Case Report
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Spindle Cell Sarcoma of the Vagus Nerve Presenting as a Neck Lump: A Case Report with Literature Review

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Abstract

Malignant Peripheral Nerve Sheath Tumour (MPNST) is a rare soft tissue neoplasm of the head and neck. This case report describes the presentation and management of a young girl with MPNST. Initially the patient received chemotherapy followed by radical neck dissection. This case study confirms that a combined-modality approach with complete surgical resection and neoadjuvant radiotherapy leads to improved outcomes in MPNSTs of the head and neck. However, more research needs to be done to explore newer treatments.

Keywords: MPNST; Sarcoma; Malignant; Peripheral nerve; Sheath; Tumours

Introduction

Malignant peripheral nerve sheath tumour (MPNST) is an aggressive mesenchymal tissue neoplasm found during childhood and adolescent phase. MPNSTs account for 5-10% of all soft tissue sarcomas, arising from peripheral nerves or cells of the peripheral nerve sheath such as Schwann cells, fibroblasts and perineural cells [1]. These cells have specific morphological features which distinguish them from one another. However, MPNST histology is very different from the others and there are only few reports that show MPNSTs have primitive neuroepithelial differentiation thus making it challenging to diagnose. In addition, few studies reveal Ewing's Sarcoma (ES) / Primitive Neuroectodermal Tumours mimic MPNST, which should be considered as differential diagnosis. To distinguish between these tumours histochemical stains are used, however these are not always reliable and result in different sensitivities and specificities, hence making it difficult to reach a diagnosis [2].

Furthermore, MPNST can either occur spontaneously or in association with Neurofibromatosis-1 (NF1). Because of the association of MPNST with von Recklinghausen's neurofibromatosis, a careful workup and family history should be obtained for genetic counselling and prognosis. However, scientific literature on these tumours is limited to small studies and case reports, thus more research needs to be done.

We present a case of spindle cell sarcoma with Peripheral nerve sheath malignant tumour.

Case History

A 16 years old girl presented with a 2 months history of a neck lump over left sternocleidomastoid muscle. On examination she was found to have Horner's syndrome, café-au-lait spots and axillary and inguinal freckling. Ultrasound scan of her neck lump suggested it to be an enlarged lymph node measuring 3x4x7cm. The Fine Needle Aspiration (FNA) cytology of this neck lump was non-diagnostic and an excision biopsy was carried out. The biopsy revealed a high-grade spindle cell tumour, likely to be a sarcoma possibly of neural origin. The chest x-ray was normal and the Computed Tomographic (CT) scan showed a large left sided soft tissue mass extending from skull base to superior mediastinum but no metastasis. The Chromosome studies revealed normal female phenotype 46XX from the tumour. Initial chemotherapy with six cycles of Vincristine, Ifosfamide, Doxorubicin and Etoposide (VIDE) was unsuccessful. This was confirmed by magnetic resonance imaging scan, which showed the lesion still extending from skull base to clavicle on the left side. The patient underwent left radical neck dissection. Intra-operatively, the tumour was seen arising from the vagus nerve. It was encasing the carotid artery and sympathetic chain. The common, internal and external carotid arteries were freed and preserved. The sympathetic chain, vagus and the spinal accessory nerves were removed enblock with the tumour. She also underwent Left true vocal cord Bioplastique injection. The histology showed spindle cell sarcoma with variable cellularity and

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hemangiopericytomatous vascular pattern. The nuclei were large round to oval and some had irregular buckled shape with numerous mitotic figures. Immuno staining was positive for BCL2 and focal staining for S 100. Desmin, Myf4, Cam 5.2, EMA, CD45, ALK1 were negative. CD99 shows cytoplasmic staining. A diagnosis of spindle cell sarcoma with Peripheral nerve sheath malignant tumour was made. Following radiotherapy, the patient continued to attend full time school. She needed to be fed by gastrostomy tube. The patient was discharged for long term follow up in the combined paediatric oncology multidisciplinary clinic.

Discussion and Conclusion

MPNST tumour is a rare childhood and adolescent sarcoma. Paediatric soft tissue sarcoma is about 7% of childhood tumors [3]. These tumours usually occur in young adults from a previously recognized neurofibroma [4]. The mean age of disease occurrence was between 29-36 years in patients with Neurofibromatosis Type I, whereas patients without Neurofibromatosis Type I tend to present in the 7th decade of life. The incidence of MPNST in patients with NF-1 is 4%-13% [4-6]. A patient with NF-1 with unresolved complaints need to be investigated due to risk of malignancy [7]. Ewing's sarcoma originating from the vagus nerve is a rare finding in teenagers but should be considered as a differential diagnosis for a neck lump.

Ewing 's Sarcoma and Peripheral Primitive Neuroectodermal Tumour (PPNET) are now considered to be in the same spectrum of malignancy The Ewing's Sarcoma Family of Tumours (ESFTs). These tumours are now known to be of neural crest origin. The chromosomal translocation [t(11;22)(q24;q12)] is present in 85% of ESFTs. ESFTs are most commonly developed in the second decade of life and show a slight male predominance [8,9]. To diagnose MPNST, ultrasound scan of neck must be done as well as chest x-ray and CT scan. Furthermore, FNA can be carried out, followed by biopsy to confirm the diagnosis. Therefore, to accurately diagnose a patient, clinical, pathological, radiological and genetic testing should be performed. Cytokeratin, S100 and CD99 stains have been associated with synovial sarcoma, MPNST and ES respectively. However, these markers are not solely reliable diagnostic tools as they have limitations in specificity and sensitivity. A study of 16 MPNST carried out by Zhu et al revealed that 75% of the samples had positive S100 staining [10]. In another study, Oslen et al., showed significant overlap between S100 and CD99, where 22 antibodies were stained in synovial sarcoma (n=23), ES (n=27) and MPNST (n=23). The cytosolic CD99 stained positive in 70% of synovial sarcomas, 93% positive in ES and only 43% positive in MPNST. Whereas membranous CD99 staining was more specific and had 85% specificity and 78% sensitivity to ES, 26% specificity for synovial sarcoma however it did not stain for MPNST. Therefore, membranous CD99 can be used to distinguish between ES and MPNST [11]. MPNSTs have always been difficult to treat. This is due to their inherently aggressive nature; however, limitations in both therapeutic and diagnostic techniques have played an important part as well. Nevertheless, mainstay of treatment is surgical resection and neoadjuvant radiotherapy. These patients need a long term follow up to monitor for any recurrences.

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