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Malignant peripheral nerve sheath tumour of the cervical symphathetic chain.

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Abstract : Malignant peripheral nerve sheath tumors (MPNSTs) are sarcomas which originate from peripheral nerves or from cells of the nerve sheath, such as Schwann cells, perineural cells, or fibroblasts. Upto 50 of MPNSTs occur in patients with NF1 demonstrating the tendency to arise from a preexisting neurofibroma. A case of left cervical symphatetic MPNST in an individual with neurofibromatosis presenting with Horner's syndrome was treated sucessfully by surgical excision and adjuvant radiation.

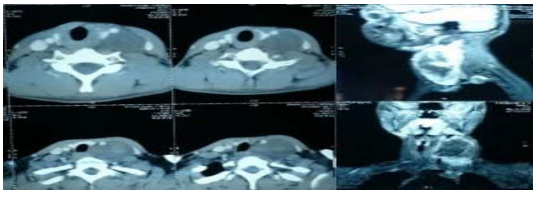
Keyword : Malignant peripheral nerve sheath tumors, neurofibromatosis, Horner's syndrome

INTRODUCTION

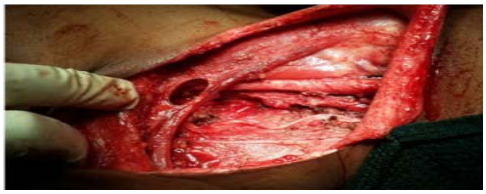
Malignant peripheral nerve sheath tumors (MPNSTs) are sarcomas which originate from peripheral nerves or from cells of the nerve sheath, such as Schwann cells, perineural cells, or fibroblasts. Upto 50 of MPNSTs occur in patients with NF1 demonstrating the tendency to arise from a preexisting neurofibroma. A case of left cervical symphatetic MPNST in an individual with neurofibromatosis presenting with Horner's syndrome was treated sucessfully by surgical excision and adjuvant radiation.

CASE REPORT

A 14-years-old boy presented with an asymptomatic left neck mass which was present since childhood but rapidly increased in size in the last two months. General examination revealed Café au-lait spots, neurofibromas, left Horner's syndrome, adenoid enlargement and high arched palate. Neck examination showed a 11*5cm globular mass deep to the left sternomastoid, gross rightsided tracheal deviation and anterior displacement of the left carotid artery. A transverse scar indicating previous biopsy was present over the swelling.



CT neck showed a 13*5*3cm multilobulated non-enhancing soft tissue density extending from C2 to C7. MRI revealed a mixed intensity lesion splaying the left common carotid artery, displacing left IJV and compressing the left lateral pharyngeal wall. Incision biopsy revealed a low-grade spindle-cell neoplasm of neural origin.IHC revealed S-100 positivity. Hence a diagnosis of malignant peripheral nerve sheath tumour of cervical sympathetic chain was made.



Surgery was proceeded by a horizontal skin incision and the mass was excised en bloc with the sternomastoid and the IJV. After the surgery the patient made uneventful recovery with little improvement in Horner's syndrome. HPE showed a low grade (1 mitotic figure/HPF) spindle-cell tumor consistent with MPNST,IHC revealed S-100 positivity. All margins were free from the tumour. The patient was administered adjuvant radiation (IMRT) and is under periodical follow-up

DISCUSSION

A sarcoma is defined as a MPNST when at least one of the following criteria is met:

1. It arises from a peripheral nerve
2. It arises from a preexisting benign nerve sheath tumor (neurofibroma)
3. It demonstrates schwann cell differentiation on histologic examination MPNSTs comprise approximately 5-10% of all soft

tissue sarcomas. They can occur either spontaneously or in association with neurofibromatosis-1 (NF1).[2] MPNSTs usually present as an enlarging palpable mass. Pain is a variable complaint. Rapid enlargement occurs more often in the setting of NF1 and should raise concern for malignant degeneration of a neurofibroma. MPNSTs arising from peripheral nerves may result in a variety of clinical patterns, including radicular pain, and motor weakness. Most MPNSTs occur in conjunction with large peripheral nerves such as the sciatic nerve, the brachial plexus and the sacral plexus. They are usually deep-seated and often involve the proximal upper and lower extremities as well as the trunk. Dermal or flat plexiform neurofibromas, commonly encountered in cases of NF-1, have not been shown to undergo malignant transformation and do not usually require close monitoring. On the other hand, larger nodular tumors associated with large peripheral nerves and deep extensive plexiform neurofibromas do have the potential to undergo malignant transformation and should be observed more diligently. In rare instances, multiple MPNSTs can arise in the setting of NF1. Most of these tumors are considered high-grade sarcomas with the potential to recur as well as to metastasize.[12] Magnetic resonance imaging (MRI) is the imaging modality of choice. To some extent, MPNSTs share basic imaging characteristics with their benign counterparts such as neurofibromas and schwannomas. These include a fusiform shape and a longitudinal orientation in the direction of the nerve. However, some distinctions are noteworthy. Large tumors (> 5 cm), invasion of fat planes, heterogeneity, ill-defined margins, and edema surrounding the lesion are more suggestive of MPNSTs .[3] Imaging studies of the chest are an important part of any initial sarcoma evaluation. MPNSTs are most likely to metastasize to the lungs, followed by the bone and finally the pleura. For this reason, a CT of the chest is the preferred imaging study to screen for distant disease. A bone scan should also be obtained to help identify metastatic bone disease.[14] FDG PET is a dynamic imaging modality which evaluates metabolic activity by quantitatively assessing intracellular glucose use. It has been shown to reliably identify areas of increased metabolic activity such as those seen in malignancies.[15] In MPNST, staging is dependent upon histologic grade, tumor size, tumor depth, and the presence or absence of metastases. In the absence of detectable metastases, histologic grade, tumor size, and tumor depth are the strongest predictors of eventual metastases. The stage is based upon imaging studies, which demonstrate the local and distant extent of the disease, and upon the histologic grade, which describes the histological characteristics of individual tumor cells.[4]

Table 1: The American Joint Committee on Cancer (AJCC) Staging System for Soft Tissue Sarcoma, 6th Edition

Stage	Size	Depth	Grade	Metastases
I	Any	Any	Low	No
II	< 5cm, any depth OR > 5cm	Superficial	High	No
III	> 5cm	Deep	High	No
IV	Any	Any	Any	Yes

A biopsy is an integral part of the staging system. It offers both a histologic tissue diagnosis and the ability to determine the grade of the lesion. This information, in turn, permits adequate planning and adjuvant treatment such as radiation or chemotherapy. In addition, this information is incorporated into the tumor staging process which provides prognostic information with regard to the disease and treatment generalizations. FNAs is a biopsy method employed to obtain individual cells for cytologic review. It can be done with a very small needle which is more easily tolerated by the patient and is often useful to establish the presence of malignant cells. However, it is not large enough to demonstrate the architectural pattern within a tumor and for this reason is not often used to make an initial diagnosis. In cases of established diagnoses, such as after surgical resection of a tumor, FNA can often be successfully used to sample tissue which is suspected to be recurrent disease. A second type of biopsy is a core needle or tru-cut needle biopsy, which uses a larger hollow-bored needle gauge to obtain a more substantial tissue sample. This type of sample offers inspection of both individual cells as well as the architectural arrangement of those cells within a given part of the tumor mass. This information is often important in establishing a histopathologic diagnosis. In many tertiary care cancer centers, core needle biopsies are often performed with either CT or ultrasound image guidance. This is an outpatient procedure and it allows for adequate tissue sampling while minimizing bleeding and minimizing contamination or seeding of surround tissue with tumor cells. In addition, it often avoids the need for general anesthesia. In some cases a formal open biopsy is required. This can either be an incisional biopsy, where a small piece of tissue is removed from the larger tumor mass, or an excisional biopsy, in which case the entire tumor is removed. In general, an incisional biopsy is recommended when a sarcoma is suspected. The general appearance of MPNSTs is one of dense cellular fascicles which alternate with myxoid regions. This swirling arrangement of intermixed dense and myxoid areas has been described as a marbleized pattern. The cells may be spindle shaped with very irregular contours. Alternatively, cells may be rounded or fusiform in shape. Nuclear palisading has also been shown but in less than 10% of cases and even then, only focally. Malignancy is suggested by features such as invasion of surrounding tissues, invasion of vascular structures, nuclear pleomorphism, necrosis, and mitotic activity. S-100 has been identified in approximately 50 – 90% of MPNSTs, however the staining pattern has been noted to be both focal and limited to few cells. Leu-7 and myelin basic protein are noted in 50% and 40% of cases respectively. [7] In general, a combination of antigens is used to help exclude other spindle cell lesions and to confirm the diagnosis of MPNST.

Surgical Treatment for MPNST

The mainstay of treatment is surgical resection. The goal of the operation is to achieve complete surgical excision of the tumor with negative (wide) margins. This offers the best outcome with respect to both local recurrence and distant metastases.[8] Radiation therapy has become an integral part of local disease control in most soft tissue sarcomas and likewise can be employed in pre-operative, intraoperative, and post-operative settings for MPNST. Together with wide surgical excision, radiation therapy offers local and overall survival rates which are similar to those following amputation, and the combined modality treatment often allows patients the option to undergo successful limb-salvage surgery. Chemotherapy is intended for systemic disease which is either too small to detect or too diffuse, rendering local treatment techniques ineffective. The use of chemotherapy is only employed in highgrade disease, in which metastatic disease is likely. The benefits of chemotherapy must be weighed against its side-effects, some of which are irreversible. For this reason, the decision to treat with chemotherapy is somewhat tailored to an individual patient and his or her individual disease. Large, deep, high grade tumors and tumors which demonstrate metastases or metastatic potential are typical indications for chemotherapy treatment.[9] Recurrence can be discussed in terms of local disease and distant or metastatic disease. The local

recurrence rate for MPNSTs has historically been reported to range from 40-65% and the distant recurrence rate has similarly been reported to range from 40-68%. Five-year survival has been reported to range from 16-52%. Longer survival has been correlated with complete surgical excision, small tumor size (<5 cm), and the presence of a low grade component.[10]

Conclusion

Cervical MPSNTs are uncommon, especially those arising from the cervical sympathetic chain, with less than 60 cases reported in the iterature. Surgery remains the mainstay of treatment along with adjuvant radiation. Genetic counselling is the best possible way in decreasing the incidence of MPNST in posterity especially in neurofibromatosis patients.

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