

Original Article

Pigmented villonodular synovitis of the elbow with radial, median and ulnar nerve compression

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Abstract: Pigmented villonodular synovitis (PVNS) is a rare, idiopathic proliferative disorder of the synovium. While, PVNS of elbow is extremely rare. We report an 82-year-old female patient with 20-year-history of gradually increased PVNS in her left elbow. The multiple masses were located in anterior, medial and lateral of elbow. Her radial, median and ulnar nerves were compressed by the tumor. We resected tumor of extra-articular part piecemeally and released the compression of nerves. After the surgery, the patient gained a functional recovery. Two years after surgery she had a tumor recurrence, but without any symptoms of nerve compression syndromes. We discussed its clinical diagnosis, radiological features, MRI findings, pathophysiology, and treatment.

Keywords: Pigmented villonodular synovitis, nerve compression

Introduction

PVNS is classified as a benign tumor, it causes locally progressive synovial proliferative lesion in the joint, bursae, and tendon sheath. It usually affects hip or knee and can also occur in the shoulder, ankle, elbow, hand and foot. PVNS can occur in all age groups. The aetiology of PVNS includes a locally aggressive neoplasm or a reactive synovitis. PVNS of elbow is extremely rare. We report a unique case of extra-articular PVNS of elbow. The mass was compressing radial, median and ulnar nerves simultaneously. No case of those nerves that are compressed by PVNS of elbow has been described in the literature before.

Case presentation

An 82-year-old female patient presented with a 20-year history of gradually increased masses in her left elbow. She felt initially mild pain and swelling of her joint, and reported no history of trauma. Her symptoms were well controlled with non-steroidal anti-inflammatory drugs. But during the previous 6 months, she felt progressive weakening the function of finger extension and numbness of forearm and hand. The mul-

iple masses were located in anterior, medial and lateral of elbow (**Figure 1**). They were soft and their margins were well-defined. Her elbow range of motion was 0° to 130° of flexion, 85° pronation, and 70° supination. Neurologic examination showed impaired function of sensory and motor deficit. Electromyography (EMG) showed that the conduction velocity of radial, median and ulnar nerves were decreased, and the action potential amplitude was decreased as well. Deep branch of radial nerve injury was slight damaged. Laboratory studies and tumor biological markers were normal. Radiographs showed lateral epicondyle of humerus bone defected with soft tissue swelling. Magnetic resonance imaging (MRI) (**Figure 2**) showed: In T1-weighted images. The lesion showed mildly low signal intensity to muscle; in T2-weighted images. it showed hyper- or isointense to muscle signal intensity; the masses were enhanced heterogeneously by the administration of gadolinium-diethylene triamine pentaacetic acid (Gd-DTPA) contrast agent, demonstrated small lytic lesions and bone defect involving the distal humerus, radial head and olecranon. Biopsy diagnosed PVNS before the operation. Considered the patient's age, the main purpose of the operation is based on releasing the com-



Figure 1. Pre-operation photograph, showing multiple large-sized masses in the elbow.

pression of nerves. The medial approach was designed to release ulnar nerve in the left elbow under general anesthesia and through an anterior longitudinal lateral approach radial and median nerves were released (**Figure 3**). We excised the masses in the extra-articular of elbow as much as possible and performed synovectomy. These masses extended from the outer layer of the capsule. The tumor presented the large size and partial necrosis of the tumor occurred (**Figure 4**). Frozen section and Histologic findings revealed PVNS and it had central zone of degeneration and necrosis. Micrograph showed numerous Mononuclear phagocytes, foamy macrophages, inflammatory cells and polynucleotic giant cells. Hemosiderin accumulation and inflammatory reaction were also observed (hematoxylin-eosin stain, original magnification $\times 400$) (**Figure 5**).

Three days after the surgery, the patient was allowed to perform rehabilitation exercise. The patient had taken Methylcobalamin Tablets for half a year (0.5 mg every 8 h; Eisai Co, Ltd Japan). One month after the surgery, her elbow range of motion recovered to the preoperative level of function. Three months after the surgery, the patient could extend her fingers and the sense of numbness decreased. Electromyography showed the recovery of normal nerve conduction velocity. Two years after the surgery, MRI presented the tumor of the antero-medial joint capsule had grown (**Figure 6**). We considered that she had a partial tumor recurrence. She had no symptom of nerve compression syndromes, we chose to continue observing and following up with the patient.

Discussion

PVNS, rare and benign, is a locally invasive disease of the synovium [1]. It affects both genders equally, and has an overall incidence of 1.8 per million [2]. Adults between thirty and forty present commonly with symptoms of pain and recurrent effusions of the joint [3]. It is clear that the knee, followed by the ankle, hip, shoulder and wrist, is the most commonly affected joint [4]. However, PVNS of the elbow is rare. It includes two different forms: localized (LPVNS) and diffuse (DPVNS) [5]. LPVNS occurs in small joints, whereas DPVNS typically affects the entire synovial lining of large joints. The aetiology of PVNS is not clear, although it has been considered to be either a locally aggressive neoplasm, a reactive synovitis or repetitive atraumatic haemarthroses distinct from haemophilia [6-8]. The use of MRI [9] can show combination of synovial proliferation and soft tissue masses, together with deposits of hemosiderin and bone erosion, is highly diagnostic for PVNS, can also adequately displays the location and extent of the lesion and its internal structure. The differential diagnosis of PVNS includes hemophilia, synovial hemangioma, synovial osteochondromatosis, and Synovial sarcoma [10]. Although recognizing the benign nature of the PVNS, its recurrence is common. The recurrence rate after surgical resection is reportedly 8 to 56% [11].

PVNS requires a synovectomy that is as extensive as possible to reduce the recurrence. Due to the complexity of the joint structure, complete resection of the synovial tissue in these joints is often difficult. Recognizing the patient's advanced age and the function of elbow hadn't been limited seriously before the operation, we didn't choose arthroscopic synovectomy and total elbow arthroplasty. The main purpose of the operation is bases on releasing nerves and excising the tumor as much as possible. Due to the large-sized mass and the need of releasing nerves, we chose combined approach. And fortunately the patient's scar formation didn't result in the limitation of elbow joint motion. Meanwhile, radiotherapy is an effective adjuvant therapy for the prevention of recurrence and complaints [12]. Only four Neural involvement of PVNS cases have been reported in the literature [13-16].

PVNS of the Elbow with three nerve compression

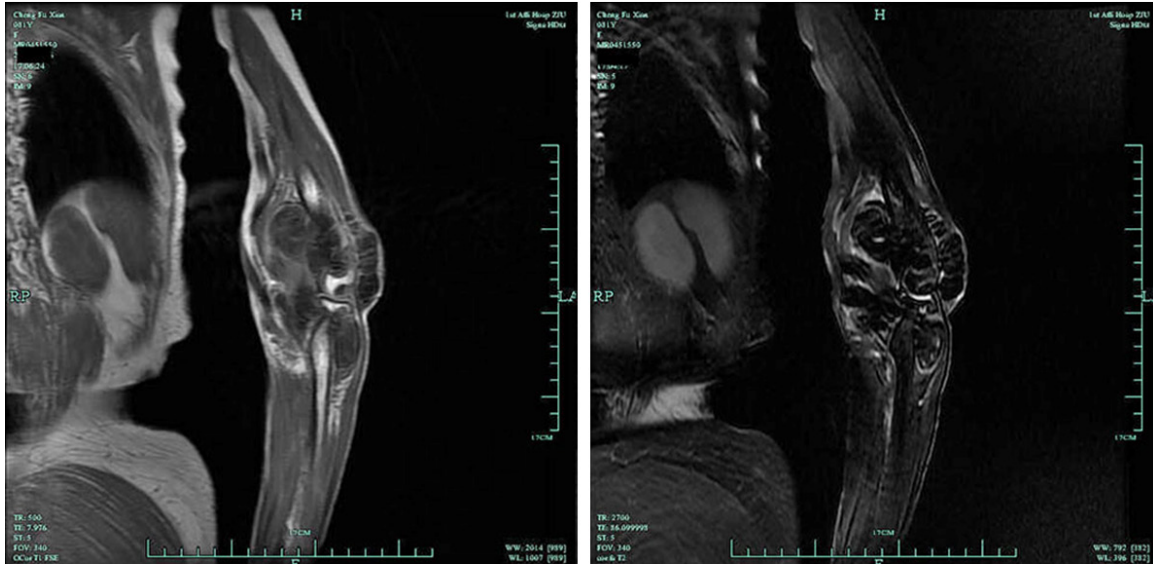


Figure 2. MRI of pre-operation showing T1-weighted images. Multiple masses located in internal and external joint showed mildly low signal intensity to muscle; T2-weighted images. they showed hyper- or isointense to muscle signal intensity.



Figure 3. The photograph during the operation, showing the released ulnar nerve, median nerve and radial nerve which were entrapped by the masses. The exfoliation was located in the capsule of the tumor tissue of the nerve surface. The partial nerve trunks were compressed into flat and thin.

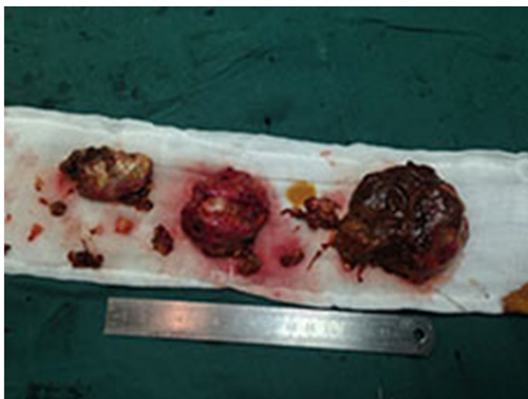


Figure 4. the resected tumor tissues, showing the masses in the extra-articular of elbow were excised basically and partial necrosis of the tumor occurred.

After releasing the compression of nerve entrapment, the patient can achieve rapidly and completely recovery. However, without finding the compression of nerve entrapment, delayed diagnosis and treatment may cause the bad outcomes.

As PVNS occurring in the elbow can compress nerves, early diagnosis and treatment are essential to

maintain joint function and avoid neuropathy. It is also important to perform the individualized treatment plan for the special cases.

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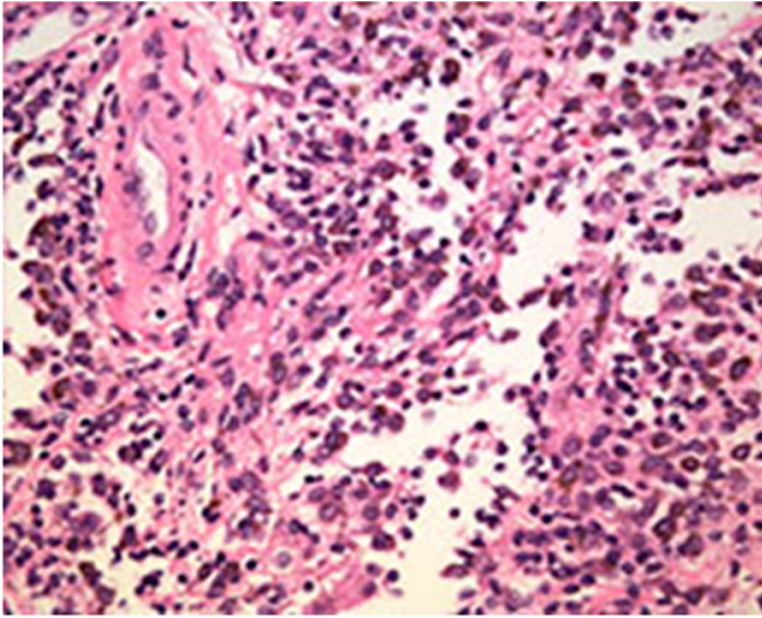


Figure 5. Pathological photograph, showing numerous Mononuclear phagocytes, foamy macrophages, multinucleated giant cell inflammatory cells and polynucleotic giant cells, hemosiderin accumulation and inflammatory reaction were also observed (hematoxylin-eosin stain, original magnification _400).



Figure 6. Two years after the surgery, MRI showed that the tumor had a partial tumor recurrence.

Written informed consent was obtained from the patient for publication of this article and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

Disclosure of conflict of interest

None.

Authors' contribution

Hui lu drafted the manuscript. Qiang chen and hui shen participated in the design of the study and performed the statistical analysis. Hui lu conceived of the study, and participated in its design and coordination and helped to draft the manuscript. All authors read and approved the final manuscript.

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