Clinical examination of brachial and pelvic plexus tumors

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✓A thorough history and physical examination are fundamental to the assessment of patients with brachial and pelvic plexus tumors. Typical of most peripheral nerve tumors, the presenting symptoms and signs are few, and if present, can be subtle. Presenting complaints may include a palpable mass lesion, either symptomatic or asymptomatic; sensory alterations; pain; motor deficits; visceral symptoms; or autonomic dysfunction. Motor deficits are usually a late feature in the pathogenesis of this lesion, and a progressive course of pain and significant sensory and motor deficits suggests a malignant pathological process. A detailed family history may reveal familial syndromes and neurocutaneous disorders that predispose the patient to neoplasia, such as neurofibromatosis. The physical examination should be conducted in a systematic fashion, looking for any cutaneous features and motor and sensory deficits. The mass should also be examined for form, consistency, and mobility. An irregular, firm, and immobile mass suggests a malignant lesion. Complete and accurate clinical information must be gathered to pinpoint the anatomical localization of the lesion and formulate a differential diagnosis.

KEY WORDS • brachial plexus • lumbosacral plexus • neurofibromatosis • peripheral nerve tumor

Symptoms and Characteristics of Brachial and Pelvic Plexus Tumors

Sensory Alterations

Sensory changes or loss is commonly an early complaint, and often it is the only complaint. Despite what may sometimes be a vague description offered by the patient, an attempt at defining the precise distribution and nature of the deficit should be made. Sensory alterations can be manifested in a number of ways: complete loss of sensation (anesthesia); decreased sensation (hypesthesia); or increased sensation (hyperesthesia). Patients may also complain of tingling, electric shock–like sensations, or the sensation of pins and needles (paraesthesia) or a disagreeable burning pain (dysesthesia). In addition, light touch or other non-painful stimuli may be perceived as painful (allodynia). The sensory changes may be very subtle initially and described by the patient as only a vague sensory alteration. In advanced cases, the functional consequences of sensory loss may manifest with a history of accidental burns, ulcer formation, inadvertent injury to digits, and dropping of objects.

Mass Lesion Characteristics

Many peripheral nerve tumors are initially found by the patient as a painless lump that is otherwise asymptomatic. Unlike nerve tumors in the limb and trunk regions, lesions involving the brachial or pelvic plexus may be very large or

A thorough history and physical examination is fundamental to the initial assessment of patients with brachial and lumbar plexus tumors. These tumors, much like other peripheral nerve tumors, usually produce few symptoms and signs, which, if present at all, can be subtle. With a thorough history and systematic neurological examination, the salient features can be elicited. Based on the information acquired from the clinical examination, the anatomical localization can be established and the differential diagnosis formulated. This information in turn leads to directed imaging and electrodiagnostic studies, as discussed in other sections of this issue of *Neurosurgical Focus*.

Patient History

Patients with brachial and lumbar plexus tumors can present with a palpable mass that is either symptomatic or asymptomatic, sensory alterations, motor deficits, pain, visceral symptoms, autonomic dysfunction, or any combination of symptoms. The presenting complaints should be characterized with regard to onset, temporal progression, distribution, quality, severity, timing, and the presence of any aggravating or relieving factors. Particular factors to note include joint position, prolonged activity, or irritation of the lesion.

Abbreviation used in this paper: NF1 = neurofibromatosis Type 1.

even massive before they produce a notable mass. Rapid changes or fluctuations in size and any association with pain or previous trauma should be ascertained in the history. Rapid growth of the mass suggests a more malignant nature, while fluctuations in size may be associated with a cystic component or bleeding within the lesion.

Visceral Symptoms

Visceral symptoms often occur as a result of the mass effect from the tumor on adjacent structures: nerves, vasculature, lymphatic system, or viscera. For example, large sarcomas in the pelvis may erode the lower lumbar vertebrae or the pelvic rim. In addition, compression of the psoas muscles or the lumbosacral plexus can cause pain on flexion or extension of the hip joint and/or a compression neuropathy. Compression of the bladder can produce irritative symptoms of frequency, urgency, and double voiding. Partial or complete obstruction of the ureters can cause hydronephrosis and renal impairment. Compression of the rectum can result in alterations in bowel habits varying from fecal incontinence or diarrhea to constipation, while compression of more proximal bowel can result in obstruction. Compression of the lymphatics or vasculature can cause edema or ischemia of the lower extremities.

Pain Quality

Pain is usually a late complaint and may be indicative of a more ominous disease process. Pain is usually caused by the focal pressure exerted by the tumor on the nerve and adjacent tissues, with resultant swelling. Associated joint stiffness, muscle and tendon shortening, and muscle fibrosis can also be the cause. Pain commonly occurs along the distribution of the involved nerve and may also be referred to adjacent tissues.⁴ The quality can vary from an aching discomfort, usually in the early cases, to a profound neuritic pain syndrome, characterized by a severe burning pain along the distribution of the nerve and sometimes accompanied by sensory changes. A well-localized pain syndrome with a trigger point over the mass lesion (which itself may also be tender) can help localize the precise nerve involved.

Motor Deficits

Motor deficits are usually a late sign, except in cases of malignant peripheral nerve sheath tumors. The weakness is usually subtle, and the patient may describe the deficit in terms of general movements or a functional impairment. A patient may complain, for example, of difficulty with buttons, knitting, and opening containers, of having to switch to the nondominant hand to perform certain tasks, of difficulty with toe-standing, or difficulty in rising from a squatting position.⁸

Autonomic Dysfunction

Both autonomic hyper- and hypofunction can accompany brachial plexus tumors. Autonomic hypofunction can include anhidrosis, coolness of the limb, or some degree of cyanosis and swelling. Abnormal vasodilation and hyperhidrosis can be seen in circumstances of sympathetic overactivity. Reflex sympathetic dystrophy, characterized by pain, changes in skin color and temperature, and variable

atrophy of the involved muscle (due to neurogenic reasons or disuse), should be considered when burning pain is associated with autonomic symptoms. A triad of miosis, ptosis, and anhidrosis is pathognomonic for Horner syndrome, which can be seen in tumors involving the lower brachial plexus (such as Pancoast tumors).

Additional Features

Obtaining thorough medical and family histories are essential. In the medical history, special attention should be paid to any previous trauma, burns, or radiation to the area in question. Any previous surgeries or mass lesions removed should be noted. In the family history, features suggestive of neurofibromatosis should be noted, with details as to which family members and the lesions they possess (brain, spinal, or nerve tumors, multiple skin lumps, or café-au-lait spots). Some cases may be so subtle that the family members may need to be examined by the physician to resolve any doubt.

Clinical Examination

Physical Examination

There are several basic principles regarding the physical examination. The limb in question and its counterpart should be fully exposed, so that the affected limb can be compared with the unaffected one. The neurological examination should proceed in a systematic fashion proximally to distally, so as not to overlook any findings.

Cutaneous Features and Neurofibromatosis

The physical examination should begin with a thorough inspection of any stigmata of neurocutaneous diseases. The most common genetic disease predisposing to neoplasia in humans is NF1.³ The skin findings in NF1 include café-au-lait macules, dermal neurofibromas, and axillary or inguinal freckling (Fig. 1). The criteria necessary to establish a diagnosis of NF1 or NF2 can be difficult to meet because of the variable expressivity. The diagnostic criteria are listed in Table 1.⁶ Patients with NF1 have a 10,000 to 100,000 times greater risk of developing malignant changes, with up to 30% succumbing to some form of cancer compared with

FIG. 1. Photograph of a huge café-au-lait spot and multiple skin neurofibromas on the abdomen of a patient with NF1.

TABLE 1
Diagnostic criteria for NF1 and NF2

NF1: diagnosis is established if any two of the following features exist
6 café-au-lait macules >15 mm in diameter if postpubertal, >5 mm in diameter if prepubertal
≥2 neurofibromas of any type, or 1 plexiform neurofibroma
axillary or inguinal freckling
a distinctive osseous lesion, e.g., sphenoid dysplasia, congenital bowing or thinning of long bone cortex w/ or w/o pseudarthrosis
bilateral optic nerve gliomas
≥2 iris Lisch nodules
a first-degree relative w/ NF1, diagnosed by the above criteria
NF2
bilateral vestibular schwannomas demonstrated by computed tomography or magnetic resonance imaging
OR
a first-degree relative w/ bilateral vestibular schwannomas AND
i) a unilateral vestibular schwannoma
ii) any two of the following: neurofibroma, meningioma, glioma, schwannoma, posterior lens opacity

patients with NF2. Malignant transformation of neurofibromas associated with NF1 occurs in about 5% of cases, and about 50% of neurogenic sarcomas are associated with this syndrome.²

Lhermitte–Duclos disease or multiple hamartomas syndrome is a phakomatosis that includes papules on the face, oral mucous membranes, and dorsal aspect of the forearms and hands. These occur in association with tonsillar B-cell hyperplasia and benign and malignant tumors of the lung, thyroid, and gastrointestinal tract. This syndrome combines the features of lipofibromatous hamartomas and neoplasms.¹ The lesions are slow-growing and often tender tumor-like expansions, usually of the larger nerves of the upper extremities.

Examination of the Mass

The tumors can range in size from large cosmetically disfiguring plexiform neurofibromas (associated with NF1) to small and difficult-to-palpate nerve tumors. In brachial plexus lesions, the mass may cause an asymmetry of the proximal limb (Fig. 2). In such cases, the lesion’s relationship to the clavicle should be determined (above, behind, below, or all three). Measurements of the maximal dimensions should be recorded, especially for comparison at a later date, if conservative management is being considered. The overlying and surrounding skin should be inspected for any discoloration, necrosis, or associated lesions. A photograph documenting the lesion is often helpful.

The exact location of the tumor should be identified and documented, along with its relationship to any adjacent nerves, blood vessels, joints, and muscles. The mass should be inspected with concomitant movement of the joint, in flexion and extension, to determine whether there is any attachment to the joint capsule or surrounding muscles; a note should also be made if any limb or joint position makes the lesion more or less prominent.

The tumor should be palpated for range of movement, particularly in relation to the long axis of the limbs. Peripheral nerve tumors move perpendicular to the course of the nerves, with minimal movement in the nerves’ longitudinal direction. It should be noted if there is any attachment to the superficial or deeper tissues or to the surrounding structures,

FIG. 2. Photograph of a patient with a large right brachial plexus mass producing supraclavicular fullness and obvious asymmetry.

such as adjacent joints. Again, examination with concomitant movement of the joint can be helpful.

The tumor should also be palpated to determine its morphological characteristics and consistency. With gentle palpation, it is often possible to appreciate whether the surface of the tumor is smooth or rough, whether there is a single nodule only or if the tumor is lobulated, and whether the edges are smooth and well-defined or irregular and poorly defined. The consistency of the tumor should be noted as well, whether the tumor is hard or soft, and if there is any associated pulsatility or fluctuance. Finally, a little firmer palpation or a gentle tap may elicit a Tinel sign, with radiating paresthesias in the distribution of the involved nerve or nerve element. For completeness of examination, the mass should be auscultated to ensure that there is no bruit present. Soft bruits may occur in very vascular lesions.

Motor Examination

Motor findings are usually a late sign in the pathogenesis of peripheral nerve sheath tumors, except in malignant peripheral nerve sheath tumors that often present with progressive and severe pain and weakness. The motor examination should start with an inspection of any muscle bulk asymmetry. This is usually quite obvious on inspection. Specific muscles and the compartments affected should be noted. When possible, an accurate assessment of muscle bulk using a tape measure should be performed. This can be done by first marking the extremity from a fixed bone landmark so that the corresponding areas of the affected and unaffected limbs can be compared.

Motor strength testing should proceed in a systematic fashion, proximal to distal. In the upper extremity this means first examining posterior parascapular and shoulder girdle muscles before proceeding to the muscles of the arm and hand. In the lower extremity, both the anterior and posterior aspects of the patient should be examined, up to and including the gluteal region. Motor strength testing should be performed with an attempt to discriminate the given movement of the limb from the specific actions of muscles, since it is the latter that provides the precise information

needed. For example, lateral abduction of the shoulder for the first 30° is produced by the supraspinatus muscle, and the next 120° is produced by the deltoid. The movement is then completed by the medial rotation of the scapula by the parascapular muscles (trapezius and rhomboid muscles). It is essential that the strength and contribution of each muscle be assessed.

With a thorough clinical examination, the clinician should be able to localize the lesion based on simple clinical findings. For example, involvement of the sciatic nerve by a tumor may lead to a unilateral foot drop; however, the same deficit can be caused by a lesion located more proximally or distally, and this possibility must be ruled out. An upper motor neuron lesion affecting the pyramidal tract, a spinal cord lesion affecting the L-5 motor neuron pool, a spinal lesion interfering with L-5 outflow, or peripheral tumor affecting the L-5 nerve root, lumbosacral trunk, sciatic nerve peroneal division, or peroneal nerve may all cause foot drop. One can distinguish upper motor neuron from lower motor neuron diseases by evaluating reflexes and muscle tone. Examination of the common L-5 innervated muscles—for example the gluteus and posterior tibialis muscles (ankle inversion)—will help distinguish a radicular and more proximal lumbosacral plexus lesion from a more distal sciatic or peroneal lesion. The short head of the biceps femoris (innervated by contributions from the peroneal division of the sciatic nerve above the knee) may help distinguish a sciatic peroneal division tumor from the more typical common peroneal involvement at the knee level.

Motor strength should be assessed for individual muscles and not particular limb movements. For example, ankle dorsiflexion should not be recorded as such, but a grade should be assigned for the function of the tibialis anterior muscle. Grading of muscle strength is usually performed according to the standard British Medical Research Council Motor Grading Scale.⁵ The major shortcoming of this system is the lack of discrimination among the ranges of muscle strength that exist between antigravity function (Grade 3) and normal motor function (Grade 5). Grade 4 strength (active movement against gravity and resistance) may represent disparate levels of function. For practical reasons, therefore, Grades 4–, 4, and 4+ have evolved in an attempt to discriminate between severe, moderate, and mild weakness, respectively. Note, however, that the likelihood of poor inter- and intrarater reliability is high with this system.

While the scope of this article does not permit a detailed analysis of the key steps in testing each muscle, certain observations are included here. Certain motor findings are pathognomonic for the involvement of a particular nerve element. Consider the patient with an upper and middle trunk brachial plexus tumor. The typical waiter's tip posture is apparent, with the shoulder internally rotated and the arm held tight against body (as the actions of the deltoid and spinatei are absent), elbow extended (biceps paralyzed), forearm pronated with the hand facing backward (as the supinator is paralyzed), and the palm up (unopposed finger flexion from extensor paresis).

Next, deep tendon reflexes should be elicited, comparing the affected to the unaffected side. Myotatic reflexes are extremely sensitive indicators of peripheral nerve pathology. It is not uncommon to find loss or diminution of ankle reflex in a patient with buttock or midhigh level sciatic nerve tumor infiltration who has complete peroneal division

involvement clinically but demonstrates nothing else (except an absent ankle reflex) with respect to the posterior tibial division.

Sensory Examination

The sensory examination is important because this component may be the only positive finding in the clinical examination, especially in an early presentation. Light touch, pinprick, two-point discrimination, vibration, and proprioception should all be tested. It is important to note that most sensory features due to peripheral nerve tumors are usually subtle, ill-defined, and variable. A profound sensory deficit in light of an early presentation is highly suspicious for a malignant lesion. It is also important to note that the lack of sensory findings does not necessarily rule out the diagnosis of a peripheral nerve tumor.

To test for sensory alteration, instruct the patient to close his eyes and then point to the area that is stimulated using light touch with a blunt object such as a dull pen tip. Although this technique will allow mapping of areas of poor or absent sensation, most patients with nerve tumors exhibit a much smaller degree of sensory alteration. Having the patient compare simultaneous stimulation can allow a more subtle loss of sensation to be discerned. Stroke the area of altered sensation simultaneously with the corresponding area on the other limb. Do this gently, using the same amount of force, and have the patient comment on any difference in the sensation. Attempts to validate such a simultaneous sensory testing paradigm using a 10-point analog scale developed by Strauch⁹ have been reported.⁷ A more thorough sensory examination can be conducted with rudimentary instruments, for example, a fresh safety pin, cotton, a 128-Hz tuning fork (for vibration and temperature testing), and either a paper clip or blunt tip callipers.

A key point in the sensory examination is that sensation should be examined in the autonomous zones of innervation where there is the least or no likelihood of sensory overlap from the adjacent nerves. Standard autonomous zones for the ulnar nerve are the volar aspect beyond the distal interphalangeal joint of the little finger. For the median nerve, it is the volar aspect of the fingers distal to the distal interphalangeal joint of the index finger and the interphalangeal joint of the thumb. While the anatomical snuffbox is considered the autonomous sensory distribution of the radial nerve, there can be variable overlap from other cutaneous nerves, for example, the lateral antebrachial cutaneous nerve.

Other Aspects of the Physical Examination

To complete the physical examination, the limbs should be examined for presence or absence of autonomic activity. Note the color, temperature, and sweating behavior and atrophic changes in skin organs and nail beds. The triad of miosis, ptosis, and anhydrosis constitutes Horner syndrome, the presence of which helps to confirm the involvement of the lower brachial plexus at a very proximal spinal nerve or lower trunk level.

Compression of a nerve often results in hypersensitivity to mechanical stimulation. The Tinel sign can be elicited by percussing over the injured nerve to produce electric shock-like sensations or paresthesias in the distribution of the nerve. The Tinel sign is often present overlying the

nerve in the area of entrapment of tumor growth and can persist for a long time, sometimes indefinitely, overlying the area of previous nerve injury.

Conclusions

The presenting symptoms and signs of peripheral nerve tumors are usually few, and if present, can be subtle. Pain, paresthesias, and/or a palpable mass are typical presenting complaints, with significant motor deficits and a progressive course of pain suggestive of a malignant pathological process. A detailed family history should always be obtained, looking for predisposing disorders such as neurofibromatosis. A systematic physical examination looking for any cutaneous and autonomic features, motor and sensory deficits, and Tinel sign should be conducted. Included as well should be a thorough examination of the mass lesion for form, consistency, and mobility, with an irregular, firm, and immobile mass suggestive of a malignant lesion. With a thorough history and physical examination, based on an understanding of the anatomy, the clinician is rewarded with enough information to pinpoint the exact anatomical location and formulate a differential diagnosis for the lesion.

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