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Central neuropathic itch as the presenting symptom of an intramedullary cavernous hemangioma: Case report and review of literature

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Keywords

Itch; Spinal cord; Cavernous hemangioma; MRI.

1. Introduction

Itch, or pruritus, is an unpleasant cutaneous sensation associated with a pressing urge to scratch. It is one of the most frequent symptoms in dermatology (e.g., in inflammatory skin disorders) and general medicine (where it can be related to cholestasis, diabetes, uremia and lymphoma).

Itch may also be observed in both peripheral and central nervous system (CNS) lesions, but it is rarely the isolated or the main symptom of neurological disorders, especially in cases of spinal cord diseases.

We report a case of localized itch as an isolated symptom of a spinal cavernous hemangioma. Surgical excision allowed relief from symptom. We reviewed different theories on the generation of itching and discussed its association with cervical lesions.

2. Case report

A 24-year-old woman presented to our neurological clinic complaining of a continuous itch, ongoing for 5 years, in the dorsal area of her right forearm, from the elbow to the little finger of the hand. Dermatological investigations found only excoriations due to scratching, without any pathological condition. The neurological examination showed only Lhermitte's sign accompanied by a burning sensation in the feet. Neither sensory deficit nor anomaly of sensory processing, such as allodynia or alloknesis, were present. Treatment with gabapentin was unsuccessful.

Electromyography and nerve conduction studies of the patient's right arm were normal. Rightsomatosensory evoked potentials (SSEP) from ankle stimulation showed a central conduction time in the normal range, but with a significant asymmetry between the left and right sides. MRI disclosed a 5-mm C6–C7 intramedullary round lesion, situated on the right posterior horn. The lesion showed a core of increased intensity surrounded by a low signal rim due to hemosiderin in T2-weighted images (*Fig. 1 and Fig. 2*). Cerebral MRI results were normal.



Fig. 1: Sagittal T2-weighted MRI scan showing a C6–C7 intramedullary lesion with a core of increased intensity surrounded by a low signal rim.

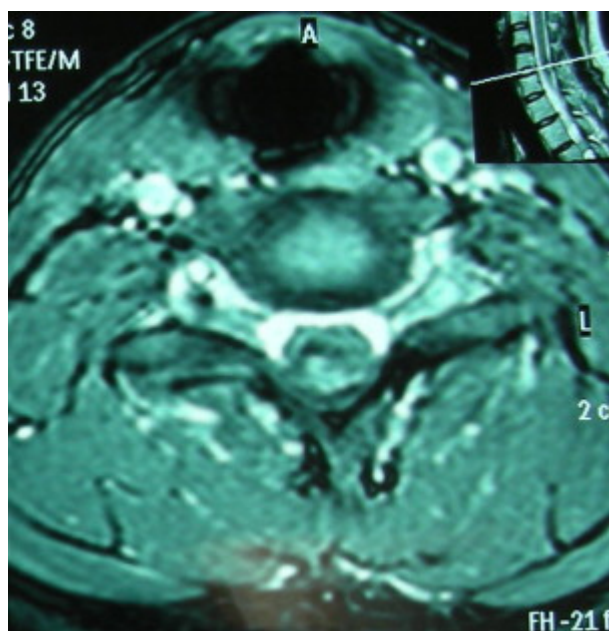


Fig. 2: Axial T2-weighted MRI scan showing the posterior horn location of the lesion.

A C6–C7 laminotomy was performed and the lesion was excised under intraoperative ultrasound guidance. Pathological examination showed enlarged thin-walled blood vessels and hemosiderin-laden macrophages consistent with a cavernous hemangioma.

A complete excision was achieved (*Fig. 3*), and the neurological status remained unchanged. At a 1-year follow-up, itching sensation was described as intermittent, markedly reduced in intensity, and limited to a tiny area of the forearm skin. The pre-operative asymmetry of SSEP was no longer detectable after surgery.

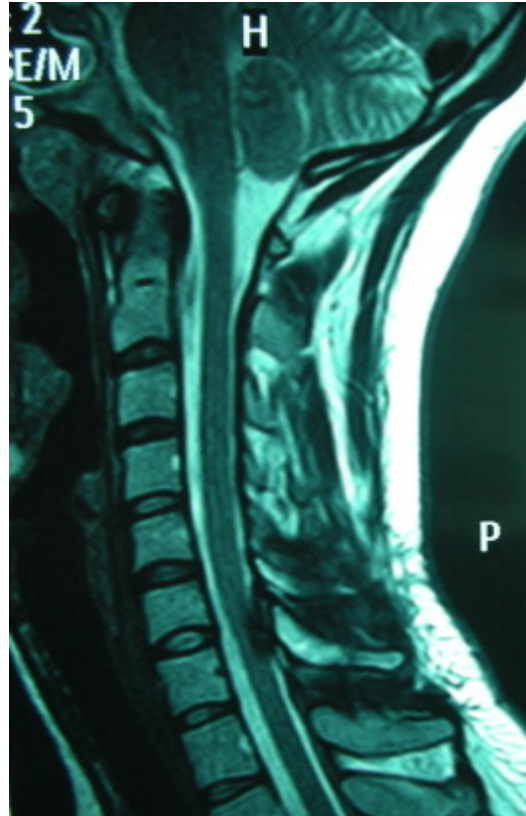


Fig. 3: Postoperative sagittal T2-weighted MRI scan showing complete removal of the lesion.

3. Discussion

Spinal cavernomas represent approximately 5% of all CNS cavernous malformations. The clinical presentation is mainly characterized by motor deficits with or without sensory loss. Radicular signs and/or symptoms may also be observed.

The natural history usually consists of micro-bleedings and subsequent gliosis that lead to a progressive myelopathy. Surgical excision can prevent progressive neurological decline and a favorable outcome is generally observed. In contrast, chronic myelopathy is not reversible and radicular pain can be relieved in less than half of patients. Macrohemorrhages and acute onset of symptoms may also appear and lead to irreversible neurological deficits.

Central neuropathic itch as an isolated symptom of spinal cavernous hemangioma has never been reported, although Dey et al. have described three cases of spinal cavernomas with itch associated to radicular pain, paresthesias and motor deficits^[1]. In two of these cases, the diagnosis was based on neuroradiological findings. The third patient presented with sudden weakness in the arm,

radicular pain and itch due to a hemorrhagic cervical cavernoma. A surgical excision was performed and allowed relief from the symptoms; nevertheless, itch recurred 2 years later.

We report a lasting itch in the C6–C7 dermatome as an isolated symptom. The improvement observed as result of removal of the cavernoma suggests a direct action of the spinal lesion in generating itch. The clinical benefits were maintained at a 1-year follow-up.

Several theories have been proposed to explain the neurophysiological basis of itch. According to the “intensity theory”, itch is considered to be closely related to pain, and it may be generated by light stimulation of nociceptors.

On the other hand, the “specificity theory” considers itch as a symptom generated by a specific and independent circuit. This hypothesis is supported by the recent identification of distinct pathways for itch and pain in cats and humans^{[2], [3]}.

Specialized C-fibers, distinct from those associated with the mediation of pain, may be activated by prurigenic mediators of the inflamed skin. The second-order sensory neurons are placed in the lamina I dorsal horn, where they most likely form an integrated system with the pain pathway that is mediated by inhibitory interneurons^[4].

Common experience and experimental results demonstrate the antagonistic interactions between itch and pain; in fact, scratch-induced pain can abolish itch.

Usual causes of itch are peripheral nerve or spinal root injuries due to Herpes Zoster or keloid scars with nerve entrapment. In these cases, pruritus is localized and accompanied by superimposed pain. Occasionally, localized itch and pain may be observed in case of nerve damage caused by degenerative vertebral diseases.

Itch in brain or spinal cord lesions is rare. Nineteen cases of central neuropathic itch (CNI) are reported in association with brain tumors, stroke, multiple sclerosis, and abscesses^[5]. In these conditions, itch is usually accompanied by other symptoms. CNI due to spinal lesions is far less frequent. Spinal plaques of multiple sclerosis can generate paroxysmal itching and pain in the same skin areas. Localized pruritus was reported as presenting symptom in a few spinal cord tumor patients and after spinal cord injury^[6].

In our case, the cavernoma might have caused a predominant degeneration of the pain over the itch neurons. Therefore, we speculate that the pain-itch network may be biased toward a predominance of the neurons generating itch. Moreover, deposition of hemosiderin and gliosis due to microhemorrhages may have created an excess of electrogenesis in the dorsal horn that most likely led to activation of second-order neurons that project via the contralateral spinothalamic tract as a central neural pathway for itch^[1].

The occurrence of localized itch without pain due to a spinal cord lesion not involving nerves or spinal roots might support the existence of distinct neural circuits for itch and pain in the dorsal horn.

4. Conclusion

A central origin of itch must be suspected if dermatological or general medical causes are excluded. Therefore, we suggest performing a cerebrospinal MRI in cases of cryptogenic pruritus, especially if a clear root distribution is present. Our anecdotal case highlights the role of surgical excision in relieving itch.

References

- [1] D.D. Dey, O. Landrum, A.L. Oaklander, Central neuropathic itch from spinal-cord cavernous hemangioma: a human case, a possible animal model, and hypotheses about pathogenesis, *Pain*, 113 (2005), pp. 233-237.
- [2] M. Schmelz, R. Schmidt, A. Bickel, H.O. Handwerker, H.E. Torebjork, Specific C-receptors for itch in human skin, *Journal of Neuroscience*, 17 (1997), pp. 8003-8008.
- [3] D. Andrew, A.D. Craig, Spinothalamic lamina I neurons selectively sensitive to histamine: a central neural pathway for itch, *Nature Neuroscience*, 4 (2001), pp. 72-77.
- [4] A. Binder, J. Koroschetz, R. Baron, Disease mechanisms in neuropathic itch, *Nature Clinical Practice. Neurology*, 4 (2008), pp. 329-337.
- [5] S. Canavero, V. Bonicalzi, B. Massa-Micon, Central neurogenic pruritus: a literature review, *Acta Neurologica Belgica*, 97 (1997), pp. 244-247.
- [6] D. Magilner, Localized cervical pruritus as the presenting symptom of a spinal cord tumor, *Pediatric Emergency Care*, 22 (2006), pp. 746-747.