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PLEXIFORM NEUROFIBROMA OF THE BRACHIAL PLEXUS – A RARE CASE REPORT

Plastic Surgery

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ABSTRACT

Neurofibroma is a common benign tumour occurring as part of an autosomal dominant disorder, neurofibromatosis type 1, leading to the formation of benign tumours or neurofibromas of the peripheral nervous system. Large neurofibromas of the brachial plexus are rare and present a difficult challenge for surgeon due to the anatomical complexity of the brachial plexus. Dermal neurofibromas usually present with swelling and occasional pain, but neurofibromas associated with the brachial plexus present with pain and neurological symptoms. These plexiform neurofibromas of the brachial plexus are known to undergo malignant transformation. Here, we present a case of a large plexiform neurofibroma affecting the left brachial plexus and extending till the elbow, confirmed with MRI and surgical debulking was done.

KEYWORDS

Brachial plexus, Tumours, Rare, Surgery, Complications

INTRODUCTION

Peripheral nerve tumours are a rare heterogeneous group of mostly benign tumours that can be divided into 2 groups, peripheral nerve sheath and non-neural sheath tumours. Schwannomas and neurofibromas are the most common tumours of the brachial plexus region, and in most series in the literature neurofibroma is more prevalent.[1-3] Peripheral non-neural sheath brachial plexus tumours are even rarer than tumours of neural origin and comprise a wide variety of benign and malignant tumours and tumour-like conditions.[2,4] Neurofibromatosis type 1 (NF-1) is one the most common autosomal dominant conditions affecting the nervous system.[5] Plexiform neurofibroma (PN) is a benign tumour that basically defines the diagnosis of NF-1. These entities consist of multiple, twisted masses that grow along the axis of a large nerve, infiltrating and separating normal nerve fascicles, composed of a mix of Schwann cells, perineurial-like cells, and fibroblasts, interspersed with nerve fibres, wire-like strands of collagen, and a myxoid matrix.[6] Tumours of the brachial plexus comprise less than 5% of tumours of the hand and upper extremities.[4] Malignant transformation is the main associated complication. The management of brachial plexus tumours requires a thorough understanding of the complex anatomy, the clinical presentation, pathologic variation, and surgical techniques for operation.[4] Surgery intervention is required when there is functional compromise, aesthetic deformity or pain.[7]

Case Report

19 year old female presented to our department with pain along the left upper limb since 2 months. She was apparently normal 2 months ago when she developed a constant dull aching non-radiating pain along the inner aspect of the left upper limb. She was able to do her daily activities but due to the increased pain, they were hindered. There was no history of weakness or swelling in the left shoulder or hand. A clinical diagnosis of a neuropathic pain was made and investigated with an MRI which showed a lesion of the left brachial plexus from roots to left elbow region which was homogeneous hypointense in T1 weighted but hyperintense in T2 wieghted images suggestive of a nerve sheath tumour. **(Fig. 1)** We planned for exploration. Under general anaesthesia and loupe magnification, a lazy 'S' incision was made in medial aspect of the proximal two thirds of the arm and deepened in layers. **(Fig. 2)** The lobulated lesion with nerve identified, debulked and sent for histopathology with the nerves intact. **(Fig. 3 & 4)** Haemostasis was secured and incision closed in layers with 3-0 polyglactin and skin staples over a suction drain. Post-operative period was uneventful with pain reduced. Histopathology revealed a plexiform neurofibroma with no evidence of malignancy.

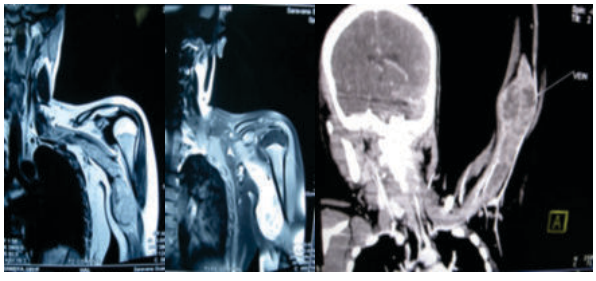

Fig. 1 – MRI Picture Showing The Lesion Involving The Left Brachial Plexus

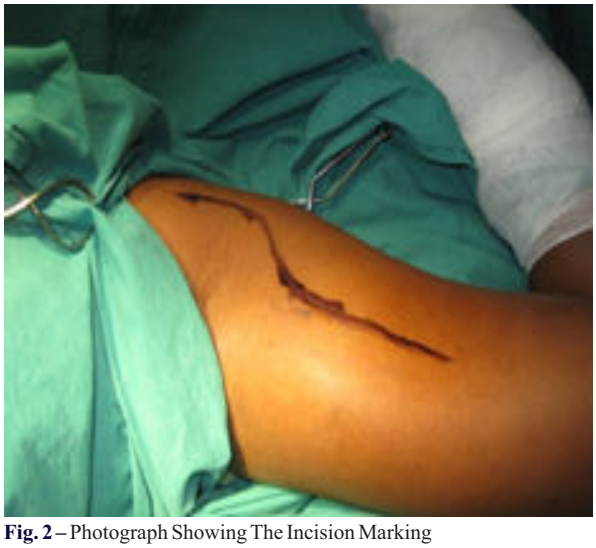

Fig. 2 – Photograph Showing The Incision Marking

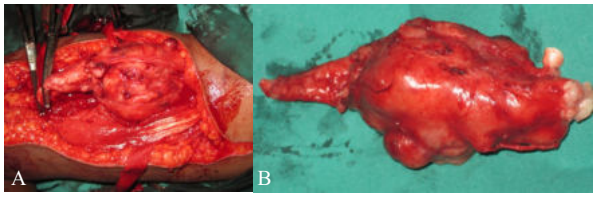

Fig. 3 – Photograph Of The Lesion In Situ (a) And After Excision (b)

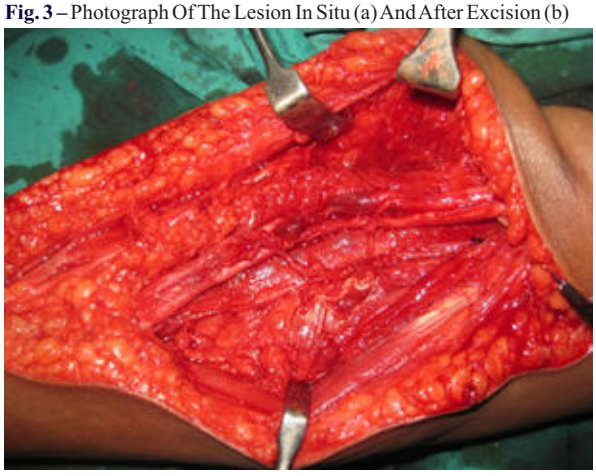

Fig. 4 – Photograph After Excision With Intact Nerves

DISCUSSION

Tumours of the brachial plexus are usually rare and present a clinical challenge for surgeons.[8] Tumours derived from perineural Schwann cells include schwannomas, neurofibromas, and malignant peripheral nerve sheath tumours. Secondary tumours involving peripheral nerves include locally invasive or metastatic carcinoma, lymphoma, and sarcoma.[9] The majority of them are solitary (up to 90 %) and not associated with neurofibromatosis type 1.[6] Plexiform neurofibroma is a common manifestation of NF-1. This entity may be classified as a benign peripheral nerve sheath tumour that involves multiple nerve fascicles or branches of major nerves.[10] In 1970 Dart et al. reported 27 cases of plexus tumors, 22 of which were nerve sheath tumours.[1] Symptoms and signs of peripheral nerve tumors are caused by direct nerve invasion, involvement of surrounding tissues, or mass effects and usually present with a painless palpable mass, local or radiating pain, motor loss, sensory loss, paresthesias, and dyspnea.[9] According to Huang et al, the most common presenting symptom is pain (70%), followed by sensory change (61%) and motor loss (52%).[4] In a study by Kehoe et al. on 104 patients who presented with a solitary benign NST, 15 of which involved the brachial plexus.[11] The primary complaint of most of them was of a palpable mass with less than half presenting with local pain or paresthesia. Siqueira et al. in a consecutive series of 18 patients who presented with brachial plexus tumours and tumour-like conditions, reported that the most common presenting symptom was mass (75%), followed by pain (58%), and sensory change (41%).[12] The duration and progression of symptoms or signs are important, as most benign tumours have a longer duration and a slow rate of progression, while malignant tumours tend to progress rapidly in size, pain, and neurologic deficits.[13] Hence, a detailed history and thorough physical examination are necessary for the diagnosis and management of brachial plexus tumours. MRI with contrast is the investigation of choice in brachial plexus lesions, to delineate the margins of the tumour from surrounding tissues.[14] According to Go et al., brachial plexus tumors show a well-defined mass with the long axis in line with the nerve of origin and a homogeneous intermediate signal intensity on T1-weighted sequences.[15] The tumours were hyperintense on T2 weighted images with a homogenous central low signal intensity (target sign) and strong enhancement following contrast administration and this sign is due to the central fibrous tissue surrounded by myxoid tissue.[15,16] The presence of the target sign does not differentiate a neurofibroma from a schwannoma.[17] They can be distinguished by determining the relationship of the tumour to the nerve of origin. In the case of a schwannoma, the nerve has an asymmetric relationship with the tumour, whereas the nerve passes through the center of a neurofibroma or is obliterated by it.[18] Fine needle aspiration cytology or biopsies are seldom performed due to the complexity of brachial plexus, possible damage to the intact fascicle or risk of hemorrhage.[4] Malignant transformation is the leading complication of plexiform neurofibromas. The lifetime risk of this transformation is estimated at 15 to 20% in patients with NF-1.[19] The clinical signs of malignant degeneration include the sudden onset of consistent pain, neurological deficit and an increase in the tumour size. Hosoi stated that MPNST develops in approximately 13% of patients with von Recklinghausen disease.[12] Brasfield and Das Gupta reported an overall incidence of 29% in a large series of patients.[8] MPNST is generally considered chemotherapy resistant tumours and the role of radiation therapy is not completely defined. However, post-operative radiotherapy is currently recommended by oncology consensus as part of standard treatment guidelines for these tumours, despite clear surgical margins.[20] The treatment of plexiform neurofibromas in patients with NF-1 consists of regular follow-up examinations involving serial physical evaluations and neuroimaging studies.[7] The choice of surgical approach is decided according to the size and location of the tumour and its relationship with adjacent structures. An anterior supraclavicular approach has been used to treat most tumours involving the roots and trunks.[4] The infraclavicular approach has been used for lesions involving the lower plexus root or trunk involved by the tumor, and patients who have had a history of radiotherapy or resection.[8] For tumours involving both the supraclavicular and infraclavicular portions of the brachial plexus, the combined or transclavicular approaches can be used.[4] The proximal and distal aspects of the tumour are identified, and the tumor is isolated from the surrounding neurovascular structures and excised or debulked.[8] Intra-operative monitoring of nerve and muscle action potentials play a useful role in the surgical management of tumours of the brachial plexus region.[4] The success rate for the surgical resection of brachial plexus tumours is high but the rate of complications are also increased. Paresthesias, weakness, and postoperative pain generally recover with time and physical therapy. A common complication of the supraclavicular approach for tumor resection is phrenic nerve paralysis, which usually improves with time.[4] Artico reported 11 tumours of the brachial plexus among the 119 peripheral nerve sheath tumors studied. In 93% of their patients with schwannomas and 81% of their patients with neurofibromas, motor function stabilized or improved postoperatively. The best surgical results were observed in the patients with schwannoma, whereas the worst results were demonstrated in those with plexiform neurofibroma of a 6-year follow-up period.[21]

CONCLUSION

The brachial plexus is involved in many tumors and tumor-like conditions of which nerve sheath tumours are common. Surgical resection of tumor is the treatment of choice in the most of the cases. The post-operative outcome is influenced by the extent of resection at surgery and pathological features of tumor. It also depends on the intraoperative microsurgical dissection technique with intraoperative monitoring, thus improving patients' outcome. In malignant tumors, surgery should be followed by adjuvant chemotherapy or radiation therapy to alleviate presenting symptom and reduction of tumor volume.

REFERENCES

1. Dart LH, Jr, MacCarty CS, Love JG, Dockerty MB. Neoplasms of the brachial plexus. Minn Med. 1970; 53: 959–964.
2. Kim DH, Murovic JA, Tiel RL, Moes G, Kline DG. A series of 146 peripheral non-neural sheath nerve tumors : 30-year experience at Louisiana State University Health Sciences Center. J Neurosurg. 2005; 102: 256–266.
3. Lusk MD, Kline DG, Garcia CA. Tumors of the brachial plexus. Neurosurgery. 1987; 21: 439-453.
4. Huang JH, Zaghloul K, Zager EL. Surgical management of brachial plexus region tumors. Surg Neurol. 2004; 61: 372–378.
5. Williams VC, Lucas J, Babcock MA, et al. Neurofibromatosis Type I Revisited. Pediatrics 2009; 133: 123-124
6. Pilavaki M, Chourmouzi D, Kiziridou A, Skordalaki A, Zarampoukas T, Drevelengas A: Imaging of peripheral nerve sheath tumors with pathologic correlation: pictorial review. Eur J Radiol. 2004; 52: 229–239.
7. Serletis D, Parkin P, Bouffet E, et al. Massive plexiform neurofibromas in childhood: natural history and management issues. J Neurosurg 2007; 106: 363-367
8. Das S, Ganju A, Tiel RL, Kline DG: Tumors of the brachial plexus. Neurosurg Focus 22: E26. 2007
9. Mrugala MM, Batchelor TT, Plotkin SR: Peripheral and cranial nerve sheath tumors. Curr Opin Neurol 18: 604–610. 2005.
10. Al-Otibi M, Rutka JT. Neurosurgical implications of neurofibromatosis type I in children. Neurosurg Focus 2006; 19: E2
11. Kehoe NJ, Reid RP, Semple JC. Solitary benign peripheral-nerve tumours. Review of 32 years' experience. J Bone Joint Surg Br. 1995; 77: 497–500.
12. Siqueira MG, Martins RS, Teixeira MJ. Management of brachial plexus region tumours and tumour-like conditions : relevant diagnostic and surgical features in a consecutive series of eighteen patients. Acta Neurochir (Wien) 2009; 151: 1089–1098.
13. Baehring JM, Betensky RA, Batchelor TT: Malignant peripheral nerve sheath tumor: the clinical spectrum and outcome of treatment. Neurology 61: 696–698. 2003.
14. Binder DK, Smith JS, Barbaro NM: Primary brachial plexus tumors: imaging, surgical, and pathological findings in 25 patients. Neurosurg Focus 16: E11. 2004.
15. Go MH, Kim SH, Cho KH: Brachial plexus tumors in a consecutive series of twenty one

patients. J Korean Neurosurg Soc 52: 138–143. 2012
16. Krandsdorf MJ, Murphey MD. Neurogenic tumors. In: Krandsdorf MJ (Ed). Imaging of soft tissue tumors. 2nd ed. Philadelphia, USA: Lippincott Williams & Wilkins, 2006: 335-348.
17. Varma DG, Moulopoulos A, Sara AS, Leeds N, Kumar R, Kim EE, et al: MR imaging of extracranial nerve sheath tumors. J Comput Assist Tomogr 16: 448–453. 1992
18. Saifuddin A: Imaging tumours of the brachial plexus. Skeletal Radiol 32: 375–387. 2003
19. 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: 1065-1074.
20. Wanebo JE, Malik JM, VandenBerg SR, Wanebo HJ, Driesen N, Persing JA. Malignant peripheral nerve sheath tumors. A clinicopathologic study of 28 cases. Cancer. 1993; 71: 1247–1253.
21. Artico M, Cervoni L, Wierzbicki V, D'Andrea V, Nucci F. Benign neural sheath tumours of major nerves : characteristics in 119 surgical cases. Acta Neurochir (Wien) 1997; 139: 1108–1116.