**A Rare Presntation Of Fibrosarcomatous Change In The Background Of Dermato Fibrosarcoma Protuberans In A Child With Down Syndrome**

Abstract

Dermatofibrosarcoma protuberans is a cutaneous Soft tissue neoplasm with potential for local recurrence but distant metastasis is rare. Trunk and extremities are most commonly involved. This case presented 12 yr old male with Down syndrome presented with a recurrent swelling in left flank since 5 yrs. He had underwent wide local excision 3 times in past 5 yr. Despite the repeated excisions, the swelling recurred again for 4th time. The patient underwent wide local excision. Tissues were subjected to histopathological and immunohistochemical test subsequently. The tumour cells show mild to moderate pleomorphism, ovoid to fusiform nuclei and brisk mitotic active (10-11/10 hpf) at places. They were reactive to CD 34. The diagnosis of Dermatofibrosarcoma protuberans with areas of fibrosarcomatous change was given.

Key Words: CD34, Dermatofibrosarcoma protuberans, Fibrosarcoma, Mitotic figures, Down syndrome