CASE REPORT

Congenital Osteolytic Dural Fibrosarcoma Presenting as a Scalp Swelling
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ABSTRACT
An extremely rare case of congenital dural fibrosarcoma is reported in a 2 months old child who presented with scalp swelling since birth. CT scan revealed an osteolytic lesion compressing the underlying atrophic brain. Tumour was completely excised and duroplasty was done with a patch graft. Postoperative CSF leak was managed with aspirations and lumbar puncture.

Key words: Congenital neoplasms. Fibrosarcoma. Intracranial tumour. Scalp swelling.

INTRODUCTION
Congenital brain tumours are uncommon. Teratoma and craniopharyngioma are frequent because of developmental defect. Astrocytomas are the next commonest. Few cases of congenital menigiomas are also reported in the literature. Congenital fibrosarcoma is a rare malignancy occurring most commonly in the extremities (71%) and in axial locations in 29% of the patients. Congenital fibrosarcoma affects chiefly the distal parts of extremities, while the adult variety usually involves the thigh. Dural fibrosarcomas are very rare tumours representing only 0.5% of all CNS sarcomas. Congenital intracranial fibrosarcoma has not been reported. Authors report a rare case of osteolytic dural fibrosarcoma presenting as scalp swelling in a 2 months old infant.

CASE REPORT
A 2 months old boy presented with left fronto-temopral painless scalp swelling since birth (Figure 1). It was firm in consistency and was fixed to bone; the overlying scalp was free all over. It was non-pulsatile and crying impulse was absent. Features of raised intra-cranial pressure were absent. CT scan of brain revealed an osteolytic lesion compressing the underlying atrophic brain (Figure 2). Complete surgical resection was performed with surrounding bony margin. Tumour was separate from the underlying brain by an arachnoid plane. Dural defect was repaired with artificial dural graft. Histopathology report revealed the lesion to be a fibrosarcoma.

He had CSF collection beneath the scalp which was managed with aspiration and lumbar puncture.

Figure 1: Scalp swelling.

Figure 2: CT scan brain showing osteolytic mass compressing underlying atrophic brain.

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Otherwise postoperative period was uneventful. No recurrence was observed at 3 months follow-up.

**DISCUSSION**

Congenital fibrosarcoma is a low grade malignant tumour which is locally aggressive. It has low metastatic rate and high local recurrence. It has good prognosis as compared to the adult type. Embryonal neoplasms arise as a result of two mutational events in the genome. The first mutation is pre-zygotic in the familial cases and second mutation is postzygotic found in non-familial cases. Initiation, promotion and progression phases of carcinogenesis can be applied to these tumours. Genetic mutation can be inherited or may be a de novo phenomenon. Chromosomal translocation and specific gene fusion result in dysregulated TrcC expression.4

Maternal or fetal exposure to radiation, drugs or viruses may be the mechanism for these tumours. A higher incidence of dural fibrosarcomas has been reported in patients with neurofibromatosis. Malat et al. have reported a case of fibrosarcoma arising from spinal dura without neurofibromatosis.5

Aydin et al. have reported a case with possible radiation induced fibrosarcoma in a case with medulloblastoma.6 Kaminski et al. have described dural fibrosarcoma in a patient receiving chemotherapy for glioblastoma multiforme in a child.7

This case presented as congenital scalp swelling with no known causative factor. CT scan was suggestive of meningioma but the histopathology diagnosis turned out to be the fibrosarcoma. Follow-up at 3 months period did not reveal any recurrence. However, long-term follow-up is planned to avoid local recurrence which is a common phenomenon in this locally aggressive tumour.

**REFERENCES**