Editorial

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Sacral Chordoma: Challenging for Resection Margin

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A chordoma is a rare, bone neoplasm that accounts for 1%–4% of all bone neoplasms [1]. Although it considered to be a low-grade neoplasm with a slow-growing pattern, its clinical course is very aggressive, with a locally-invasive, highly-recurrent character [2]. It has been reported to be predominant in men, and its peak incidence is between 50–60 years of age [3]. It arises from the sacrococcygeal region in approximately 40%–50% of cases, from the base of the skull (spheno-occipital/nasal) in 35%–40% of cases, and from vertebral bodies in 15%–20% of cases [2, 4, 5].

A chordoma usually present clinically with pain as the cardinal symptom. Neurologic symptoms tend to vary based on the location of the lesion. Jeys et al. [6] reviewed 33 patients with a chordoma arising from the sacrococcygeal region. Pain, typically dull and worse with sitting, was the most common presenting symptom in 85% of the patients. Pain commonly developed in the lower back and the sacrum. Other painful regions were the legs, buttocks, and the perianal and perineal regions. The classic symptoms of cauda equine (saddle anesthesia, leg parathesia, motor weakness, bladder dysfunction, bowel dysfunction) occurred in 70% of the patients. Urinary incontinence and outflow obstruction were frequent bladder dysfunction symptoms. The majority of bowel dysfunctions were due to constipation.

Unfortunately, these symptoms are nonspecific in nature, and a chordoma has a slow-growing pattern with an insidious onset of symptoms. Therefore, it is often found at the late stages of the disease. Jeys et al. [6] reported that the mean duration of symptoms prior to diagnosis was 120 weeks (2.3 years; range, 0.5–8.0 years). The mean maximum tumor size at resection was 8.3 cm, with a mean volume of 614 cm^3.

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Differential diagnosis is important for the surgical and the post-surgical management of a chordoma. The accurate diagnosis of tumors is of valuable prognostic significance. Fine-needle aspiration biopsy or core-needle biopsy in the case of bony lesions has been suggested to be the most oncologically sound approach to establish a diagnosis before resection, with care to avoid tumor seeding [5, 7]. The excellent capabilities of magnetic resonance imaging and computed tomography allow precise delineation of tumors in terms of volume and anatomic characteristics, and aid in diagnosis [8]. In this study, three of eight patients were misdiagnosed and had undergone primary surgery with an R1 margin at other hospitals. After re-resection, all three cases had recurrences. On the contrary, four patients had been diagnosed with preoperative core-needle biopsy and underwent primary surgery with a R0 resection. One of the four patients had a local recurrence after 18 months. The patient had an initial 12.5-cm × 13.0-cm tumor infiltrating the mesorectum [9].

With early accurate diagnosis, a complete *en bloc* resection with a R0 resection margin is a key point in the treatment of a chordoma. Rates of local recurrence, as well as survival, appear to depend on the achievement of a R0 resection margin. Tzortzidis et al. [10] reported that aggressive surgical approaches to achieve total resection resulted in long-term control in 50% of the cases. Aggressive surgical resection and reconstruction may require a multidisciplinary team approach.

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