Case Report

Malignant Peripheral Nerve Sheath Tumor

Rare Neurological Isolated Neoplasm

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ABSTRACT

Primary malignant schwannoma is a rare neoplasm of nerve sheath origin. It is a cancer of the connective tissue surrounding nerves. Given its origin and behavior it is classified as sarcoma. The estimated incidence of Malignant Peripheral Nerve Sheath Tumor (MPNST) in general population is 0.001% and in patients with Neurofibromatosis 1 (NF-1) is 2-5%. It is an uncommon spindle cell sarcoma accounting for approximately 5% of all soft-tissue sarcomas. They are highly aggressive and occur in the second or third decade. This neoplasm usually affects the extremities. There is strong association between MPNSTs and neurofibromatosis (NF-1) and previous irradiation. We present the case of a 61-year-old woman manifesting with recurrent sciatica near for the fourth and fifth lumbar vertebral bodies. She underwent resection of a mass at the L4-5 level that was subsequently recognized as a malignant peripheral nerve sheath tumor.

Keywords: Malignant Peripheral Nerve Sheath Tumor (MPNST); Neurofibromatosis; Protein S100

INTRODUCTION

Malignant Peripheral Nerve Sheath Tumor (MPNST) is extremely rare entity. However, it is of diagnostic importance because it is one of the most aggressive soft-tissue sarcomas. It is also called as neurofibrosarcoma as it arises from the neuroectodermal lining of the peripheral nerves. MPNSTs are often associated with NF1 in 30-50% and with previous irradiation in 4-11% while 0.001% arise de novo in the general population [1,2]. They often affect the head, trunk, and extremities; they lead to poor overall survival [3].

CASE REPORT

A 61-year-old female patient presented with history of persistent pain in the lowest segment of the lumbar region and sciatica since two years, without favourable evolution using medical treatment. Physical examination revealed a sensibility on her lumbar vertebrae near for the L4-L5. Nerve function and the remainder of general examination were normal without skin, skeletal or ocular anomalies. Computed Tomography (CT) scan showed a medullar necrotic rounded mass lesion at the L4-L5 level. Histopathological examination of the specimen revealed a tumor composed of spindle shaped cells with elongated hyperchromatic nuclei, arranged in long and short fascicles. Cells were arranged around hyaline bands in the form of cords and at places they were arranged in whorls (Figure 1). Some of the tumor cells had marked nuclear pleomorphism. The stroma showed extensive areas of necrosis and hemorrhage. The blood vessels showed hyalinized walls. Mitotic figures were more than five per high-power field (Figure 2). Immunohistochemically, the tumor cells showed diffuse and strong positivity for S-100 (Figure 3). However, they were negative for Pan-cytokeratin (AE1/ AE3), Desmin, and Human Melanoma Black (HMB) 45. Based on this data, a diagnosis of MPNST was made.

DISCUSSION

MPNSTs are malignant tumors developing from cells of peripheral nerve tissue [2]. They are also called neurofibrosarcomas, malignant schwannoma or neurogenic sarcoma and account for approximately 5-10% of all soft-tissue sarcomas and about one-fourth to one-half occur in the setting of NF-1 [4]. The MPNST is typically a disease of adult life, as most tumors occur in patients of 20-50 years of age [5]. It does not exhibit any sex differences [4]. The age of onset tends to be lower in cases associated with NF-1 [6].
CONCLUSION

Our case point out that in case of persistent pain and resistant to medical therapy, associated with alterations of sensitivity, neoplastic causes including the tumors of the nervous system should be considered. Although malignant medullary tumors are rare, MPNSTs should be included in the differential diagnosis of the nervous system tumors. Complete resection and administration of routine adjuvant chemotherapy are the treatment of choice.

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