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SURGICAL MEDICAL SCIENCES / CERRAHİ TIP BİLİMLERİ

Current Approaches in the Diagnosis, Treatment and Management of Sacral Chordomas

Sakral Kordoma Tanı ve Tedavisinde Güncel Yaklaşımlar

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Abstract

Objectives: Sacral chordoma (SC) is a rare, aggressive tumor with a high recurrence rate and originates from notochordal remnants. SC is difficult to differentiate with its non-specific symptoms such as lower back pain. Surgical treatment and radiotherapy are first-line treatments. Resection with wide margins is vital for the prevention of recurrence.

Materials and Methods: Nine patients diagnosed with SC between January 2014 and December 2021 were retrospectively analyzed. All adult patients with SC were included in the study. Pediatric population was excluded from the study. Pathology, radiology and surgical records of the patients' were used. Mean age was 53.2.

Results: Two out of 9 patients died during follow-up and 5 patients had local recurrence. No metastasis was observed.

Conclusion: Current evidence shows that surgical treatment is necessary for SC's treatment. Radiotherapy is also an important aspect of the treatment. Existing evidence should be meta-analyzed for a better understanding of SC's treatment and outcomes.

Key Words: Chordoma, Sacrum, Recurrence

Öz

Amaç: Sakral kordoma (SK), yüksek nüks oranına sahip nadir ve agresif bir tümördür ve notokord kalıntılarından kaynaklanır. SK, alt sırt ağrısı gibi spesifik olmayan semptomlarıyla diğer patolojilerden ayırt edilmesi zor bir durumdur. Cerrahi tedavi ve radyoterapi birinci basamak tedavilerdir. Geniş sınırlarla rezeksiyon, nüksün önlenmesi için öneme sahiptir.

Gereç ve Yöntem: Ocak 2014 ile Aralık 2021 tarihleri arasında SK tanısı konmuş 9 hasta retrospektif olarak analiz edildi. Tüm erişkin hastalar çalışmaya dahil edildi. Pediatrik popülasyon çalışma dışı bırakıldı. Hastaların patoloji, radyoloji ve cerrahi kayıtları kullanıldı. Ortalama yaş 53,2 idi.

Bulgular: Dokuz hastanın takip süreci boyunca 2'si hayatını kaybetti ve 5 hastada lokal nüks görüldü. Metastaz gözlenmedi.

Sonuç: Mevcut kanıtlar, SK tedavisi için cerrahi müdahalenin gerekliliğini göstermektedir. Radyoterapi de tedavinin önemli bir kısmını oluşturmaktadır. SK tedavisinin ve sonuçlarının daha iyi anlaşılması için mevcut kanıtlarla yapılacak meta-analize ihtiyaç duyulmaktadır.

Anahtar Kelimeler: Kordoma, Sacrum, Nüks

Introduction

Sacral chordoma (SC), also known as notochordal sarcoma, is a rare and malignant tumor of the sacral axial skeleton with a high recurrence rate whilst being the most common primary malignancy of the sacral region (1,2). SC's origin is unknown. However, it is believed that it originates from notochordal remnants (3,4). It is most commonly seen in the axial skeleton and skull base (4). Most of the SC are sporadic but T (brachyury) gene duplication supports familial chordoma diagnosis (5).

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SC usually presents with non-specific symptoms such as pain in the lower back or gluteal area and it is hard to distinguish from other sacral pathologies (6). In addition to lower back pain, symptoms regarding defecation and urination are frequently seen in the patients. Hence, early diagnosis is very difficult while being very important for effective treatment. Mostly, diagnosis is delayed until bladder or bowel function is compromised or the mass to be detected by a rectal or vaginal examination. With today's advanced imaging techniques, we are able to detect these masses easily and they help us understand the tumors' characteristics and anatomical features (7).

SC has a genetic perspective which has yet to be discovered. Up-regulation of *sphingosine kinase 1* gene is associated with the development of SC, high recurrence rates and local invasion (8).

In SC's surgical treatment, wide resection is remarkably important for recurrence. It is proved that tumors which are not resected with wide margins have high recurrence rates (9). On the other hand, wide resection margins can cause harm to nerve roots, pelvic bone and supportive ligaments which will consequently cause long term disabilities (10).

Treatment for SC also includes neoadjuvant and adjuvant radiotherapy. Some studies confirmed radiotherapy and surgery combination lead to low recurrence rates compared to surgical treatment alone (11,12). Literature shows particle therapy is more effective than conventional radiotherapy (13). Nishida et al. (14) revealed that carbon ion radiotherapy is more effective in preventing recurrence and preserving bowel and bladder functions than surgery (13). A limitation of this study is the sample size is small. We believe there is a need for more evidence on this subject. Currently, the combination of surgical treatment and radiation therapy is the most effective treatment plan according to the literature (14).

Materials and Methods

We retrospectively analyzed 9 patients who were operated on for SC at our institution between January 2014 and December 2021. All adult patients with SC were included in the study. Pediatric population was excluded from the study. Pathology, radiology and surgical records of the patients' were used. Ethical approval was obtained from Ankara University School of Medicine Human Research Ethics Committee (approval no: 2022000106-1, date: 09.03.2022).

There was 3 female (33%) and 6 male (66%) patients in this study. Mean age was 53.2 years (18-75). 1 patient (11%) was operated in our institution for recurrence. Only biopsy and tumor ablation were done on 2 patients (22%). Tumor size was calculated using patients' preoperative magnetic resonance imaging or computed tomography. Mean tumor size was 54.5 x 48.7 x 39.3 mm (Transverse x Craniocaudal x Anteroposterior).

We used posterior approach to the sacrum and sacrectomy was performed in all cases. All cases were operated with the same technique and the same team.

Statistical Analysis

All statistical analyses were done using SPSS software (version 22.0; SPSS, Chicago, IL, USA) (15).

Results

Patients' characteristics are shown in Table 1. Follow-up time was described as from the time of surgery to the last check-up or the time of death. All of the patients were followed up and the mean follow-up time was 46.6 months (18-72). No local incision site complications were seen during follow-ups. The mean duration of hospital stay was 16 days (2-20).

The most common symptom was back and leg pain (Table 2) and one patient's diagnosis was made incidentally after imaging

Table 1. Characteristics of study population

Patient	Age	Sex	Delay*	Tumour location	Tumour size (TVxCCxAP)	RT	Resection	Complication	Recurrence	Time to recurrence
1	75	F	36 months	S2-S3	105x130x80	No	STR	Incontinence	Yes	1.5 years
2	58	M	1.5 months	S2	130x110x40	Yes	STR	Rectum perforation	Yes	5 years
3	58	M	8 months	L4-S1	37x40x62	No	STR	-	Yes	4 years
4	51	M	3 months	S4-S5	35x25x15	No	TR	-	No	-
5	69	M	15 days	S1	25x20x17	No	Biopsy	-	No	-
6	37	F	24 months	S2	32x17x22	No	Biopsy	-	No	-
7	18	M	4 months	S1-S3	32x23x36	No	GTR	-	No	-
8	49	M	4 months	S1-S2	24x23x17	No	GTR	Incontinence	Yes	3 years
9	64	F	12 months	S3	70x51x65	Yes	GTR	-	Yes	1.5 years

*Time between symptom onset and surgery
STR: Subtotal resection, TR: Total resection, GTR: Gross total resection, TV: Transverse, CC: Craniocaudal, AP: Anteroposterior, RT: Radiotherapy

Table 2. Symptom characteristics

Symptoms	Number of patients
Back pain	8
Leg pain	5
Urinary/bowel incontinence	2
Rectal fullness sense	1
Incidental diagnosis	1

done for a coccyx fracture and the patient was asymptomatic before the fracture. Two of the patients (22.2%) died during the follow-up. Surgical treatment delayed an average of 10.3 months (15 days-3 years) after symptom onset. One patient's cause of death was acute stroke and the other is unknown. Rectum perforation, urinary and fecal incontinence were the postoperative complications seen in three (33.3%) patients. None of the patients received chemotherapy after or before surgical treatment. 2 patients underwent radiotherapy. No metastases were seen in all of the patients. Five patients (55.5%) had local recurrence and 2 patients had recurrence despite patients getting radiotherapy after surgery.

Discussion

Our study's patients showed known factors to prevent recurrence may not be sufficient. Although current evidence shows radiotherapy prevents recurrence, 2 patients received radiotherapy and showed recurrence despite radiotherapy (14,16,17).

Surgical treatment of SC has mainly two aspects which are surgery with wide resection margins and radiotherapy. According to literature, a wide resection margin lowers the recurrence rate (9). However, resection surgery can cause harm to important anatomic structures and long term disabilities may occur but in our study group none of the patients developed disabilities.

Another treatment for SC is particle therapy and is more effective than conventional radiotherapy (13). However, in literature particle therapy was studied on a small sample size and in our study none of the patients administered particle therapy for adjuvant treatment. We believe there is a need for more evidence on this subject.

Study Limitations

We have a small sample size in our study and SC are uncommon condition in daily neurosurgery practice, these are limitations of our study. We believe there is a need for more study on this rare condition.

Conclusion

There is a need for an up-to-date systematic review and meta-analysis of current evidence on SC's surgical treatment. Current evidence on SC treatment shows us the importance of wide resection margins and radiotherapy. We hope our study will contribute to future systematic reviews.

Ethics

Ethics Committee Approval: Ethical approval was obtained from Ankara University School of Medicine Human Research Ethics Committee (approval no: 2022000106-1, date: 09.03.2022).

Informed Consent: Retrospective study.

Peer-review: Externally peer-reviewed.

Authorship Contributions

Concept: M.Z., E.S.A., Ö.O., B.C.A., S.H., E.B.M., Ö.K., Ü.E., Y.Ş.Ç., Design: M.Z., E.S.A., Ö.O., B.C.A., S.H., E.B.M., Ö.K., Ü.E., Y.Ş.Ç., Data Collection and Processing: M.Z., E.S.A., Ö.O., B.C.A., S.H., E.B.M., Ö.K., Ü.E., Y.Ş.Ç., Analysis or Interpretation: M.Z., E.S.A., Ö.O., B.C.A., S.H., E.B.M., Ö.K., Ü.E., Y.Ş.Ç., Literature Search: M.Z., E.S.A., Ö.O., B.C.A., S.H., E.B.M., Ö.K., Ü.E., Y.Ş.Ç., Writing: M.Z., E.S.A., Ö.O., B.C.A., S.H., E.B.M., Ö.K., Ü.E., Y.Ş.Ç.

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