Arch. Psychiat. Nervenkr. 227, 33—37 (1979)

Archiv für Psychiatrie und Nervenkrankheiten
Archives of Psychiatry and Neurological Sciences
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High Cervical Neurinoma (C1/C2) Diagnosed Falsely as Multiple Sclerosis Because of Trigeminal Neuralgia

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Summary. Remitting paresis of the left leg accompanied by left trigeminal neuralgia led to the diagnosis of multiple sclerosis in a 46-year-old woman.

Over the following 6 years, an incomplete syndrome of the spinal cord developed along with bilateral trigeminal pain. Neuroradiological and neurosurgical exploration revealed a neurinoma located ventrolaterally at C1/C2 on the left side.

It is emphasized that since trigeminal fibres descend as far as the upper part of the C2 segment, trigeminal neuralgia should not be considered as an exclusively supraspinal symptom.

Key words: Trigeminal neuralgia – Multiple sclerosis – Cervical neurinoma – Differential diagnosis.

Zusammenfassung. Es wird über eine 46jährige Patientin mit einem hochcervicalen Neurinom (C1/C2) berichtet, bei der aufgrund des initialen Verlaufes mit remittierender Beinlähmung und Trigeminusneuralgie die Diagnose einer Multiplen Sklerose gestellt wurde.

In den folgenden 6 Jahren entwickelte sich ein inkomplettes hohes Querschnitssyndrom mit bilateralen Trigeminusschmerzen. Neuroradiologische und neurochirurgische Exploration ergaben ein links ventro-lateral gelegenes Zwerchsackneurinom in Höhe C1/C2.

Es soll hervorgehoben werden, daß eine Trigeminusneuralgie nicht von vornherein als ein supraspinales Symptom gewertet werden darf, da Trigeminusfasern bis zum C2-Segment absteigen.

Schlüsselwörter: Trigeminusneuralgie – Multiple Sklerose – Zwerchsackneurinom – Differentialdiagnose.

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0003-9373/79/0227/0033/$ 01.00

Introduction

The fact that slowly growing intraspinal tumours may occasionally be diagnosed falsely as multiple sclerosis (MS) is well documented in the literature [2, 5, 8, 9, 10]. The justification for reporting yet another case of this sort is the rare and remarkable concurrence of symptoms—a remitting attack of leg paresis together with trigeminal neuralgia (TN)—which (obviously taken as a multifocal process) led to the initial diagnosis of MS.

TN occurs 150—300 times more frequently in MS patients than in the remaining population (for complete references, see Huhn et al., 1973). In view of this manifest syntropy, it is not surprising that in patients with remitting spinal symptoms TN might be readily accepted as a supraspinal sign supporting the diagnosis of MS. In cases of TN/MS syntropy, foci of demyelinization are found especially at the entrance of the sensory root of the pons [4]. The aim of this short casuistic report is to stress the dangers of asigning TN a supporting role in the diagnosis of MS and to emphasize that apparent dissemination of symptoms in time and space, a typical feature of MS, can also result from spinal tumours.

Case Report

Personal History. Apart from an occasional bout of lumbalgias the 46-year-old woman had been in good health until 6 years ago when she experienced a sudden weakness in the left leg. Her condition improved after treatment in a health resort. One year later, however, paresis of the left leg recurred and was accompanied by attacks of neuralgic pain in the region of the lower left canine tooth. This pain could not be triggered by touch or jaw movements. The patient entered a neurological clinic where the symptom constellation of remitting spastic paresis of the left leg together with trigeminal neuralgia led to the diagnosis of MS.

Cerebrospinal fluid (CSF) contained 12 cells per cmm as well as a slightly elevated total protein content and increased gamma globulins. Subsequently, spastic plegia of the left leg, spastic paresis of the left arm, and weakness in the right extremities developed. TN was now experienced also in the region of the right mandibular, but was still more pronounced on the left side. Unable to walk, the patient was admitted to the same neurological hospital again 3 years later, at which time CSF contained 3 cells per ccm and the total protein content was at the upper limit of the normal range. Although treatment with carbamazepine partially alleviated the paroxysmal and neuralgic character of the facial pain, a slight but constant pain remained in the pre- and subaural regions and in the upper neck. Cervical X-rays revealed some degeneration and spondylarthrosis at C5/6.

Neurological Findings

When the patient entered our hospital, she showed an almost complete spastic tetraplegia; deep tendon reflexes were hyperactive and Babinsky responses were present. Sensory examination revealed modest deficits of pain and temperature perception in the trunk and extremities, whereas touch and vibration were perceived normally. Movement of the head was still possible, though slightly painful. The ocular fundi showed neither papilloedema nor optic atrophy. There was a moderate decrease in pain perception in the region of the mandibular and maxillary branches of the left trigeminal nerve, but corneal reflexes were not

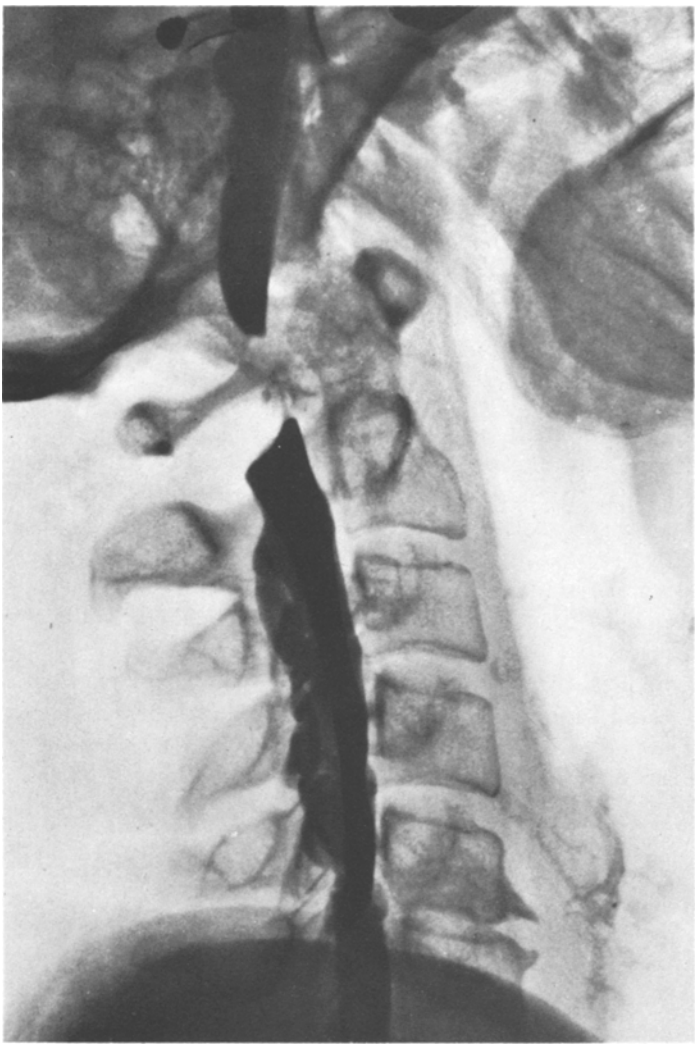

Fig. 1. Cervical myelogram revealing a space-occupying lesion at the C1/C2 level

impaired. The remaining cranial nerves functioned normally, including the XIth supplying the sternocleidomastoid muscle. There was relative incontinence of the bladder. The vital capacity was reduced to lower levels of resting respiration. The patient was fully oriented and intellectually intact.

Laboratory Findings and Neurosurgery

Lumbar puncture yielded clear spinal fluid under normal pressure; 3 cells, 104 mg/dl protein content, 13.5% gamma globulins. A computed tomogram of the head revealed no abnormalities within the region of the brain stem. Cervical

X-rays indicated spondylarthrosis at C5/C6. Duroliopaque myelography demonstrated the presence of an intraspinal space-occupying process located ventrolaterally at the level of the left C2 foramen which was thus enlarged (see Fig. 1). A tumour removed from this region was subsequently shown by histology to be a neurinoma.

Discussion

Pain and motor disturbances are by far the most common complaints [8] in patients with spinal neurinomas [7]. The initial failure to detect a high cervical neurinoma in the present case must be related to the fact that it produced a trigeminal pain syndrome falsely attributed to a brain-stem lesion. There would appear to be two pathogenic mechanisms which could possibly have contributed, perhaps conjunctly, to a spinally mediated trigeminal pain syndrome: (a) affection of the first (and second) cervical sensory roots ascertained during the operation could well have produced an irritative lesion, leading to a type of cervical neuralgia. The sensory distribution of the region of the lower jaw angle would comply with this assumption; (b) the typical character of paroxysmal pain and its localisation deep within the mandibular teeth suggest that the pathogenic mechanism could also have been an irritative lesion of the caudal part of the spinal trigeminal nucleus.

It must be remembered, firstly, that a considerable number of trigeminal fibres descend as far as the upper part of the C2 segment, building the caudal part of the spinal trigeminal nucleus, and secondly, that dorsal root fibres of the upper cervical nerves also end in the nucleus of the spinal trigeminal tract [6]. From neurophysiological and clinical work, it is generally accepted that the most caudal parts of this nucleus and its continuation into the dorsal horn at the level of C1 and C2 are the regions that receive the fibres mediating pain [1, 4]. This view is supported clinically by the medullary trigeminal tractotomy performed by Sjöqvist [3, 11]: since the majority of fibres in the caudal part of the spinal trigeminal tract are thin and because pain is assumed to be mediated by thin and partly unmyelinated fibres, Sjöqvist transected the spinal trigeminal tract at the level of the obex in patients with TN; the patients were usually relieved of the pain attacks and there was no longer any pain in the skin and mucous membranes of the face. One final point should be emphasized since it might be a further reason why the cervical neurinoma was overlooked: clear-cut, spontaneous remissions of motor deficits are not specific for MS; they also occur, for example, with mechanically induced spinal tumours [9, 10]. One essential difference between spontaneous remissions in these two cases, however, is that in mechanically induced spinal tumours exacerbations always affect the same system, whereas exacerbations in MS cause deficits in different systems [9].

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Received March 8, 1979