Case Report

Rhabdomyosarcoma : a rare case of rhabdomyosarcoma of alveolar type of hand in a 18 year old male

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Rhabdomyosarcoma(RMS) is a malignant tumor arising from skeletal tissue. It is the most common sarcoma of childhood comprising of 3-4% of childhood malignancies with 2/3rd of cases being reported in children below age of 10 years. Adult rhabdomyosarcoma is rare, and with involvement of hand is still rarer. In this present report we document the clinical, radiological and histopathological findings of a 18 year old male with rhabdomyosarcoma of hand whose FNAC & Incisional biopsy suggested Synovial sarcoma and MRI gave a provisional diagnosis of Pigmented Villonodular Synovitis. [*J Indian Med Assoc* 2019; **117:** 34-5]

***Key* words : Rhabdomyosarcoma, hand.**

Rhabdomyosarcoma (RMS) is a rare, highly malignant tumour of mesenchymal origin thought to arise from cells committed to a skeletal muscle origin occurring in the paediatric age group. It accounts for nearly 65% of all sarcomas in patients of 15 years of age and younger and approximately 7% of all childhood malignant solid tumours[1]. About 20% of the reported cases involve the extremities. It was an uncommonly entertained diagnosis in the hand until the publication of Potenza and Winslow in 1961[2].

These tumours have been classified into four subtypes on a histological basis: embryonal, alveolar, botryoid and pleomorphic[3]. Alveolar type is the most common type that involves the extremities and has got poorer prognosis. Intergroup Rhabdomyosarcoma Study Group (IRSG) has evolved a staging system for this tumour[4] which incorporates the elements of Tumour, Node and Metastases,as well as on the site of occurrence of the primary tumour. This is because the site plays a major determinant in the recurrence and the prognosis of the disease. Though these tumours apparently carry a poor prognosis, when it occurs in the hand it is quite possible to have a long-term disease-free state by a combination of early detection and appropriate treatment.

Case Report

A previously healthy 18 years old male presented to our OPD with a swelling over dorsal aspect of right hand (Fig 1), which had been increasing for 4 months. Swelling was associated with mild Pain particularly on extension of fingers with little impairment in flexion and extension of 4th phalanx.

Examinations — Examination of swelling confirmed a firm to hard mass of size 4.5×3.5×1 cm non adherent to skin extending from head of 4th metacarpal to distal carpal bones, it was fixed firmly to underlying structures the swelling was nontender non pulsatile and there was no local rise of temperature.

Investigations — His routine blood biochemical and urine examinations were found to be normal. X-Ray of hand revealed cortical erosion of the 4th metacarpal. FNAC of the swelling showed few large round cells with loose chromatin and inconspicuous nuclei, scanty cytoplasm and a few spindeloid cells giving a provisional histological diagnosis of a biphasic malignant tumor possibly a synovial cell sarcoma. MRI (Fig 2) hand showed a hyperintense to isointense mass encasing the 4th metacarpal with cortical irregularity giving a diagnostic possibility of giant cell tumor of tendon sheath possibly a Pigmented Villonodular Synovitis, Incisonal biopsy was performed which also gave a provisional diagnosis of synovial cell sarcoma. Chest X-Ray was found to be normal.

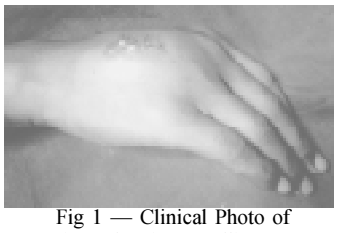

Fig 1 — Clinical Photo of Patient Showing Swelling Over Dorsal Aspect of Right Hand

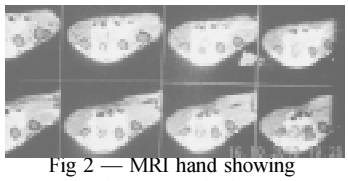

Fig 2 — MRI hand showing Hyperintense to isointense soft tissue mass lesion

Management — Thinking in line of management of synovial cell sarcoma wide margin excision by ray amputation was proposed to the patient. Necessary informed consent was taken from both patient and his parent and he was operated and mass was removed by wide margin excision by ray amputation in which 4th finger and entire metacarpal were removed (Fig 3) with transposition of 5th metacarpal to capitate. The mass as seen grossly during surgery was firmly adherent to the metacarpal and was not encapsulated the mass was sent for histopathology. Sections of mass with H/E stains with 5 x magnification showed small round blue cells with fibrovascular septa (Fig 4) and rhabdomyoblast seen with 40x magnification confirming diagnosis of RMS of alveolar type. The tumor was classified according to Intergroup RMS study Postsurgical pathological system as stage lla the patient was discharged on 1oth post operative day and currently is on chemotherapy.

Discussion

Malignant lesions of hand which arise primarily from tissues other than skin are rare and our knowledge is derived primarily from small series and case reports. In a review of 200 cases of sarcomas Hark and Cerney found that only 5% of them were rhabdomyosarcomas. This is apparently due to the fact that the skeletal muscle cells are completely differentiated and do not undergo cellular division in postnatal life as do those of other tissues[5].

Rhabdomyosarcoma in the hand is even rarer and the first major report was that of Potenza and Winslow. In the Clinico-pathological study by Linscheid of the rhabdomyosarcoma of the extremities and limb girdles at Mayo clinic of 87 cases of pleomorphic rhabdomyosarcoma he encountered only five tumours were located in

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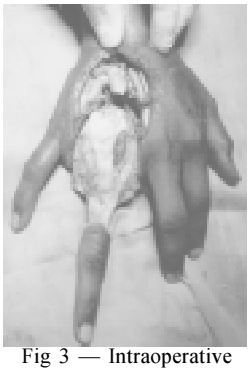

Fig 3 — Intraoperative Photograph

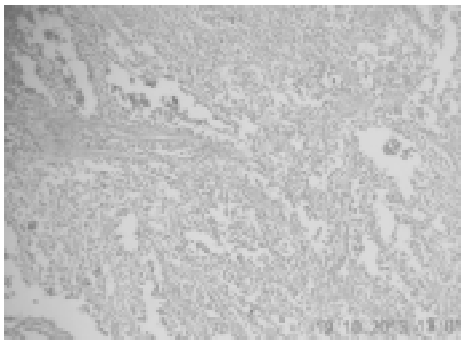

Fig 4 — H/P using H/E stain under light microscope with 5 x magnification showing small round blue cells with fibrovascular septa

the hand[6]. They advised radical excision as an important component of treatment. They also concluded that lesions of the upper extremity had better prognosis than the lesions of the lower extremity and more distal lesions had a better prognosis than the more proximal lesions.

Response to chemotherapy may be predicted by the identification of gene fusions and chromosomal rearrangement. In cases of rhabdomyosarcoma the presence of one or the other of the gene fusions has been shown to have prognostic significance, because they can distinguish between a very high risk subgroup (PAX3-FKHR) and a favourable outcome subgroup (PAX7-FKHR). Multivariate analysis demonstrated a significantly increased risk of failure (P=0.025) and death (P=0.019) in patients with metastatic disease if their tumours expressed PAX3-FKHR[7].

Prognostic factors include Clinical group,age,Stage and histologic subtype. Most common sites of metastasis include draining lymph nodes, Lungs & bone marrow.Many of the extremity tumours in the literature had metastasis at the time of initial presentation. They had a poor prognosis irrespective of the treatment modality. The subgroup of rhabdomyosarcoma primarily occurring in the hand could have a good prognosis because they can be detected early[8]. Surgical technique plays an important role.

For the tumours of the hand radical excision holds the best prospect of cure. This is preferred even at the cost of functional disability. Reconstructive surgery could be done later to enhance function. Radiotherapy after the amputation of the primary area has not found to be of significant use. All cases must have chemotherapy. The overall survival from this tumour is improving currently because of the use of therapeutic agents such as vincristine, actinomycin-D, cyclophosphamide and adriamycin plus cisplatinum, as well as external beam radiation therapy. For tumours locally controlled with surgical resection, five-year survival approaching 70% has been reported[9].

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