Case Report

Plexiform Schwannoma of Brachial Plexus in Axilla: A Rare Case Report

Yun Xiao1, Shu-qing Feng2, Tao Luo1, Yong-sheng Zan1, Li Li1, Li-qi Shen1

1Trauma Center, The First People’s Hospital of Yunnan Province/The Affiliated Hospital of Kunming University of Science and Technology, Kunming, Yunnan, China; 2Ultrasound Center, The First People’s Hospital of Yunnan Province/The Affiliated Hospital of Kunming University of Science and Technology, Kunming, Yunnan, China

Abstract

The most common tumours in the brachial plexus are benign schwannomas, followed by neurofibromas and malignancies, originating from the peripheral nerve sheath. The clinical manifestations of brachial plexus tumours are variable according to their location, extension, neurological elements involved and pathology. Brachial plexus tumours are rare in the upper extremity, and axillary schwannoma is uncommon. This case reports a 59-year-old woman with a tumour in her left axilla for two years, gradually enlarging with numbness in her left little finger. Microsurgical interfascicular dissection operation was performed to remove the tumour. Numbness disappeared after the procedure, and no tumour recurrence was observed during the 30-month follow-up. To the best of our knowledge, plexiform schwannoma of the brachial plexus in the axilla has not been reported so far. In this article, such a case is reported, where this tumour was diagnosed by the histopathological examination and confirmed with immunohistochemistry.

Keywords: Axillary, Brachial Plexus, Plexiform Schwannoma

Introduction

Schwannoma is a benign encapsulated nerve sheath tumour that arises from the schwannoma cells along the nerve route and can affect the third to twelfth cranial nerves, peripheral and autonomic nerves. The aetiology of schwannoma is still unknown, but it is generally accepted that there is a strong association between neurofibromatosis stype2(NF2) and plexiform schwannoma. Schwannomas rarely involve the brachial plexus and account for about 5% of all schwannoma cases. The tumour usually presents as a palpable tumour in the supraclavicular region, with or without upper extremity sensory and motor abnormalities and muscle atrophy in severe cases. Subclavian and axillary brachial plexus schwannomas are rarely reported. The tumour appeared as a nodular or ovoid large tumour; only 5% of schwannomas develop into plexiform. The diagnosis of plexiform schwannoma can be made by ultrasound, computed tomography (CT) and Magnetic resonance images (MRI). Resection is the management choice in most benign and malignant brachial plexus tumours. Generally speaking, good results are expected after surgical resection of brachial plexus schwannomas, depending on the surgeons’ skills, techniques, and experiences, especially if intraoperative neuromonitoring is used to help reduce damage to standard nerve fibres.

Case Presentation

A 59-year-old woman presented to our outpatient orthopaedic centre due to a slow-growing tumour in her left axilla on October 10, 2019. She said the tumour was noticed for the first time two years ago during a routine physical examination. After she visited a local hospital, surgeons operated on her and removed the tumour. The postoperative
pathology revealed neurofibroma; they didn’t give her any special treatment. Ten months later, she found a new tumour in her left axilla. The tumour grew slowly without pain or any other symptoms until the tumour had gradually increased in size and was associated with occasional numbness over her left little finger, so she came to our department. We were afraid of the risk that the tumour may undergo malignant degeneration and invade other normal nerves. Nothing special was reported concerning her medical record and family history.

Physical examination showed a multinodular, localized, longitudinally, soft, non-tender tumour in her left axilla. No movement of the tumour by shoulder flection and extension or rotation, motor defects, muscle atrophy, abnormal radial pulse, and capillary refill. Tinel’s sign was positive, and numbness in her left little finger. All laboratory investigations and radiographs were normal. Ultrasound demonstrated a soft tissue of 86 mm x 20 mm x 17 mm with a striated solid hypoechoic structure in the left axilla, which suggested the possibility of neurofibromatosis (Figure 1). CT revealed multiple soft tissue nodules in the left axilla, possibly of brachial origin (Figure 2). MRI showed plexiform tumours in the left axilla which were closely related to the medial tract (Figure 3).

Surgical treatment was determined under the diagnosis of schwannoma, which was exposed through a 15 cm long incision extending from the axilla to the proximal forearm under general anaesthesia through the anteromedial infraclavicular approach on October 14, 2019. After dissecting the subcutaneous tissue, the tumour was identified along the medial tract of the brachial plexus. Exploration revealed the tumour, which was a smooth solid plexiform formation with nerve fascicles stretched and displaced over the lateral of the tumour without nerve enlargement (Figure 4). After identification of the tumour, all attention was devoted to the identification, isolation, and mobilization of all adjacent plexus elements. Under the guidance of intraoperative neuromonitoring, we completely removed the
Figure 2. CT findings. A: mediastinal window showed that no significant abnormalities were observed in the mediastinum, and multiple soft tissue nodules were observed in the left axilla, with a larger transverse diameter of about 26mm (red arrow) B: lung window enhanced scan showed multiple soft tissue nodules in the left axilla. An enhanced scan showed increased signal, complete capsule and clear boundary of surrounding tissue. Multiple nodules were observed in the upper lobe and middle lobe of the right lung (red arrow).

Figure 3. MRI findings. A-B: multiple T1 long T2 signal lesions of unequal size, continuous, multinodular and round shape were observed under the left axilla, with clear boundaries and plexiform change signals. The tumour showed the same intensity as the muscle on T1-weighted images (A-red arrow) and hyperintense on T2-weighted images (B-red arrow). C-D: Three-dimensional volume enhancement scan showed uniform enhancement, with diameters between 1.6-2.1cm. Combined with brachial plexus imaging, the lesions were closely related to the medial tract, located below the axillary artery, which appears to be a conventional nerve sheath tumour (red arrow).
tumour and a small amount of brachial plexus sheath. The whole tumour was excised for biopsy, and the final histological examination showed the diagnosis of schwannoma (Figure 5). Postoperative recovery was uneventful, with healed surgical wound and no signs of infection, no neurological deficit. Numbness disappeared after the operation, and the woman returned to work on postoperative day 21. She remained asymptomatic at the 30 months follow-up appointment, with no tumour recurrence.

Discussion

Neurofibromatosis Type 2 (NF2) is a tumour suppressor gene syndrome characterized by a predisposition to schwannomas of the cranial, spinal, and peripheral nerves, meningiomas, and ependymomas. Peripheral nerve sheath tumours are common in patients with NF2 and occur in more than 40% of patients\(^6\). Peripheral sheath tumours of NF2 often present as asymptomatic, which may lead to functional
deficits of the affected nerves as the tumour grows\textsuperscript{5}. These tumours may originate from major peripheral nerves, including the brachial plexus. The most common tumours of the brachial plexus are benign schwannomas, followed by neurofibromas and malignancies\textsuperscript{6}. They are usually seen as solitary or multiple tumours; only 5\% of schwannomas develop into plexiform\textsuperscript{4}. Most tumours of the peripheral nerve sheath with the plexiform growth pattern are neurofibromas. Distinguishing plexiform schwannoma from plexiform neurofibroma is clinically relevant because plexiform neurofibroma may undergo malignant degeneration\textsuperscript{9}. At the same time, thoroughly examining any patient with a cervical or axillary tumour is crucial to avoid misinterpretation as lymphadenopathy.

The clinical manifestations of brachial plexus tumours are caused by direct nerve invasion, infiltration of surrounding tissues, or local tumour effects. Schwannomas in this area usually appear as a localized slow-growing tumour but may present as nerve compression symptoms\textsuperscript{9}. Examining the brachial plexus with ultrasound is effective because it allows many parts of the brachial plexus and surrounding soft tissues to be evaluated with high spatial resolution. Brachial plexus ultrasound is helpful for assessing sheath tumours, perineurial fibrosis, metastasis, some inflammatory neuropathy, neuralgic muscular atrophy, and post-traumatic sequelae\textsuperscript{11}. CT scans can identify vasogenic tumours by depicting the relationship between the tumour, nerves, and blood vessels and can also exclude metastatic tumours\textsuperscript{12}. MRI is reliable in differentiating between tumours and vascular anomalies in patients with neurofibromatosis. In addition, the MRI can show the multifocal nature of a plexiform. With the improvements in magnetic resonance scanners, coils, and pulse train technology, MRI became the best imaging modality, allowing us to perform routine, high-quality brachial plexus imaging. With an understanding of the anatomy of the brachial plexus and familiarity with the common pathologies that affect the area, radiologists can provide valuable imaging evaluation for patients with brachial plexus tumours\textsuperscript{13}. In our case, the MRI diagnosis of the extent of nerve involvement was accurate and compatible with surgical findings, which was very important for surgical planning. Biopsy and histopathological examination are the only definitive way to diagnose plexiform schwannoma\textsuperscript{14}.

The pathologic features of the plexiform schwannoma are similar to those of conventional schwannoma. Still, it is characterized by multinodular and plexiform growth patterns, mainly the Antoni A cell component, and the absence of the Antoni B region\textsuperscript{15}. Most brachial plexus benign tumours can be completely and safely resected through microsurgical techniques and intraoperative electrophysiology\textsuperscript{16}. Surgical removal is recommended as the preferred treatment for tumours that cause neurological deficits and discomfort, gradually increase in volume and are suspected to be malignant tumours\textsuperscript{17}. Complete removal of the tumour and preservation of the nerve is the ideal goal. Since the schwannoma is well encapsulated, it is almost always possible to enucleate and separate the tumour from the nerve sheath and microsurgical interfascicular dissection operation is a viable treatment of axillary plexiform schwannoma\textsuperscript{18-20}.

Sho Kohyama et al. reported a case of a patient with a plexiform tumour from the left C5 to C7 nerve root along the course of the brachial plexus to the left brachia. Tumour invasion was extremely extensive, and the tumour excision failed. Part of the musculocutaneous nerve was sent for biopsy, and therefore a latissimus dorsi muscle transposition was performed to restore elbow flexion. The final diagnosis of plexiform Schwannoma was histologically confirmed\textsuperscript{21}.

Brachial plexus schwannomas have a good prognosis. The eventual functional outcome after the excision of plexiform schwannoma depends on preserving unaffected fascicles through careful interfascicular dissection. In this patient, the sensory ulnar digital branch was compressed. After removing the tumour and part of the nerve sheath with the help of intraoperative neuromonitoring, numbness disappeared from her little finger, and there was no tumour recurrence at the 30 months follow-up appointment.

**Conclusion**

In conclusion, axillary plexiform schwannoma is extremely uncommon, which is a slow-growing benign tumour affecting the brachial plexus with different clinical presentations. It is essential to distinguish plexiform schwannoma from plexiform neurofibroma and lymphadenopathy. The diagnosis of plexiform schwannoma can be made with ultrasound, CT and MRI. MRI is the best imaging modality that can show a plexiform multifocal nature and be very important for surgical planning. In our case, the axillary plexiform schwannoma was symptomatic. Therefore, we performed a microsurgical interfascicular dissection operation, resulting in excellent results.

**Consent to publish**

*Written informed consent was obtained from the patient for the publication of this case report and accompanying images.*

**References**