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Case Report

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A Giant Soft Tissue Fibrosarcoma of Posterior Head and Neck: A Case Report

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Abstract

Sarcomas are malignant neoplasms originating from mesodermal tissues that constitute connective tissues of the body. This seven years old girl was born after an uncomplicated full term pregnancy. Her physical status at birth was unremarkable. Her younger sister is healthy. At age of one and a half year she developed swelling over posterior head and neck region that was gradually increasing. The rate of growth was more rapid from October 2014. She was admitted in National Institute of Neurosciences & Hospital (NINS). Local examination showed a giant swelling 22X17X21 cm in size over posterior head and neck region, fixed to the sub-occipital bone, firm in consistency. Her general and neurological examination revealed no abnormalities.The authors present a case of a giant soft tissue fibrosarcoma of posterior head and neck of a seven years old girl. The case has been discussed. [Journal of National Institute of Neurosciences Bangladesh 2015;1(2): 65-68]

Keywords: Sarcoma; soft tissue fibrosarcoma; children; head and neck

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Introduction

Sarcomas are malignant neoplasms originating from mesodermal tissues that constitute connective tissues of the body[1]. They are rare group of malignancies that constitute less than 1% of body's tumors, including those of the head and neck region[2-5]. 5–15% of adult sarcomas are in the head and neck region, while 20% of them arise from bones and cartilages and 80% arise in soft tissues[3,6-9]. Of soft tissues sarcomas, 80–90% affects adults and 10–20% is seen in children[6]. 7% of all pediatric malignancies are soft tissue sarcomas[7]. Bone sarcomas are very rare and 2600 cases are diagnosed with bone sarcoma in United States of America which constitutes only less than 0.2% of malignant tumors[10].

Soft tissue sarcomas (STS) have tri-modal age distribution with peaks in less than 10 years of age, between 11 and 40 years and the last is over 40 years[6]. In head and neck region, based on histological subtyping 50% of sarcomas are: osteosarcoma, rhabdomyosarcoma, malignant fibrous histiocytoma, fibrosarcoma and angiosarcoma [6,11]. In this paper, seven years old girl with a giant soft tissue fibrosarcoma of posterior head and neck region was reported.

Case Presentation

This seven years old girl was born to healthy unrelated parents after an uncomplicated full term pregnancy. Her physical status at birth was unremarkable. Her younger sister is healthy. At age of one and a half year she

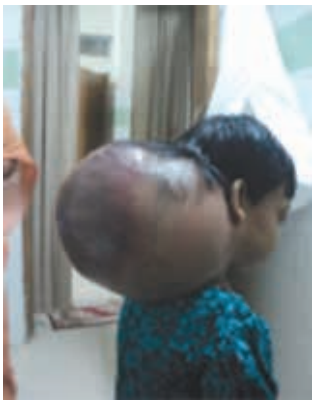
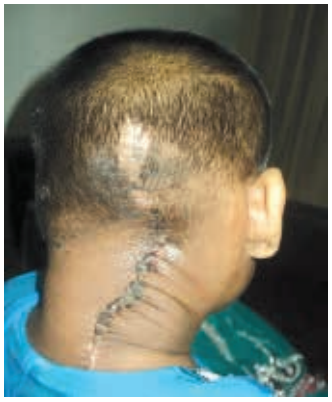

Figure 1: 7-year-old girl with soft tissue fibrosarcoma of posterior head and neck, Figure 2: After complete removal

developed swelling over posterior head and neck region that was gradually increasing. The rate of growth was more rapid from October 2014. She was admitted in National Institute of Neurosciences & Hospital (NINS) in may 2015. Local examination showed a giant swelling 22X17X21 cm in size over posterior head and neck region, fixed to the sub-occipital bone, firm in consistency. Her general and neurological examination revealed no abnormalities.

Neuroimaging studies: CT-scan of brain showed large mass lesion with central hypo dense area. MRI of brain showed large heterogeneous intensity mass lesion in sub-occipital region extending in neck region posteriorly. There is intense heterogeneous enhancement in post contrast study.

Operation and postoperative courses: The whole tumour was excised on 30.05.2015. The tumour was severely adhered to sub-occipital bone. Tumour was dissected sub-periosteal, started from above, convexity of sub-occipital bone changed to concave. Tumour was then dissected from posterior and side of neck meticulously. Feeder arteries were cauterized. Tumour was removed completely in a single mass. The tumour displayed big round mass of solid white tissue. After successfully achieving haemostasis, wound was closed in layers. 6 units blood were transfused during 7 hours operation. After an uneventful postoperative course, the patient was discharged on 21.06.2015 with advice to attend cancer hospital for further treatment.

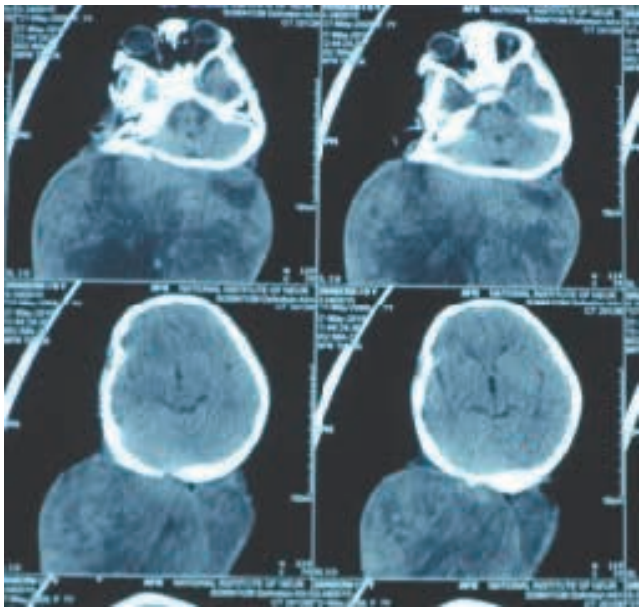

Figure 3: CT scan Brain showed large mass lesion with central hypodence area

Histopathological findings: Histopathological findings revealed fibrosarcoma. It is made of spindly cells present in fascicles. Moderate number of mitosis and foci of coagulative necrosis are present.

Discussion

Head and neck soft tissue sarcomas are rare and represent 1 to 5% of all corporal neoplasias[2]. The cause of the majority of sarcomas is yet unknown; however, it is thought that a number of environmental and genetic factors are closely linked to the development of these types of neoplasias. The survival range depends on the histological grade and the clinical stage. At five years, survival is approximately 60 to 70% and with local control, it becomes 60 to 80%[3]. The survival range depends on the histological grade and the clinical stage. Approximately 10 to 30% of patients presented with distant metastases within the first two years[5]. In general, young patients with low-grade, small and superficial sarcomas have a better prognosis than high-grade sarcomas[7].

Fibrosarcoma is defined as a malignant spindle cell tumor that shows a herringbone or interlacing fasicular pattern without the expression of other connective tissue cell markers[11]. Fibrosarcoma can arise in soft tissues or within bones. Intra-ossesous fibrosarcoma may develop enosteally or possibly periosteally, affecting the bone by spreading from adjacent soft tissue.

Fibrosarcoma can occur in any location, but the bone extremities are the main affected sites; occurrence in the maxilla is rare, with an incidence ranging from 0 to 6.1% of all primary fibrosarcoma of the bone. The mandible is the most common site for fibrosarcomas[12-13]. The clinical behavior of fibrosarcoma is characterized by a high local recurrence rate and a low incidence of locoregional lymph node and/or distant hematogenous metastases. However, hematogenus metastases may involve the lungs, mediastinum, abdominal cavity and bone[14].

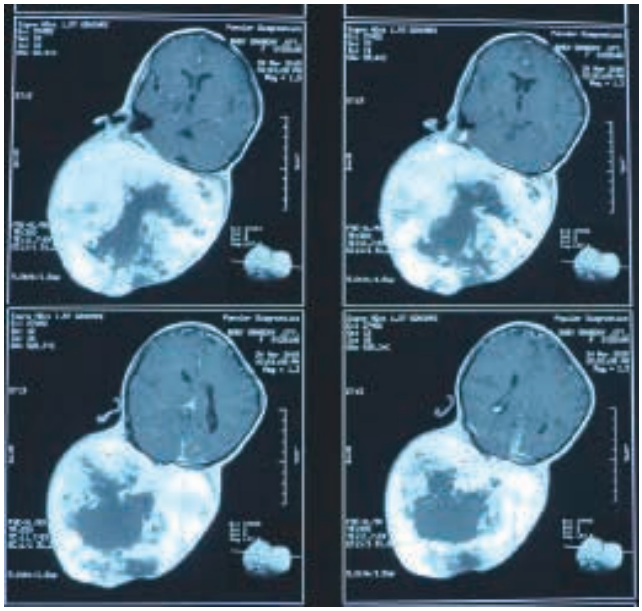

Figure 4: MRI of Brain showed large heterogenous intensity mass lesion in sub-occipital region extending in neck region posteriorly. There is intense hetrogenous enhancement in post contrast study

Local recurrence poses a serious and complex problem, particularly with occurrence of mediastinum infiltration, local destruction, airway compression, esophageal compression and extension. Radiation therapy is generally considered only in cases for which resection is impossible; chemotherapy is only used for palliative treatment. Prognosis is directly related to adequate, complete resection, which obviously requires early detection before the extensive involvement of soft tissue.

The histological appearance of fibrosarcoma does not allow a distinction between a tumor of the bone from one arising in soft tissue[15]. Histologically, the degree of differentiation is variable, from being comparable to a benign fibroma to a highly anaplastic tumor, thus presenting a diagnostic dilemma to histopathologists. Fibrosarcoma can be graded as either a low or high grade of malignancy. Low-grade fibrosarcoma shows spindle cells arranged in fascicles with low to moderate cellularity and a herringbone appearance. This type of fibrosarcoma has a mild degree of nuclear pleomorphism and rare mitosis, with a collagenous stroma.

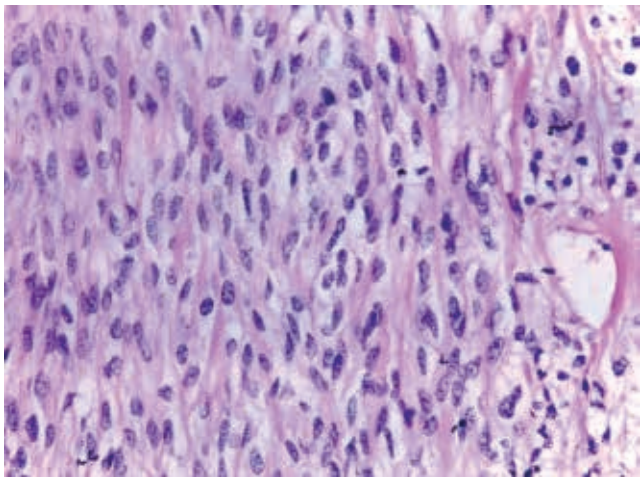

Figure 5: Microscopic image of Fibrosarcoma

High-grade lesions show an intense nuclear pleomorphism, greater cellularity and atypical mitosis. The nuclei can be spindle shaped, oval or round. The histological appearance of high-grade fibrosarcoma may be similar to other tumors, such as malignant fibrous histiocytoma, liposarcoma or synovial sarcoma. The positive immune-staining for vimentin, together with negative staining for muscular immune-markers, helps to diagnose fibrosarcoma[16].

Conclusions

Patients with sarcomas greater than 5 cm in clinical stage III or IV with positive surgical margins, can die from progression and metastases. To determine how a neoplasm is likely to behave, with or without treatment, it is necessary to know certain facts about the disease. The management of soft tissue sarcomas of the head and neck is particularly challenging and depends upon some prognostic factors. The assessment of prognostic factors, which correlate baseline clinical and experimental co-variables to outcomes, is one of the major objectives of clinical research.

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