular mass lesion- usually hypoechoic.

CT

CT scans show well defined soft tissue density mass lesions with moderate heterogenous post contrast enhancement as well as adjacent bone destruction.

MRI

It depicts better soft tissue / local spread (intracranial)

Signal specifics include:

TIWI

Iso to hypo -intensity, signal similar to muscle

Areas of haemorrhage within the lesion are seen in some subtypes

T2WI

Hyperintense with prominent vascular flow voids

TI Post Contrast Images (Gadolinium)

Show mild to moderate heterogeneous enhancement

Treatment and Prognosis

Up to 20% of patients have metastases at the time of diagnosis (Kransdorf *et al.*, 2006). These are typically to lung and bone marrow.

Patients are treated with a combination surgical excision, chemotherapy and radiotherapy:

- **Surgery:** Complete resection of primary tumour and is done sometimes after down-staging by chemoradiotherapy.
- **Chemotherapy:** Drugs include vincristine, cyclophosphamide, dactinomycin, Adriamycin, ifosfamide, VP-16.
- **Radiotherapy:** External beam radiation.

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