KJP The Korean Journal of Pain
| Editorial |
Korean J Pain 2017 July; Vol. 30, No. 3: 163-164
pISSN 2005-9159 eISSN 2093-0569
https://doi.org/10.3344/kjp.2017.30.3.163

Diagnosis of occipital neuralgia due to upper cervical chordoma

Department of Anesthesiology and Pain Medicine, Yonsei University Wonju College of Medicine, Wonju, Korea

Young Bok Lee

Chordoma is a relatively rare tumor that primarily occurs in the axial skeleton. Approximately 50% of chordoma originate from the sacrococcygeal area and 6% of chordoma arise from the cervical spine [1,2].

Chordoma originated from the cervical spine as a slowly growing tumor, which makes the tumor symptomatic at relatively later stages. Upper cervical tumors often are accidentally discovered during investigation of cervical pain or radiculopathy. The tumor causes compression of the cervical nerve roots or gives rise to referred pain in the neck due to invasion into the cervical facet joints. Occasionally, the lesion is mistaken for a facet joint pathology or cervical disc herniation. Although the tumor is histologically benign, it is clinically malignant as it infiltrates the neighboring tissues, such as bone, lymph nodes, skin, liver, and brain [3].

Upper cervical chordoma frequently becomes clinically symptomatic when it has grown considerably in size. Nerve compression or intraspinal invasion may result in severe neurologic complications. Upper cervical tumors have invaded the second cervical spine root, and neuralgia of the greater occipital nerve has led to occipital headaches. When the mass invades the atlanto-occipital joint or the zygapophysial joint of the second and third cervical vertebrae, referred pain is experienced in the lower occipital and posterior upper cervical areas. If the symptoms persist without appropriate treatment, they may progress to neuropathy. Once the neuralgia becomes a neuropathy, symptoms may persist despite tumor resection.

Upper cervical chordoma is included in the diagnosis of lesions found in the upper cervical region. Imaging is particularly important for the diagnosis and management in patients with symptomatic lesions. The tumor usually shows inhomogeneous contrast enhancement of varying intensity on CT and MRI. While no imaging features are pathognomonic, the CT findings are classical for chordoma. MRI may underestimate soft tissue invasion as found on operative assessment. It should be included in the differential diagnosis for pathologic masses with symptoms. Definitive diagnosis of the chordoma requires histologic evaluation in which physaliferous cells with cytoplasmic vacuoles and positive periodic acid-Schiff (PAS) staining are observed [4].

REFERENCES

1. Currier BL, Papagelopoulos PJ, Krauss WE, Unni KK, Yaszemski MJ. Total en bloc spondylectomy of C5 vertebra for chordoma. Spine (Phila Pa 1976) 2007; 32: E294-9.
2. Boriani S, Chevalley F, Weinstein JN, Biagini R, Campanacci L, De Iure F, et al. Chordoma of the spine above the sacrum.

Received June 24, 2017. Accepted June 26, 2017.
Correspondence to: Young Bok Lee
Department of Anesthesiology and Pain Medicine, Wonju College of Medicine, Ilsanro 20, Wonju 26426, Korea
Tel: +82-33-741-1536, Fax: +82-33-742-8198, E-mail: yblee@yonsei.ac.kr

Treatment and outcome in 21 cases. Spine (Phila Pa 1976) 1996; 21: 1569–77.
3. Jiang L, Liu ZJ, Liu XG, Ma QJ, Wei F, Lv Y, et al. Upper cervical spine chordoma of C2–C3. Eur Spine J 2009; 18: 293–8.
4. Bjornsson J, Wold LE, Ebersold MJ, Laws ER. Chordoma of the mobile spine. A clinicopathologic analysis of 40 patients. Cancer 1993; 71: 735–40.