A Rare Case of Juvenile Fibrosarcoma of the Eyelid

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ABSTRACT
Juvenile fibrosarcoma is not an uncommon tumor in children; however, eyelid involvement is extremely rare. Very few cases have been reported in the literature. This is the first case of JFS of the eyelid in Tribhuvan University Teaching Hospital and the first diagnosed case in Nepal. Histologically, it is similar to adult fibrosarcoma but it has an excellent prognosis. Surgical excision is the mainstay of treatment; however chemotherapy has been proved effective. Here we present a case of juvenile fibrosarcoma of the eyelid in a four year female child presenting with a rapidly enlarging, painless, left upper eyelid mass. An incisional biopsy, from the eyelid mass, showed typical histopathological features of juvenile fibrosarcoma.

Key words: Juvenile fibrosarcoma, eyelid, a rare case

INTRODUCTION
Juvenile fibrosarcoma (JFS) in children is histologically similar to classic adult fibrosarcoma but it is considered a separate entity because of its favorable prognosis.1,2 JFS accounts for approximately 13% of fibroblastic-myofibroblastic tumors in children and adolescents and 12% of soft tissue malignancies in infants.2 Common sites of JFS include distal soft tissue of extremities (61%) followed by trunk (19%) and head & neck region (16%).2 JFS presenting as an eyelid mass is extremely rare and only very few cases have been reported so far.3 Even in a large series of population-based studies on malignant eyelid tumors, not even a single case was identified.4 Clinically, the patient presents with a solitary, rapidly enlarging, painless, non-tender mass which may exceed 30 cm². Histologically, the tumor is cellular and composed of immature, spindle-shaped, fibroblastic cells in a herringbone pattern or interlacing fascicles with variable mitotic activity.5 In the present case, the patient presented with a rapidly growing, left upper eyelid mass and an incisional biopsy showed typical features of juvenile fibrosarcoma.

CASE REPORT
This four year female child presented with a rapidly enlarging, painless swelling of the left upper eyelid of two months’ duration. Swelling was spontaneous to begin with and no history of any trauma discovered. Milestones of the baby were normal and she did not have any significant past, personal or family history. Clinical examination revealed a 5x4 cm tumor of the left upper eyelid with an intact, erythematous, tense, overlying skin without any tenderness (Figure 1.). Her vision or the right eye was 6/9 and the vision of the left eye could

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not be assessed because of the tumor. An incisional biopsy was sent for histopathological examination.

**HISTOPATHOLOGICAL EXAMINATION**

Grossly, the specimen was composed of multiple pieces of gray-white tissue and the entire specimen was submitted for histopathological examination. Histopathological examination of hematoxylin and eosin (H&E) stained sections revealed a cellular soft tissue neoplasm composed of uniform population of immature spindle cells arranged in intersecting fascicles and herringbone pattern. Tumor cells showed scanty cytoplasm, elongated mildly pleomorphic nuclei, inconspicuous nucleoli, hyperchromatic chromatin and variable numbers of mitotic figures (0-3 per high per field). Variable amount of collagen was noted between the tumor cells. Histopathological diagnosis was juvenile fibrosarcoma (Figure 2.).

**DISCUSSION**

Juvenile Fibrosarcoma accounts for 13% of fibroblastic-myofibroblastic tumors in children and adolescents.\(^1,2\) Common sites of involvement are distal extremities (61%), trunk (19%) and head & neck region (16%).\(^2\) Actual frequencies of JFS involving the eyelid has not been described so far, as very few cases have been published. Many studies have been published involving large number of cases of tumors of eye and ocular adnexae; however, fibrosarcoma of the eyelid has not been identified in any of these studies.\(^3-9\) Basal cell carcinoma, squamous cell carcinoma and sebaceous carcinoma are the most frequently encountered malignant tumors of the eyelid in most series.\(^5,6,7,8,9\) Sarcomas of the eyelid other than fibrosarcoma, however, have been described by Margo and Mulla in large population based studies.\(^4\) Clinically a rapidly growing painless swelling of eyelid over a period of a few weeks to months is the usual presentation of this tumor.\(^3\) The overlying skin is often tense, erythematous and ulcerated.\(^2\) In the study of Weiner and Hidayat, the age of the patients ranged from new-born to eight years and there was male predominance.\(^3\) The present case was a four years female with history of the upper left eyelid mass growing rapidly over a period of two months. In addition, this case also showed erythematous, tense, but non-ulcerated skin overlying the tumor. Histologically, the tumor was cellular and was composed of immature, spindle cells in a classic herringbone pattern or interlacing fascicles with variable mitotic figures, as described by Weiner and Hidayat.\(^5\) The nuclei were elongated with tapering ends with uniformly distributed hyperchromatic chromatin and inconspicuous nucleoli. A variable amount of collagen was seen in between the tumor cells. Giant cells, necrosis, hemorrhage and calcification which can be present in JFS\(^2\) were lacking in the present case. Histological differential diagnoses included rhabdomyosarcoma, fibromatosis and fibrous histiocytoma,\(^3\) but they were ruled out.

Despite rapid growth and a high degree of cellularity, most JFS have favorable prognosis compared with adult fibrosarcoma.\(^1,2\) Mortality ranges from 4% to 25%, and the recurrence rate is 5% to 50%.\(^2\) Metastasis is rare.\(^2,3\) In the study of Weiner and Hidayat, two of five tumors recurred locally but were non-metastasized.\(^3\) No definitive morphological or genetic prognostic factors have been identified.\(^3\) Although surgery is the mainstay of treatment, chemotherapy has been proven effective.\(^2\) The outcome of the patient in the present case could not be ascertained as she did not show up for follow-up.

**CONCLUSION**

Juvenile fibrosarcoma of the eyelid is a rare malignant tumor. Morphologically, the tumor is similar to the usual
adult fibrosarcoma but because of its excellent prognosis after surgical excision or chemotherapy, it is considered a separate entity. Because of its rarity in the eyelid, JFS is often forgotten in differential diagnosis of eyelid masses. Diagnosis established by biopsy of a specimen and the patient can be completely cured if diagnosed early. However, regular follow-up is needed for early detection of local recurrence, if any.

REFERENCES