PLEXIFORM SCHWANNOMA OF RIGHT WRIST: A RARE CASE REPORT

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Abstract

Plexiform schwannomas, which make up 3% to 19% of all schwannomas, are tumors of Schwann cells in the myelin sheath, can account for 5% of benign soft tissue neoplasms [4]. Both sexes are equally affected, and the third and sixth decades of life are the most typical times for it to occur. Diagnostic and therapeutic challenges arise because they continue to be asymptomatic; current recommendations include imaging followed by excision.

Keywords: Plexiform Schwannoma; Benign neoplasm.

Introduction

The most prevalent types of nerve tumors in the hand and upper extremities are peripheral nerve sheath tumors (PNSTs). There are two distinct types of peripheral nerve sheath tumors: benign peripheral nerve sheath tumors and malignant peripheral nerve sheath tumors [1-3]. Schwannomas are benign tumors of the peripheral nerves [13, 14]. Verocay provided the first description of them in 1908 [15]. Without regard to race or gender, schwannomas typically develop during the third and fifth decades of life [13, 16]. They typically form as solitary tumors with diameters between 1.5 and 3 cm [17–19]. 12 to 19% of schwannomas involve the upper extremities, while 13.5 to 17.5% include the lower extremity [27]. Schwannomas are primarily found on the volar surface of the upper extremities [13,14,17,18]. The tumors have a gradual, noninfiltrating growth and are completely enclosed [28,30]. Schwannomas can be difficult to investigate and treat because they might go years without showing any clinical symptoms. If the tumor starts to compress the affected nerve in the extremities, it manifests clinically as localized pain and paraesthesia. Ultrasonography (USG), MRI, and clinical characteristics can support the diagnosis. Tumor excision is the most modern surgical technique. In this case study, we discuss a 37-year-old woman who has a plexiform schwannoma on her right wrist.
A 37 years old female came to hospital with the complaints of swelling over the right wrist for past 10 years. Initially the swelling was small and gradually progressed to attained the present state. The swelling wasn’t associated with pain. She gave history of similar complaints 20 years back in the same hand for which excision biopsy was done. She was conscious oriented, her vitals were stable. On examination, there were two swellings of sizes 3x4cm and 3x3cm, one over the hypothenar aspect of right hand and another over the Volar aspect of right wrist. The swelling had well differentiated margins, skin over the swelling appears to be normal. Consistency of the swelling is firm, surface of the swelling was irregular, cross fluctuation was present, trans illumination was negative.

Ultrasound imaging done showed possibility of ganglion and MRI done revealed possibility of lymphatic or low grade venous malformation.

Fig 1. Swelling of right wrist -volar and hypothenar aspect

Fig 2. MRI image of right wrist swelling
She was then planned for surgery after obtaining anaesthesia fitness. She underwent excision biopsy under aseptic precautions under regional block and biopsy was sent for Histopathological examination. Postoperatively she was uneventful. She was on continuous compression dressing for consecutive three days.

Fig 3. Dissecting the right wrist swelling.

Fig 4. Excision biopsy of right wrist swelling

Fig 5. Histopathological examination – Antony A & Antony B bodies
Her Histopathology report showed features consistent with plexiform schwannoma. For further immunohistochemistry S-100 proteins study was done which showed strong positive. As her condition improved, she was discharged with regular outpatient follow up.

Discussion

Plexiform schwannoma was first identified by Harkin et al. in 1978, which is very uncommon and it is a benign form of Schwann cell proliferation that manifests as a multinodular growth pattern [5]. The head and neck region, upper extremities, and other body sections are where these schwannomas are most frequently found. It typically manifests as a solitary, slow-growing tumor in the dermis or subcutaneous tissue. It is rarely painful and can also grow quickly and acutely [7,8]. Before receiving treatment, symptoms may be persistent for a month to 30 years [8].

Uncertainty surrounds the causes of solitary plexiform schwannoma. However, the origin of numerous tumours is complicated and frequently linked to Gorlin-Koutlas syndrome, schwannomatosis, neurofibromatosis type II, and with a family history or traumatic past [10]. Antony A tissue, which is composed of dense and spindle-shaped Schwann cells in a palisading pattern - verocay bodies or loose hypocellular region - Antony B bodies, is prevalent in the histological appearance of plexiform schwannomas and is positive for S100 expression [12]. Additionally, MRI scans are occasionally employed to support the diagnosis. This finding points to plexiform schwannomas in a multinodular heterogeneous lesion that has tumors with low signal intensity on T1-weighted images and high intensity on T2-weighted images. Because they show the size of the lesion and how it affects nearby structures, MRI images are very useful for preoperative planning [8]. Successful treatment of schwannomas is a result of early precise diagnosis. In this case report, cytology was used to provide the initial diagnosis of plexiform schwannoma.

The suggested therapeutic option for schwannoma is surgical excision, which involves carefully removing the tumour from the nerve sheath without causing any harm to the nerve or other structures. In our case, a surgical loupe was used to precisely perform microsurgical dissection. In the end, no nerve was damaged.

Conclusions

The advent of effective microsurgical dissection techniques has made plexiform schwannomas of the extremities, which are an uncommon tumour, linked with an excellent prognosis of there is no multiple sensory or motor nerve fibres involvement. Imaging methods and a histological examination, as observed in our patient, are the foundations for the diagnosis of plexiform schwannoma. In order to improve management and outcomes, more research must be conducted.

References


