Otolaryngology online journal

ISSN: 2250-0359 Volume 4 issue 1 2014

MALIGNANT PERIPHERAL NERVE SHEATH TUMOR OF THE NECK

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Abstract

Malignant peripheral nerve sheath tumors are highly aggressive tumors characterized by rapid growth with infiltration of surrounding tissue and hematogenous metastases. We described a 32-old male patient with malignant peripheral nerve sheath tumor of the head and neck. Surgery was the chosen method of treatment by the dissecting the mass. Even chemotherapy were administered after surgery, local recurrence occurred in the second month of the follow-up period. Then the patient underwent radical neck dissection under general anesthesia. Close follow-up was made, at first year disease had no recurrence. We reported this case and reviewed the literature regarding malignant peripheral nerve sheath tumor by reason of this case.

Introduction

Malignant peripheral nerve sheath tumor (MPNST) refers to any malignant tumor arising from major or peripheral nerve branches or sheath of peripheral nerve fibers of Schwann cell origin or pluripotent neural crest cell origin [1]. MPNST account for approximately 5-10% of all malignant soft-tissue tumors, of these only 8-15% occur in the head and neck [2,3]. The parotid area and infratemporal fossa were the commonest sites [4]. Surgical resection with adjuvant radiotherapy (ART) (even with clear resection margins) is the recommended treatment modality [4-7]. Because of the tumor's high rate of recurrence, radiotherapy is needed even when resection margins are clear. There is little reported in literature on these tumors and little information is available on the clinical management of MPNST occurring in the head and neck area.

Case report

32-old male patient presented with a right sided painfull cervical mass which had been growing gradually over the previous 3 months. Movements of his neck, motor and sensory function of his right arm were normal. We obtained magnetic resonance imaging (MRI), which showed that a well-defined, lobulated, enhancing mass had arisen from the right neck. (*figure 1*) The patient underwent complete excision of the cervical mass under general anesthesia. On gross examination, the tumor measured 4x4x3 cm. (*figure 2*) Histologic examination revealed that the tumor was largely made up of spindle cells that had a myxomatous appearance secondary to the presence of abundant intercellular connective tissue mucin. (*figure 3*) The spindle cells contained small hyperchromatic nuclei and long, wavy cytoplasmic processes. The histology report confirmed that the tumor arose from a large myelinated nerve and was made up of interlacing spindle cells set in a mucinous matrix. Focally, these cells coalesced to form acinar and cribriform patterns. Immunohistochemistry yielded a strong cytoplasmic staining for S-100 protein. (*Figure 4*) Ki 67 labeling index was over than 50%. (*Figure 5*) Vimentin was positive and CD34, Desmin, Aktin, CD117, BcL2, CD5, CD99 were negative. Based on these operative and histologic findings the tumor was diagnosed as MPNST. Following the surgery, chemotherapy (CT) were administered. He had local recurrence of tumor in the second month of the follow-up period despite postoperative CT. Then the patient underwent radical excision of the cervical mass under general anesthesia and received ART. The patient was without disease at first year.

Discussion

MPNSTs are seldom found in the head and neck area [6,7]. These tumors affect primarily the 30-50 year old age group, and occur with equal frequency in males and females. The estimated incidence of MPNST is 0.001% [1]. Ghosh et al [6] published one of the largest series of MPNSTs, involving 115 patients, of whom 14% presented with a head and neck tumor mass. Nthumba et al [8] have been reported 333 patients with nerve sheath tumors, of which 31 were MPNSTs and 2 of them localized in the head and neck region. The diagnosis is based on histology and confirmed by a positive reaction to immunohistochemical markers [9]. There is a wide histologic spectrum. The most common is that of a highly cellular, spindle cell tumor with a variable degree of nuclear pleomorphism. Immunophenotyping shows focal staining for S-100 and CD57.6 MPNST prognosis are extremely poor. Poor prognostic indicators are a lesion greater than 5 cm diameter, deep seated, high grade or recurrent [7]. Some authors believe that prognosis depends on histologic findings such as cellularity, pleomorphism and mitotic activity and to the size of the primary tumor [5,6,10]. Hruban et al [11] observed 43 cases and 54% of these cases had local recurrence 65% of them distant metastases to the lungs and bone, the five year survival rate is 34%. MPNST's infiltrate local tissue and spread periferentially [5]. Regional lymph node involvement is less than 1% of deep located disease [7]. The diagnosis is based on histology and confirmed by a positive reaction to immunohistochemical markers. Because of the great density of vital structures and unresectable areas in the head and neck, such as the skull base, internal carotid artery and cervical vertebrae, failure to perform adequate wide excision is common and generally associated with an unacceptably high local recurrence rate [3,12-14]. Even with negative surgical margins, up to 50% of MPNSTs recur locally, often on multiple occasions [3,14] This marked tendency for local recurrence has been attributed to the ability of this tumor to infiltrate surrounding tissues and invade perineural areas. Surgery with adjuvant radiotherapy is the choice of treatment for MPNST, requiring radical resection [4]. Because of the infiltrating character of the neoplasm and the close location to the major vessel and nerves, total surgical excision of the MPNSTs of the head and neck is very difficult [4,13,14]. But patients in whom a total tumor resection was impossible showed lower survival rates [13]. As the recurrence rates are very high and as effective adjuvant therapies is not work, to perform en bloc resection with tumor-free margins. Wide surgical excision is the only reliable treatment modality [13,14]. Because of the risk of early and late recurrences and second malignancies necessitates postoperative RT [2,4,5,7]. Basso-Ricci [15] observed that 14 of 25 (56%) patients were free of disease 3 years after combined therapy consisting of surgery and postoperative radiation. This percentage is higher compared with other series of patients who prevalently underwent only surgery [2,3]. CT remains controversial [2,4]. But some authors advocated adjuvant CT. After treatment, close follow-up is advised for these patients. Our patient had local recurrence of tumor in the second month of the follow-up period despite postoperative CT. Following recurrence he recieved adjuvant RT and was without disease at first year.

Conclusion

MPNST is a rare aggressive malignant tumor in the head and neck area, with a particularly high rate of local recurrence, although distant metastases can also occur.

The tendency to local recurrence contributes to the poor prognosis of the neoplasm, especially in the head and neck region. Free margin surgical resection is principally needed for definitive treatment.

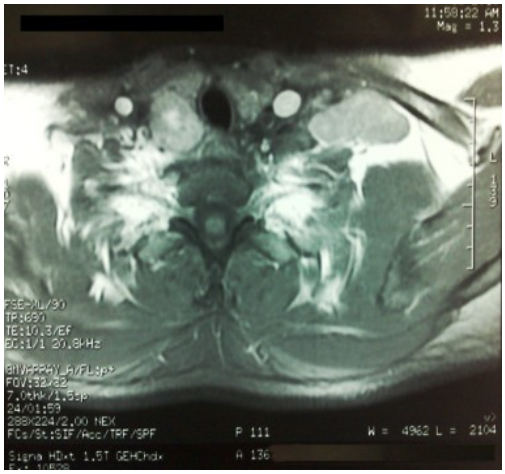

Figure.1: Axial section, magnetic resonance imaging of tumor

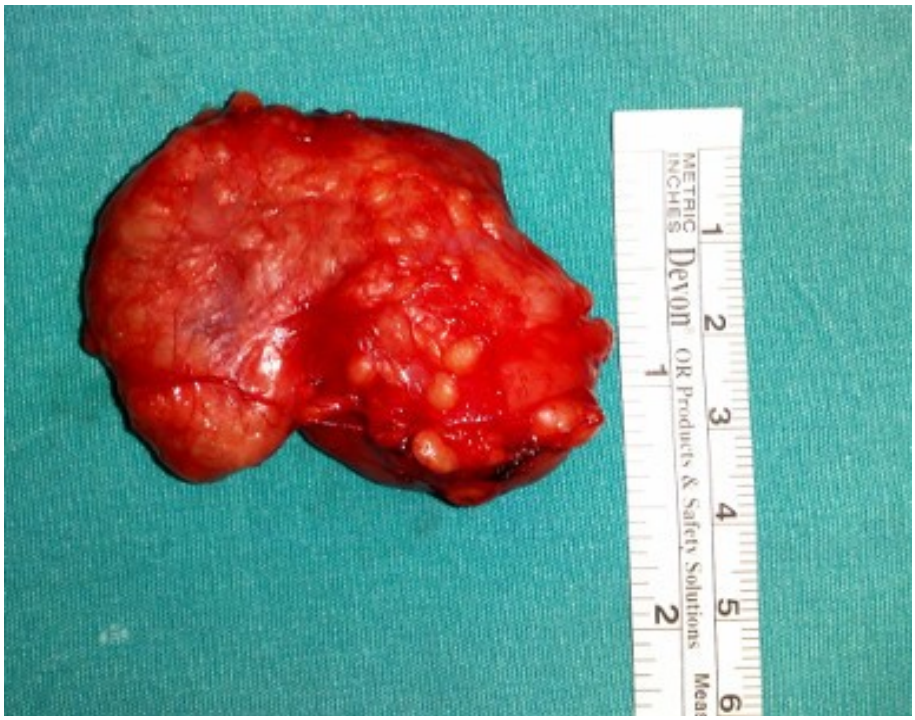

Figure.2: Macroscopic view of tumor

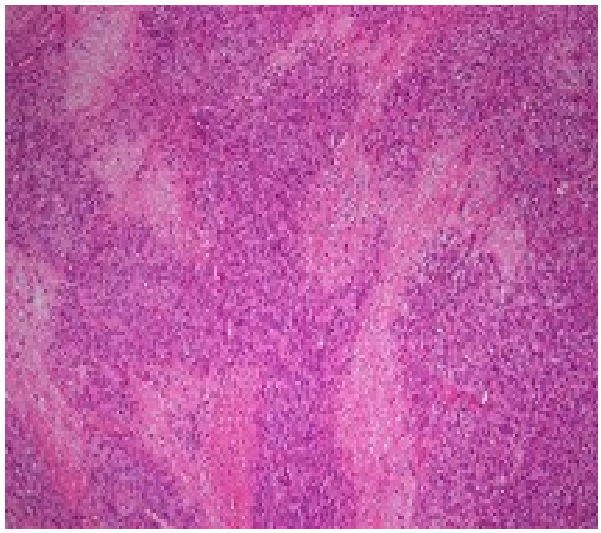

Figure.3: Geographic necrosis and cellular spindle cell areas are seen

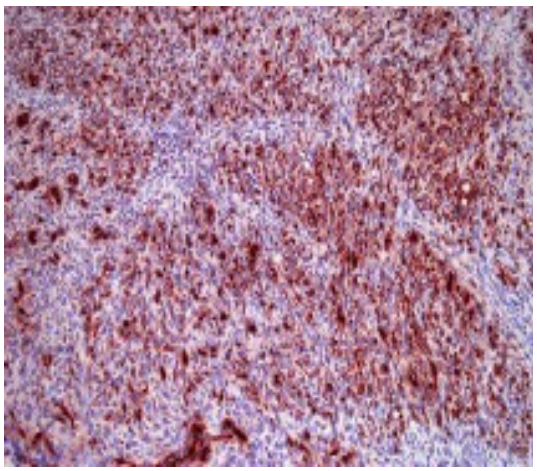

Figure.4: Focal areas showing strong immunoreactivity for S100 protein

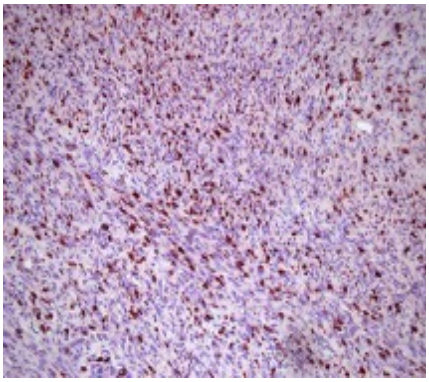

Figure.5: High Ki67 proliferatif activity was observed

References

1. Gupta G, Mammis A, Maniker A. Malignant peripheral nerve sheath tumours. Neurosurg Clin N Am. 2008;19:533-43.

2. Ducatman BS, Scheithauer BW, Piepgras DG, Reiman HM, Ilstrup DM. Malignant peripheral nerve sheath tumors. a clinicopathologic study of 120 cases. Cancer. 1986;57:2006-21.

3. Hoffmann DF, Everts EC, Smith JD, Kyriakopoulos DD, Kessler S. Malignant nerve sheath tumors of head and neck. Otolaryngol Head Neck Surg. 1999;99:309–19.

4. Wong WW, Hirose T, Scheithauer BW, Schild SE, Gunderson LL. Malignant peripheral nerve sheath tumor: analysis of treatment outcome. Int J Radiat Oncol Biol Phys. 1998;42:351-60.

5. Grobmyer SR, Reith JD, Shahlaee A, Bush CH, Hochwald SN. Malignant peripheral nerve sheath tumor: molecular pathogenesis and current management considerations. J Surg Oncol. 2008;97:340-9.

6. Ghosh BC, Ghosh L, Huvos AG, Fortner JG. Malignant schwannoma. A clinicopathologic study. Cancer. 1973;31:184-90.

7. Anghileri M, Miceli R, Fiore M, et al. Malignant peripheral nerve sheath tumors: prognostic factors and survival in a series of patients treated at a single institution. Cancer. 2006;107:1065–74.

8. Nthumba PM, Juma PI. Malignant peripheral nerve sheath tumors in Africa: a clinicopathological study. ISRN Surg. 2011; doi:10.5402/2011/526454

9. Cabibi D, Zerilli M, Caradonna G, Schillaci L, Belmonte B, Rodolico V. Diagnostic and prognostic value of CD10 in peripheral nerve sheath tumors. Anticancer Res. 2009;29:3149-55.

10. Kahraman A, Yildirim I, Okur E, Demirpolat G. Horner's syndrome from giant schwannoma of the cervical sympathetic chain: case report. B-ENT 2009;5:111-4.

11. Hruban RH, Shiu MH, Senie RT. Malignant peripheral nerve sheath tumors of the buttock and lower extremity; A study of 43 cases. Cancer. 1990;66:1253-65.

12. Valeyrie-Allanore L, Ismaili N, Bastuji-Garin S, et al. Symptoms associated with malignancy of peripheral nevre sheath tumours: a retrospective study of 69 patients with neurofibromatosis 1. Br J Dermatol. 2005;153:79-82.

13. Minovi A, Basten O, Hunter B, Draf W, Bockmuhl U. Malignant peripheral nerve sheath tumors of the head and neck: management of 10 cases and literature review. Head Neck. 2007;29:439-45.

14. Hujala K, Martikainen P, Minn H, Grenman R. Malignant nerve sheath tumours of the head and neck: four case studies and review of the literature. Eur Arch Otorhinolaryngol. 1993;250:379-82.

15. Basso-Ricci S. Therapy of malignant schwannomas: usefulness of an integrated radiologic surgical therapy. J Neurosurg Sci. 1989;33:253-7.