CASE REPORT

INTRANEURAL PERINEURIOMA OF THE BRACHIAL PLEXUS

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ABSTRACT

Intraneural perineurioma (IP) is a rare and slowly progressing disease which is characterized by spindled perineurial cells, arranged in pseudo-onion bulb whorls around one or more centrally situated Schwann cells. Diagnosis of the disease is based mainly on histologic findings and immunohistochemical profile. Although cytogenetic and molecular genetic studies are of limited value, they may play an important role in the differential diagnosis of the IP, with many other overlapping soft tissue tumors.

Keywords: Intraneural perineuroma, Brachial plexus, Tumour

BRAKİYAL PLEKSUSUN İNTRANÖRAL PERİNÖROMASI

ÖZET

İntranöral perinöroma (İP) iğsi perinöral hücrelerin ortalarında schwan hücreleri bulunarak veya bulunmayarak oluşturdukları soğan benzeri histopatolojik yapı ile karakterize nadir görülen ve yavaş ilerleyen bir hastalıktır. Tanısı büyük oranda histolojik bulgulara ve immunhistokimyasal profile dayanır. İP diğer yumuşak doku tümörlerinden ayrılmasında sitogenetik ve moleküler genetik çalışmalar önemli rol oynar.

Anahtar Kelimeler: İntranöral perinöroma, Brakiyal pleksus, Tümör

INTRODUCTION

Intraneural perineuroma (IP) is a benign and slowly progressing rare clinical entity which generally tends to affect the peripheral nerve sheaths of the thick nerves of the upper extremities. It rarely involves the cranial nerves[1-5]. The illness is associated with a slow, painless and progressive loss of motor function and occasional sensory loss in the affected nevre[1,5]. The process occurs in a wide age range, from 2 to 64 years, and there is no sex dominance[5]. The segment of involved nerve reported in the literature ranges from less than a centimeter to 18 cm in size[5].

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CASE REPORT

A 76-year-old man presented with a 2-year history of a slowly enlarging painless mass on the left side of his neck and progressive loss of strength in his left upper extremity. He had no other significant local or systemic symptoms and signs of infection. Neurologic examination revealed a total motor loss in the muscles of the left upper extremity which were innervated via the nerves of the left brachial plexus. A fine needle aspiration biopsy was performed 2 years previously elsewhere with a diagnosis of adipose tissue. His medical history was significant for Multiple Sclerosis, diagnosed 25 years previously.

Physical examination revealed a 4x3 cm round, fixed and rubbery mass with palpation over the left supraclavicular area. There were no signs of infection on the overlying skin and no other lesions on the face, trunk or extremities. A magnetic resonance imaging (MRI) of the neck demonstrated a mass 3cm in diameter, probably originating from a peripheral nerve at the left supraclavicular region. Thickening of the peripheral nerves in the adjacent of the mass was also reported.

Intraoperatively, a rubbery, brown 5x4 cm mass adherent to and surrounding the left brachial plexus was excised together with the involved nerves. Pathologic evaluation of the involved fusiform nerves revealed pseudo-onion bulbs composed of concentric layers of proliferated perineurial cells, ensheething a central axon and Schwann cell, in cross section. A hematoxylin and eosin (H&E)-stained specimen section showed numerous pseudo-onion bulbs with concentric intraneural lamellar proliferation of the perineural cells (Figure 1). Immunostain for epithelial membrane antigen (EMA) demonstrated a membranous pattern of reactivity in the perineurial cells of the pseudo-onion bulbs (Figure 2). Immunostaining for S-100 protein revealed the positively-stained Schwann cell sheaths at the centers of the pseudo-onion bulbs (Figure 3). Neuronfilament protein (NFP) positive axons were seen to lie at the centers of the pseudo-onion bulbs (Figure 4).

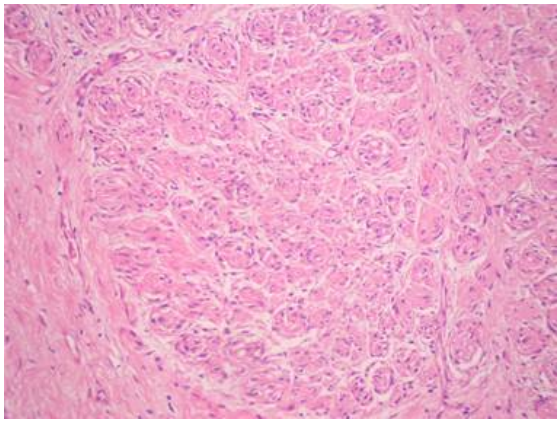

Figure 1: Hematoxylin and eosin (H&E) stained specimen section showing numerous pseudo-onion bulbs with concentric intraneural lamellar proliferation of perineural cells (original magnification x 400).

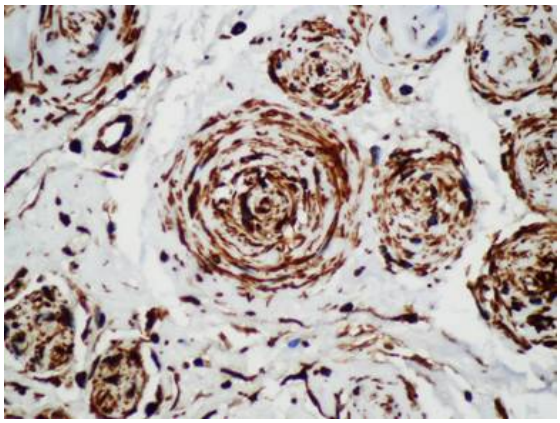

Figure 2: Immunostaining for epithelial membrane antigen (EMA) demonstrated a membranous pattern of reactivity in perineurial cells of the pseudo-onion bulbs (original magnification x 400).

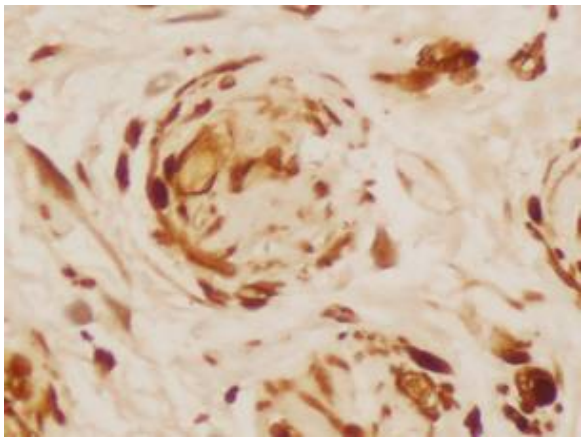

Figure 3: Immunostaining for S-100 protein revealed the positively stained Schwann cell sheaths at the centers of the pseudo-onion bulbs (original magnification x 400).

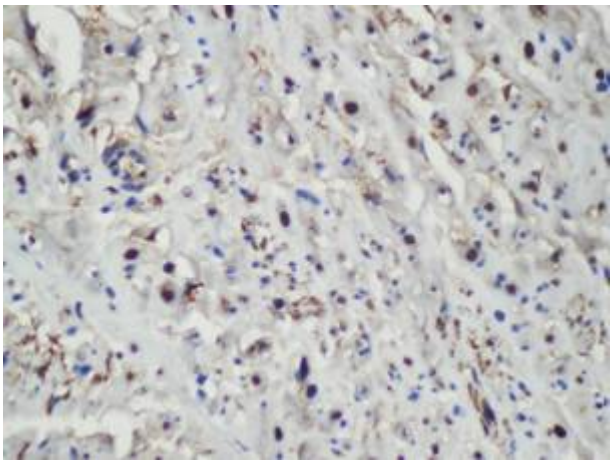

Figure 4: Neuronfilament protein (NFP) positive axons were seen to lie at the centers of the pseudo-onion bulbs (original magnification x 400).

DISCUSSION

The disease was described by Lazarus and Trombetta in 1978[2]. The term ‘’perineurioma’’ covers two distinct but similiar entities, one an extraneural soft tissue tumor, the other a permeative intraneural benign tumor[1,3]. Extraneural perineuriomas are benign tumors which occur primarily in the subcutaneous tissues of the trunk and limbs as a painless, solitary nodule or mass[1,4]. Occasionally, they can occur in the dermis and deep soft tissues, rarely, they can be found in the head and neck area, retroperitoneum, brain, kidney and intestines[1,4]. The term intraneural perineurioma was suggested by Emory et al[3] in order to unite the clinical entities previously known as localized hypertrophic neuropathy, hypertrophic mononeuropathy, localized hypertrophic neurofibrosis, intraneural neurofibroma, and hypertrophic interstitial neuritis.

IP is an uncommon benign tumor characterized with the neoplastic proliferation of the perineurial cells. The axons and Schwann cells of the nerves are protected with layers of epineurium, perineurium and endoneurium. The perineurium, which is composed of perineurial cells, is located between the epineurium and endoneurium. It was a subject of debate whether the proliferation of the perineurial cells of IP is a neoplastic or reactive process due to repeated trauma and resultant loss of integrity of the normal perineurial barrier[3]. Evidence for the theory of trauma and nerve entrapment is not convincing. On the other hand, cytogenetic studies have revealed that the IP show chromosome 22 abnormalities; monosomy or deletion of the 22q11~q13.1 bands[3]. Although, previously, some authors had advocated this process as reactive, lately, IP has been accepted as a benign neoplasm in the literature[1,4].

Diagnosis of the disease is based on mainly histologic findings and the immunohistochemical profile[1,4,5]. Although a number of stains can be used for diagnosis in the histologic examination and immunohistochemistry; hematoxylin and eosin (H&E), epithelial membrane antigen (EMA), S-100 protein, neuronfilament protein (NFP) are the most valuable[1,4]. The affected nerves characteristically reveal the concentric intraneural lamellar proliferation (pseudo-onion bulb) of the perineural cells resulting in the sausage-shaped swelling of a segment of the nerve. Immunohistochemistry shows EMA-positive (Figure 2) perineurial cells surrounding Schwann cells and a central core of axons, positive for S-100 (Figure 3) and NFP (Figure 4) respectively.

Differential diagnosis of IP includes localized hypertrophic neuropathy, hereditary hypertrophic neuropathies (Charcot-Marie-Tooth and Dejerine-Sottas), neurilemmoma, neurofibroma, and neuromas[1,3,4,5]. Although pseudo-onion bulb whorls of IP are composed of EMA-positive and S-100 negative perineurial cells, the pathologies mentioned above are characterized by true onion bulb whorls of uniform S-100 positive and EMA-negative Schwann cells with a centrally located myelinated axon. Extraneural perineurioma (a benign tumor unassociated with nerves) and IP are related only in their cytogenesis and they can be easily differentiated with their presentation site and clinical appearance[3].

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