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Case Report

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Carpal tunnel syndrome due to malignant peripheral nervous system tumor; a rare entity

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Abstract

Carpal tunnel syndrome is a common disabling problem due to nerve entrapment. Though intrinsic risk factors aggravating this condition is well known malignant peripheral nervous system tumor as an entity responsible for this condition, to the best of our knowledge, is previously unrecorded. Pitfalls in the histological diagnosis of this condition are elaborated.

Keywords: Carpal Tunnel Syndrome, Malignant Tumor, Significance of pathological findings- Pit falls.

INTRODUCTION

Carpal tunnel syndrome (hence forth CTS) is an enigmatic disabling condition, widespread in western countries seen in 6% population in these countries and is responsible as one of the major causes leading to much abstenteeism in work related activities; particularly acute in those performing repeated movements with their hand and wrist. Carpal tunnel, a bony canal consisting of carpal ligament, contains nine flexor tendons and median nerve. This is a neuropathic condition caused by entrapment of this nerve; its function being delimited by carpal bones and ligament [1].

The main complaint for which our patient sought medical attention was pain in the hand, tingling and nunbness in the distribution of median nerve and reduced grip strength and functioning of the hand; similar to complaint usual in a case of this nature [2].

In spite of numerous papers elaborating on the various aspects of this disease proper, the most important cause responsible for this disease remains idiopathic. Though factors aggravating this condition are known fully, malignant peripheral nervous system tumor (hence forth MPNST) responsible as an intrinsic entity causing CTS is poorly documented. This paper highlights the link between this disorder and emphasizes the need for proper histological assessment.

CASE REPORT:

A 34-year-old male came to our medical attention for a complaint of inability to grasp and hold an object in his right palm. Earlier, on and off, he was suffering from pain and parasesthesia of hand; sometimes extending upto elbow even. On examination he had marked atrophy of thenar muscles and sensory loss. A marble sized swelling was noticed at the wrist which was fixed to deeper structures. A planned surgery revealed a tumor arising from median nerve which was fusiform in appearance. Microscopically the tumor showed alternating areas of hypo and hypercellularity with extensive foci of necrosis (Fig 1). Bundles of normal nerve elements coexisted with those of plexiform (Fig 2) features. There was a direct transition between a benign cellular tumor and sarcomatous growth. The sarcomatous cells were arranged in herring bone pattern along with foci of hemangiopericytoma and herniation of neoplastic cells into the walls of small blood vessels. Hypocellular areas showed uniform nuclei with buckled or wavy appearance (Fig 3). Since the diagnosis was obvious in H and E stain, special stains; in particular immunochemical stains were

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not needed for histological diagnosis.

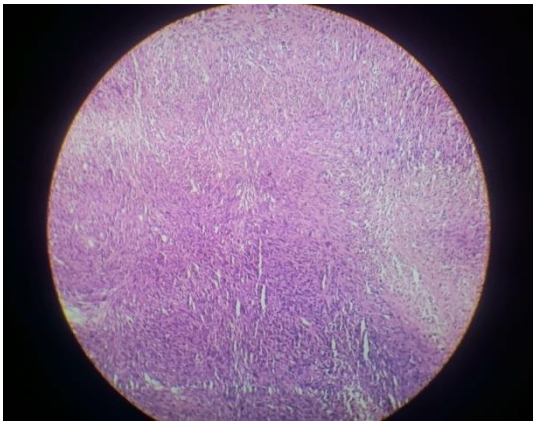

Figure 1: Sarcomatous tumor showing alternating areas of hypo and hypercellularity with extensive foci of necrosis

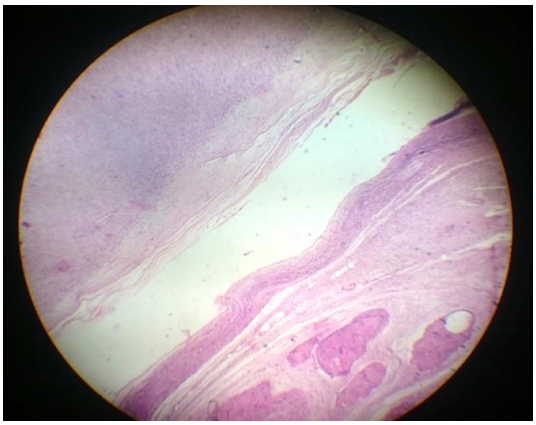

Figure 2: Sarcomatous tumor showing alternating areas of hypo and hypercellularity with extensive foci of necrosis

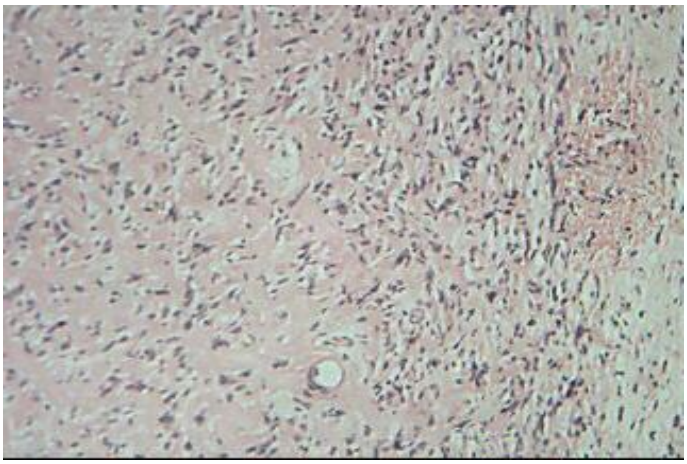

Figure 3: Sarcomatous tumor showing alternating areas of hypo and hypercellularity with extensive foci of necrosis

DISCUSSION

MPNST, commoner as soft tissue tumor, arises most often sporadically without preexisting neurofibroma (henceforth NF) or neurofibromatosis-1 (hence forth NF1). Occasionally less than 10% of such tumors take origin from preexisting NF or NF1 (characterized by a constellation of manifestation like café au lait spots; larger than 1.5 cms, one or more NF of any type, one or more Plexiform NF, freckling in axilla, optic glioma, two or more lisch nodules, dysplasia of sphenoid bone and a first degree relative with NF-1) [3]. The origin of the tumor under discussion is probably from preexisting plexiform NF; considered to be a precursor of malignancy. In this case, both classical and histological evidence for solitary NF and plexiform NF are present intimately mixed. While clinically it never poses any difficulty in diagnosing CTS, the herring bone pattern of MPNST, the predominant appearance of this tumor in this case, may lead to an alternate histological diagnosis of fibrosarcoma [4], which is almost an indolent tumor biologically unlike MPNST. The presence of marbellisation pattern, myxoid stroma, wavy buckled nuclei, of neoplastic cells, positive immunochemical reaction with S100, differentiates MPNST from fibrosarcoma; which is also clinically more distinct. Another sarcomatous tumor is synovial sarcoma with similar histological features and which arise in this region and require immunochemical means to differentiate between both; particularly when both show glandular differentiation. S-100 is usually negative in synovial sarcoma (hence forth SS) while in 50% of MPNST, it is positive; likewise, EMA, CEA are positive in synovial sarcoma. Glandular differentiation is well marked in SS while it is subtle and rare in MPNST. The presence of goblet cells in glands of MPNST is to be contrasted against SS wherein no such findings are recorded [5]. The predominance of NF differentiation, particularly in cellular type requires application of criteria postulated by late Scheithauer *et al* [6]. Atypical NF is often confused with MPNST and can be excluded because it lacks hypercellularity of the latter and pleomorphic nuclei, seen in this condition are scattered and degenerative.

Conflict of interests:

The authors declare that there is no conflict of interest regarding the publication of this paper.

CONCLUSION

Though surgery is often curative, cases of low grade MPNST as is the case in our patient requires careful monitoring for future recurrences.

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