PAEDIATRIC HAND TUMOURS-SYNOVIAL SARCOMA : A RARE MALIGNANCY

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Introduction

The rarity of soft tissue sarcoma overall and their even rarer presentation in distal portion of an extremity has limited the number of patients in most reports. Among soft tissue tumours, synovial sarcoma is uncommon & occurs rarely in children. Till date only one case of synovial sarcoma of the left palm has been reported in the English literature [1]. Here we describe another case of this rare entity and its management.

Case Report

A 6-years-old male child was admitted with complaints of gradually increasing swelling over the palm of left hand since 3 years. To start with the swelling was of a grain size & was noticed by the parents. Onset was spontaneous and initially the course was progressive, subsequently increasing rapidly to attain the present size. There was no history of pain. Examination revealed a small, 3x3 cm irregular swelling present over hypothenar region of left palm. It was firm, nontender and fixed (Fig 1). No distal neurovascular deficit was detected. A single 2x1 cm soft, mobile and nontender lymph node was palpable in the left axilla which was biopsied and turned out to be reactive hyperplasia. No other lymph nodes were detected clinically or per-operatively. Radiographs of hand and chest were normal. Computed tomography scan of hand revealed no evidence of any bone erosion. An incisional biopsy was done which was reported as synovial sarcoma with predominant epithelial component. Local wide excision with 2cm margin clearance was carried out on 26 March 1998 at CH(SC) Pune. The skin loss was made up with application of a split skin graft. Post operative recovery was uneventful. Histopathological examination of the excised specimen confirmed the diagnosis of synovial sarcoma (epithelioid variant) with incomplete clearance of deeper aspect. The immunohistochemistry profile of the tumour revealed cytokeratins (CK) strongly positive, vimentin and epithelial membrane antigen (EMA) positive and smooth muscle antigen (SMA) focally positive. Radiotherapy 6600 cGy was given. Patient was under regular follow up till the radiotherapy treatment was going on but subsequently lost to follow up.

Discussion

Malignant soft tissue sarcoma of hand is rare in children. Most large series on hand tumours report findings from adult patients. The overwhelming majority of reported soft tissue tumours of hand and wrist have been benign. In contrast to the predominance of synovial sarcoma in adults, most cases of soft tissue sarcoma in children are rhabdomyosarcoma.

Fig. 1 : Photograph of left palm showing growth in the hypothenar region

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Fig. 2 : Computed tomography scan of hand showing extent of the tumour with no bony erosion

Synovial sarcoma rarely occurs before 10 years. The most common site is in the region of the knee and distal portion of the thigh, followed by foot and ankle. It is rare in the hand. Till date only one case of synovial sarcoma of the palmar aspect of the left hand is reported [1].

Growth of the synovial sarcoma is very slow. The time elapsed between the first symptom and diagnosis generally ranges from 2 to 4 years. In this case it was 3 years. Unlike most other nonrhabdomyosarcoma soft tissue tumours of hand, synovial sarcoma frequently spreads to regional lymph nodes. However, axillary lymph nodes were free of metastasis in our case.

The synovial sarcoma is composed of two morphologically distinct types of cells that form a characteristic biphasic pattern. The biphasic synovial sarcoma includes epithelial cells with a surrounding spindle or fibrous component. The spindle cells stain positive for cytokeratin and epithelial membrane antigen (EMA). Vimentin is demonstrable in spindle cells but absent in epithelial cells.

The varied clinical presentation of synovial sarcoma of the hand may impede an early diagnosis. Furthermore, after the diagnosis has been confirmed, there is some difference of opinion as to the best method for staging these lesions and their treatment. The proposed current approach to the management of soft tissue sarcoma of hand includes a high index of suspicion that a mass in the hand may be a sarcoma, local staging with use of magnetic resonance imaging, distant staging with examination of regional lymph nodes and a computed tomography scan of the chest [2]. Wide local excision with at least a 2 cm margin is the treatment of choice. When this surgery is likely to result in significant functional disability then limited excision & adjuvant radiation is the alternative option. Amputation is the last option.

In our case we did wide local excision with sacrifice of both branches of the ulnar nerve. Since the deeper margins were positive histologically and malignancy was of high grade, radiotherapy (6600 cGy) was given. The favourable prognostic variables in this case are small tumour size (< 5 cm diameter), a primary site in hand, a younger age & a predominant epithelioid pattern.

In summary, because of rarity of synovial sarcoma, there are no clear guidelines on their management. A functional limb saving attitude without compromising the principles of malignancy should be individualized.

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