Rare disease

Benign palmar schwannoma – a rare case in a handball player

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Summary

Peripheral nerve sheath tumours (PNSTs) of the hand are rare. Schwannomas, also known as neurilemmomas, are the commonest benign PNSTs. They arise from a proliferation of Schwann cells. Aetiology is unclear. In this report, the authors present the case of a lifelong Rugby Fives (a handball variant) player who developed a symptomatic benign schwannoma at the impact point on his palm. To our knowledge, there are no documented cases of upper limb schwannomas which may be related to repetitive trauma from sport.

BACKGROUND

Schwannomas, or neurilemmomas, are the commonest benign peripheral nerve sheath tumours (PNSTs), accounting for 5% of all tumours in the upper extremities¹ but remain a rare differential for hand lumps. They are generally solitary, slow-growing lesions, although rarer, multiple lesions can occur in individuals especially in association with either neurofibromatosis or schwannomatosis.^{2–3} Microscopically, they are homogenous, well-differentiated and encapsulated tumours deriving from schwann cells (glial cells providing neuronal support in the peripheral nervous system, primarily for myelination).⁴ While malignant transformation is exceedingly rare and tumours remain slow-growing, malignant PNSTs have a very poor prognosis.⁵ Although benign, schwannomas can become symptomatic due to mechanical compression and present with pain, swelling or a lump or sensory change. Surgical management is enucleation/excision without disturbing the surrounding nerve fascicles and recurrence is rare.^{6,7}

Fives is a handball game which resembles squash or racquetball; however hands are used in place of racquets. There are several versions of Fives, the most well known of which are Eton Fives and Rugby Fives – the names refer to the independent schools in which the game flourished in the late 19th and early 20th century.^{8,9} Though cork based and covered with leather, the balls are small and hard and can be propelled at significant speeds.

Due to the rarity of these benign schwannomas, description in the literature is restricted mainly to case reports or case series. In a literature search for schwannomas in the hand and associated MeSH terms, the last case report described a child with an intraosseous schwannoma.¹⁰ There are no articles relating schwannomas to sport or repetitive trauma.

CASE PRESENTATION

A 70-year-old left hand dominant gentleman presented to his general practitioner with a 3 year history of pain in his right palm when playing Fives. There was no palpable lump or obvious swelling and no pain on palpation. He regularly played Rugby Fives, a variant of handball/squash whereby players hit a small hard ball with their hands,

protected only by thin leather gloves. He had started playing as a young boy at school and had played 1½ to 2 hours a week every week for a sum total of 33 years. He suffered from significant impact pain with radiation in the ulnar nerve distribution and ultimately this pain was preventing him from playing. His symptoms were reproducible on clapping his hands together.

He was tender to palpation in the palmar aspect of his right hand over the base of the third and fourth metacarpal bases, corresponding to the repetitive point of impact of the Fives ball.

INVESTIGATIONS

A magnetic resonance scan (MRI) showed a defined lesion deep to the flexor tendons which, radiologically, suggested a ganglion at the level of the metacarpals (figure 1).

DIFFERENTIAL DIAGNOSIS

Tumours of the hand are common but (excluding cutaneous malignancies) are benign in the significant majority.¹¹ Early accurate diagnosis bearing in mind an index of suspicion of malignancy can avoid mismanagement and loss of function. The most common tumour from soft tissue – including neural tissue – is a ganglion cyst (aetiology unknown; a theory related to degeneration of mucoid connective tissue had been proposed) and the most common bone tumour is the enchondroma (benign cartilaginous neoplasms in intramedullary bone).¹² After that, in order of decreasing frequency come inclusion cysts, giant cell tumours, granulomas, haemangiomas and lipomas. In terms of malignancies, cutaneous tumours (squamous cell carcinoma is most common primary in the hand) are much more common than bone or soft tissue tumours.

TREATMENT

The patient underwent an excision procedure of the lesion via a zig-zag approach over the area of pain. The common digital nerves (CDN) and arteries were identified, and a soft ovoid lump was identified arising from the CDN to the fourth web space. This was enucleated from the nerve (figure 2). The tissue was sent for histology which demonstrated a benign schwannoma.

Figure 1 MRI scan of hand.

OUTCOME AND FOLLOW-UP

Postoperatively, he underwent hand therapy and at 4 week follow-up he was pain free. At 10 weeks postprocedure he was fully rehabilitated and had resumed his previous level of sporting activity.

DISCUSSION

Schwannomas are rare and so are often misdiagnosed or are diagnosed only on histology postoperatively,¹³ as in this case. As such, information on the aetiology of schwannomas remains scarce and at present most of the evidence in the current literature is hypothetical or anecdotal. Many schwannomas occur as solitary lesions and are often found incidentally;¹⁴ however, multiple schwannomas can occur mostly related to conditions comprising the neurofibromatosis (neurofibromatosis type II – NF2 and schwannomatosis).¹⁵ The increased incidence of these conditions is reflected in the evidence base – a genetic basis has been identified in NF2. The NF2 gene on chromosome 22 controls the production of merlin, a tumour suppressor.^{16 17} It is not unreasonable to consider there may be a de novo genetic element in solitary schwannoma aetiology.

While the null hypothesis in this case would dictate that a link between playing Fives (or any sporting activity involving repetitive blunt trauma) and the development of a schwannoma was coincidental, clearly the inability to prove causality does not exclude the possibility of contribution to aetiology. Given that the patient's symptoms were only evident when he was playing sport and he had previously been asymptomatic despite the duration of his Fives involvement, this might suggest that it had developed as a consequence.

There have been isolated cases reporting the association of schwannomas with foreign bodies^{18 19} and there

Figure 2 Intraoperative photo.

has been suggestion that microtrauma to the skin may stimulate the growth of schwann cells.²⁰ We speculate that playing Fives regularly may have facilitated dermal implantation of foreign material (leather fibres from the protective gloves) by microabrasion/trauma thereby stimulating schwannoma growth.

Learning points

- ▶ Schwannomas should be included in the differential diagnoses for soft tissue lumps of the upper limb.
- ▶ Schwannomas are rare.
- ▶ There may be a link between dermal abrasion/microcellular trauma and schwannoma aetiology.

Competing interests None.

Patient consent Obtained.

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