Pudendal neuralgia due to solitary neurofibroma

Case report

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An unusual case of pudendal neuralgia due to a solitary neurofibroma of the perineal region is described. The authors outline the long clinical history. There was complete pain relief after removal of the lesion.

KEY WORDS · pudendal neuralgia · solitary neurofibroma · neurofibromatosis · von Recklinghausen's disease · pudendal nerve · peripheral nerve

PUDENDAL neuralgias requiring direct surgical treatment are rare. We report the case of a patient who underwent surgery for a neurofibroma of the perineal region after presenting with the classic neuralgic syndrome of the pudendal nerve.

Case Report

This 28-year-old woman came for our evaluation in December, 1978. She had an 18-month history of paroxysmal attacks of pain involving the right perineal region. Pain was described as occurring laterally and radiating to the ipsilateral labium majus, following the distribution of the pudendal nerve. These neuralgic episodes had gradually increased over the months both in frequency and intensity; they were reported as spontaneous as well as elicited by the lightest touch. Sitting and coitus had become impossible. Both rectal and vaginal explorations caused unbearable pain.

The patient had undergone a number of local anesthetic blocks among other treatments, with poor or transient relief. Psychotherapy had been ineffective. Family and medical history was noncontributory. No stigmata of multiple neurofibromatosis (von Recklinghausen's disease) were detected.

Physical examination revealed a localized area in the external aspect of the right perineal region, gentle manipulation of which triggered intense pain radiating to the midline and lasting 5 to 6 seconds. Pelvic roentgenograms were normal.

Surgical exploration was carried out on December 29, 1978. The perineal fascia was incised and a gray-red pea-sized nodule was easily identified on macroscopic examination. The lesion appeared not to be encapsulated, but showed lateral mobility when displaced by the finger. No connections with nerve bundles were apparent, and the nodule was removed. Histological sections demonstrated that the tumor was neurofibromatous in nature (Fig. 1).

The patient was discharged symptom-free, and has remained well for 2 1/2 years since surgery.

Discussion

Neuralgic syndromes affecting the perineal region are ascribed to involvement of the pudendal nerve or, less commonly, of the third sacral root. Painful paroxysms are more frequently “primary” or “pure” (idiopathic) pudendal neuralgia. Rarely, pain is symptomatic of vertebral malformations, such as spina bifida or tumors of the cauda equina; sometimes, it follows sacral fractures or surgery of pelvic organs.

Peripheral neurofibromas most frequently involve the nerve trunks of the upper extremity, particularly the ulnar and radial nerves. These tumors are usually found in patients with von Recklinghausen's disease. They often reach considerable size before producing pain, and cause little or no neurological deficit.

We have categorized the tumor removed from our patient as a “solitary” neurofibroma, using this term for lesions that are found in the absence of stigmata of von Recklinghausen's disease. They are entirely
benign, display a slowly progressive history, and are painful only in rare cases.\(^3,6\)

Solitary neurofibromas have become recognized as a distinct entity since 1962, when Heard\(^2\) reported on 46 such peripheral tumors. There were 35 superficial and 11 deep lesions. Nine patients with superficial tumors suffered pain, which was strictly local and exhibited no radiating features. In contrast, two patients with deep tumors experienced paroxysmal paresthesias in the distribution of the nerve involved; these two cases may be regarded as having neuralgic syndromes and resemble our case.

Despite the current acknowledgment of solitary neurofibromas as distinct nosological entities, Russell and Rubinstein\(^7\) warned that these lesions must always be regarded with caution, since other peripheral manifestations of multiple neurofibromatosis may appear many years after discovery of the first lesion. Therefore, our patient is being followed carefully in order to recognize any delayed evidence of this disease.

We wish to emphasize the long interval that elapsed after onset of the pain before adequate treatment was instituted. The possibility of such a distressing course should be kept in mind, especially in the case of patients who may be labeled as suffering from “psychalgia.”\(^4,6\)

**References**


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