Intraneural mucoid pseudocysts
A REPORT OF TEN CASES
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A mucoid pseudocyst of a peripheral nerve is a rare and benign tumour of controversial origin. We have reviewed ten patients with a mean follow-up of 3.2 years. The tumour affected the common peroneal nerve in eight and the ulnar nerve in two. The mean time between the onset of symptoms and diagnosis was 7.4 months (1.2 months to 2 years). On examination, there was pain in eight patients and swelling in seven. Motor deficit in the corresponding nerve territory was found in all. The diagnosis was usually confirmed by MRI. Treatment was always surgical.

All the patients recovered, with a mean time to neurological recovery of 10.75 months. Recurrence was seen in only one patient and was treated successfully by further surgery. Our results are similar to those reported by other authors. A successful surgical outcome depends on early diagnosis before neurological damage has occurred.

Neural mucoid pseudocysts are rare, benign, peripheral nerve tumours which were first described by Zum-Bush in 1895. Their origin is characterised by the infiltration of the neural sheath by a mucoid substance which forms an intraneural tumour, compressing the nerve fascicles towards the periphery. There is no epithelial lining and the pathogenesis is controversial. We report the treatment and outcome of these intraneural cysts.

Patients and Methods
Between 1981 and 1999 we treated ten patients, eight men and two women with a mean age of 48 years (27 to 78).

The mean follow-up was for 3.2 years (1 to 7.4). Eight patients presented with lesions of the common peroneal nerve at the level of the neck of the fibula and two with lesions of the ulnar nerve (elbow and wrist). Seven complained of local pain and one of radiating pain. Two had isolated motor paralysis of the common peroneal nerve. Swelling was never a presenting complaint. The clinical signs appeared after exertion in four patients. The mean time between the onset of symptoms and diagnosis was 7.4 months (1.2 months to 2 years). Table I gives the details of the clinical findings.

Plain radiographs showed 10° of genu varum in one patient, and 25° of cubitus valgus in another, the latter as a sequel to a displaced fracture of the elbow in childhood. We performed ultrasound studies on three patients. Two studies showed a hypoechoic zone (liquid tumour) within the common peroneal nerve. CT was carried out on two patients and showed a cystic, hypodense, homogeneous swelling. MRI was undertaken on five patients and confirmed in each the liquid nature of the tumour. The swelling was intermediate on the T1- and hyperintense on the T2-weighted scan. This was unchanged after administration of intravenous gadoliumum. The nerve appeared to be thickened and laminated. In two patients, examination showed no connection with the adjacent joint. The EMG identified a neurological deficit in eight and a conduction block in three.

All patients underwent surgery, in nine at initial presentation and in the tenth after a recurrence. In each case the surgical management was similar. The operation was performed under pneumatic tourniquet using an approach which allowed dissection of the nerve trunk, divisions and branches. The fibrosascular arch was divided (e.g. peroneal canal or the arch of flexor carpi ulnaris). After epineurotomy (Fig. 2) using an operating microscope,
the contents of the pseudocyst were evacuated, preserving neural integrity. The resulting cavity was not closed. An exoneurolysis, with nerve stimulation, was also undertaken. Any communication between the pseudocyst and the joint was carefully sought. The wounds were closed after haemostasis and the joint was splinted for two weeks.

Results

Macroscopically, the tumour presented either as a single mass or as several translucent areas embedded in the nerve trunk. The contents resembled yellow jelly. Neither the cysts nor the nerve had connections with the adjacent joints. After evacuation of the pseudocyst, exoneurolysis confirmed that the nerve fibres were in continuity and neither strangled nor compressed. Histological examination showed no evidence of an epithelial lining.

The mean time of postoperative recovery of any motor or sensory deficit was 10.75 months (1.6 to 17.7). The age of the lesion determined the speed of the recovery. One patient had three recurrences after complete neurological recovery. These were always heralded by the reappearance of a nodule on the nerve and followed after a few months by a neurological deficit. Evacuation of the contents of the pseudocyst led to a permanent cure.

Discussion

Neural mucoid pseudocysts are rare and benign nerve tumours. There are few reports of series; usually only isolated cases have been described. There has been a preponderance of cases reported in the legs, in most affecting the common peroneal nerve at the neck of the fibula. Other sites are near a joint or a narrow compression tunnel such as the arch of flexor carpi ulnaris at the elbow. Lesions may occur in motor or sensory nerves but mostly in mixed nerves. They are uncommon in the peripheral nerves of the upper limb. Although they usually affect the ulnar nerve at the elbow, cysts have also been reported at the following sites: 1) the posterior interosseous nerve at the level of brachioradialis; 2) the median nerve at the level of pronator teres and in the carpal tunnel; 3) the ulnar nerve in Guyon’s canal and at the level of the deep palmar aponeurosis; and 4) the digital nerves and their dorsal branch. One case has been described of cysts in different nerves in the same thumb.

Neural mucoid pseudocysts often affect middle-aged men and usually present with pain or the symptoms of nerve compression. The appearance of clinical signs after exertion is characteristic. A history of acute minor trauma is often noted. The pain may be due to intracystic bleeding. Soon afterwards a neurological deficit in the corresponding nerve territory appears and the pain settles briefly. The time between the onset of symptoms and diagnosis is shorter than for other nerve tumours and varies between 3 and 7.4 months. This is perhaps because of rapid growth with associated nerve deficits, which are alarming for the patient, and the superficial nature of the tumours.

Pain is usually intermittent and a positive Tinel’s sign is uncommon. A swelling or nodule on the course of the nerve may be found. A motor deficit was present in all our patients and sensory changes in approximately 50%. Plain radiographs are usually normal. Although ultrasound can identify the location and nature of the cyst, MRI is more helpful. It clearly demonstrates the cystic nature, anatomical location, and extent of the lesions. It can also assess the state of the nerve. An intermediate T1-weighted image with a hyperintense bright signal on the T2-weighted image remains unchanged after an intravenous injection of gadolinium. The contents are heterogeneous. MRI allows differentiation between an adjacent articular synovial cyst and a cystic schwannoma.

EMG and nerve-conduction studies should be performed to document any neurological deficit and to help to localise lesions. A conduction block suggests a favourable prognosis.

Intraneural mucoid cysts can be confused with other nerve tumours, nerve-entrapment syndrome or an adjacent, compressive, articular synovial cyst. With the latter, the articulation is often abnormal. The cyst can develop far
from the damaged joint and histological examination will show an epithelial lining. At the neck of the fibula an intraneural mucoid pseudocyst can be confused with a nerve-entrapment syndrome, effort palsy of the common peroneal nerve or sciatica. At the elbow, it can be confused with an extrinsic nerve compression as may be seen in post-traumatic fibrosis, osteochondromata, muscle anomalies and rheumatoid cysts.

Treatment is always surgical. Nerve resection and grafting must not be performed even if the lesions appear to be extensive. Intrafascicular dissection is contraindicated because of the risk of fascicular damage. It is essential to maintain nerve continuity, first by incision and drainage of the contents of the cyst after epineurotomy, then by division of the neighbouring fibromuscular arch. An exoneurolysis is also performed. Complete resection of the pseudocyst is dangerous if not impossible. There is no plane of dissection between the tumour and the adjacent fascicles. Although a communication between the pseudocyst and the joint is rare it must nevertheless be sought. The contents are similar to those of synovial cysts. The intracystic liquid is an acellular mucopolysaccharide which differentiates it from other liquid neural tumours of peripheral nerves such as a cystic schwannoma. The cystic wall has a fibrolamellar pattern and contains some inflammatory cells but no epithelial lining. The laminated nerve fibres on the external surface of the pseudocyst are often flattened and form part of the wall. They are elongated and, in the presence of severe compression, can be destroyed.

Long-standing tumours, the mean time to neurological recovery, which occurs in most cases, is ten months. Pain disappears rapidly after decompression and recovery in our patients was complete within a few months. This is in contrast to other published series. Recurrences are rare, but may begin with the appearance of a new nodule. We observed one patient who experienced three recurrences.

A successful outcome after surgical treatment depends on the early diagnosis, before neurological damage has occurred. Patients with pseudocysts on proximal nerve trunks achieve better results than those with distal lesions. This may be because of the less regenerative ability of more distal nerves. Long-term follow-up should include clinical examination and MRI if necessary.

References