ORIGINAL RESEARCH PAPER

Pathology

PRIMARY INTRACRANIAL RHABDOMYOSARCOMA WITH SCALP EXTENSION – DIAGNOSTIC ROLE OF CYTOLOGY

KEY WORDS: Intracranial, Rhabdomyosarcoma, Pediatric, Scalp, Cytology

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ABSTRACT

Primary intracranial rhabdomyosarcoma (PIRMS) is a rare tumor of mesenchymal origin. It commonly affects the pediatric population with rapid downhill progress and a predilection for the cerebellum. We present a case of PIRMS in frontoparietal location with scalp extension.

INTRODUCTION

PIRMS is an extremely rare entity, being common in young children, especially involving cerebellum. [1,2,3,4]

CASE STUDY

A 5-year-old male patient presented with scalp swelling, headache and episodes of unconsciousness for 2 months. Radioimaging showed an extra-axial lesion in the right frontoparietal region measuring 5.5×4×4.5 cm. Mass was isointense on T1W and hyperintense on T2W with intense contrast enhancement. The lesion was eroding skull and abutting onto scalp, suggesting a primitive neuroectodermal tumor.

Fine needle aspiration cytology from scalp lesion showed small round cell population. **(Figure 1 and 2).**

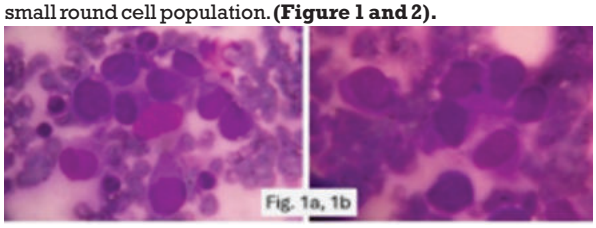

Figure 1a. Round tumor cells with occasional tadpole cell. **Fig 1b. R**osette formation (Cytology smears, Giemsa stain)

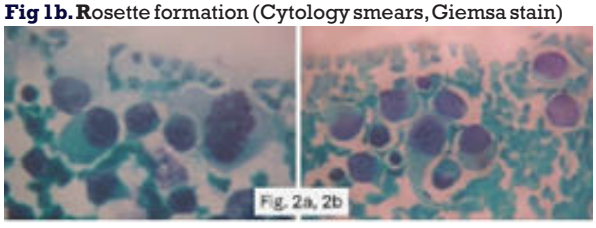

Fig. 2a: Tadpole cells with pleomorphism and mitoses. **Fig. 2b:** Tumor cells with cytoplasmic striations (Cytology smears, Papanicolaou stain)

On surgical resection, the tumor was relatively circumscribed. Histopathology confirmed rhabdomyosarcoma **(Figure 3a, 3b).** The tumor was positive for desmin and myogenin, but negative for GFAP.

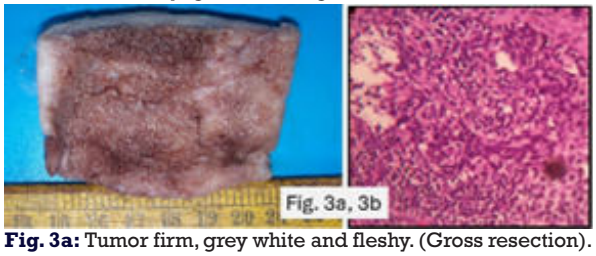

Fig. 3a: Tumor firm, grey white and fleshy. (Gross resection).
Fig. 3b: Sheets of rounded tumor cells. (H&E stain, 10 x)

PIRMS is an extremely rare entity. In pediatric population is involves cerebellum, while adults show predilection for supratentorial location. PIRMS may arise from heterotopias or pluripotent mesenchyme. [1,2,3,4]

Surgical resection followed by adjuvant radiotherapy is advised. PIRMS has a dismal prognosis. Our patient expired within 3 months of diagnosis.

CONCLUSIONS

PIRMS is rapidly growing, widely invasive tumor with high mortality. Cytology can aid in early diagnosis of this rare entity.

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