INTRODUCTION

Schwannomas are also known as neurilemomas, are common benign peripheral nerve tumours originating from schwann cells of nerve sheath. They are usually solitary, encapsulated and homogeneous masses and present with slowly growing masses, sometimes associated with pain and paresthesia. Pre-operative evaluation is based on ultrasonography and magnetic resonance imaging, but final diagnosis requires histopathology. It is important to distinguish plexiform schwannoma from plexiform neurofibroma because of the possibility of malignant transformation in plexiform neurofibroma. Plexiform schwannoma may be confused with neurofibromatosis and this can lead to overtreatment. We present a very rare case of solitary mass which had a plexiform type multicentric extension: hybrid schwannoma of the median nerve of a 20 years old girl.

CASE REPORT

A 20 years old girl presented to the outpatient clinics of the orthopaedics department with slowly growing mass on her right hand and forearm which had been there for 6 years. She was complaining of mild pain and paresthesia for the last one year. She remembered no history of trauma or infection. There was no family history of neurofibromatosis and no associated clinical properties like café-au-lait spots. On physical examination, painless, mobile, solid mass was palpable at volar forearm and radial border of the right hand.

There was no erythema or warmth and no objective motor and sensory deficit. Tinel's sign was positive when tapping over the carpal tunnel.

Laboratory tests (complete blood count, blood biochemistry, erythrocyte sedimentation rate) were in normal ranges. Ultrasound imaging showed that the mass was multiple, solid and separate from the adjacent flexor muscles and tendons. Magnetic resonance imaging (MRI) showed multiple smooth, well-defined, fusiform masses that exited the nerve within the right hand and forearm on T1-weighted and STIR images (Figure 1a and 1b). Intermediate to low signal on T1-weighted and diffuse increase in signal on T2-weighted images; the tumour was considered to be schwannoma.

Surgical exploration showed that the entire median nerve was grossly enlarged from the proximal to the distal segments. There was a continuous multinodular mass involving the median nerve. Also there was two satellite masses at the same median nerve trunk. One of them was adjacent to the main mass, other was located on the digital nerve of the index finger (Figures 1c - 1f). The sheath of the nerve was incised longitudinally to minimize damage to the nerve fascicles and the masses were resected en-bloc by sharp dissection using microscope with no complication (Figures 1c - 1i). At the proximal part, tumoural invasion into the fascicules of median nerve made the dissection and the resection impossible even under the microscopy so that the proximal part of the tumour was not excised to avoid nerve damage. Pathological examination was carried out on a frozen tumour section and schwannoma was corroborated.

At the final follow-up (12 months after the operation), there was no sensory and motor deficit and clinically any recurrence or muscle atrophy were not observed.

ABSTRACT

Schwannomas are the most common benign neoplasms of the peripheral nerves in the upper extremity originating from the schwann cells of nerve sheath. They account for 5% of all tumours in upper extremity. Schwannomas are usually solitary, encapsulated and homogeneous masses and present with slowly growing masses.1 Plexiform type peripheral nerve schwannomas can also be seen which are uncommon benign masses.2 Most common findings of schwannomas are painless swelling or sporadic pain and sometimes paraesthesiae or neurological deficiency when it grows into the nerve or grows within an enclosed space such as the carpal tunnel.3 We report an unusual case of a solitary mass which had a plexiform type multicentric extension and was diagnosed to be a hybrid schwannoma of the median nerve of a 20 years old girl.

Schwannoma of median nerve

DISCUSSION

Schwannoma is the most common benign neoplasm of the peripheral nerves in the upper extremity, however, both schwannomatosis and plexiform schwannoma is an extremely rare benign neurogenic mass. The mass appears most often on trunk, head and neck, or upper extremity. They may be found anywhere from the brachial plexus to end of the digital nerve. Just 5% of them develop into the plexiform type and these are reported schwannomas larger than 5 cm. Schwannomas commonly develop as slowly-growing, isolated masses less than 3 cm diameter. A few studies reported schwannomas larger than 5 cm. The mass slowly pushes the nerve fascicles aside, so the symptomatology is very often minimal and surgical enucleation can be performed with no loss of nerve function. In this case, the tumour had multicentric solitary masses and plexiform area, after resection of some masses and intact digital nerve, after removed mass. The histopathological appearance (HEx50): Antoni A (β) area, highly cellular, tightly compact spindle-shaped cells and Antoni B (α) area, loose myxoid, loosely arranged cells (g). Immunohistochemical study (S100x100); using anti-S-100 protein staining, stained the nuclei and cytoplasm of nearly all of the tumour cells. Gross appearance of the schwannoma following removal from the median nerve.

REFERENCES