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Abstract

Background: Long term side effects of cancer treatments can have a significant effect on functional mobility. Radiation therapy can have significant effects on all body tissue, including muscle and connective tissue. Surgical resection of malignant tumors also carries post-operative side effects that can influence function. The purpose of this case study is to highlight the successful treatment of neck pain despite limited treatment options from a complex past medical history. **Case Description:** This case study details the treatment of a 56-year-old female with a history of a clivus chordoma. Adverse effects following surgical resection, cervical fusion, and radiation therapy compelled this patient to present to an outpatient orthopedic clinic with complaints of neck pain and the presence of multiple functional deficits. **Intervention:** The intervention focused on manual therapy to address soft tissue adhesions in the upper quarter, therapeutic exercise to increase thoracic mobility, neuromuscular re-education of the transverse abdominus and diaphragm, and education for the successful integration of therapy principles to activities of daily living and exercise. **Outcome Measures:** The outcome measures utilized with this patient were limited to the Focus On Therapeutic Outcomes survey. **Discussion:** Several publications on the topic of clivus chordoma treatment, radiation therapy, and the overall neuromuscular complications experienced by cancer survivors acknowledge the role that physical therapy must play in the successful rehabilitation of these patients. Review of the related literature provides support for the interventions utilized in this case study including soft tissue mobilization, joint mobilization, therapeutic exercise, and neuromuscular re-education.

Keywords: physical therapy; radiation fibrosis; chordoma; cervical fusion; neck pain

Background

In the United States, predictions estimate that the number of cancer survivors will exceed 18 million by the year 2020.[1] Many of these patients present to outpatient physical therapy clinics with direct complications from the cancer or as the result of treatment such as surgery, chemotherapy, or radiation therapy. Of importance to this case are the sequelae of adverse effects suffered by a survivor of a clivus chordoma. Chordomas are a form of aggressive malignant neoplasm derived from notochord remnants. Representing up to 4% of tumors originating in bone, chordomas are rare and typically slow growing but have a high likelihood of local recurrence and the potential to metastasize and infiltrate local structures.[2-5] It is estimated that between 25 and 39% of chordomas develop at the clivus and involve the occipital-cervical (OC) joint[3,4]. Because chordomas are locally aggressive, likely to recur, and resistant to chemotherapy, the primary treatment approach is surgical resection of the tumor followed by radiation therapy.[4]

Surgical resection of this type of tumor is complicated by the tendency of chordomas to invade important structures including the brainstem and spinal cord.[5] The proximity of the clivus to critical vascular and neurological structures including the brainstem, optic nerve, cochlea, and pituitary gland can make achieving a full surgical resection of the tumor extremely difficult, if not impossible in some cases.[4,6] In addition to the challenges presented by the location of the tumor, the aggressive nature of the tumor compromises the stability of the OC joint as the tumor causes destruction of bone and ligaments in the region.[3,6] For this reason, fusion of the cervical spine is often performed in conjunction with or prior to resection of clival chordomas.[3] Complications of OC joint fusion include vertebral artery injury, dura tears, cerebrospinal fluid leakage, nerve or spinal cord injury, screw failure, bone fusion failure, and wound infection.[7] Surgical damage to nervous structures can occur from direct transection or from ischemia, traction, or compression due to positioning during surgery.[8]

Radiation therapy is considered a best practice treatment of head and neck cancers due to the difficulty the location presents in achieving a complete surgical resection.[5,9] It is well recognized that radiation therapy improves outcomes in chordomas, yet exact treatment parameters are not well established[5]. The goal of radiation therapy is to kill fast dividing tumor cells, but despite efforts to limit exposure to critical tissues it can produce a variety of unwanted side effects.[9] Any structures in the radiation field can be damaged by the treatment, including the spinal cord, nerve roots, peripheral nerves, muscles, ligaments, skin, and fascia.[9-11] In many cancers treated with radiation therapy, such as breast or prostate cancer, there are typically no neuromuscular effects. However, these adverse effects are expected following the treatment of head and neck cancers due to the delivery of high doses of radiation in the vicinity of critical structures.[9] Radiation fibrosis, sometimes referred to as Radiation Fibrosis Syndrome (RFS), defines the pathological fibrotic processes and consequent clinical presentation of affected tissues following exposure to radiation therapy.[12] Complications of RFS include the progressive thickening of skin, subcutaneous tissue, muscle fibers, ligaments, tendons, nerves, and bone. Adhesions between adjacent tissues are also common. These changes reduce overall compliance of involved tissues and lead to functional and cosmetic changes that can impact quality of life.[11] The exact pathophysiology of these adverse effects are not entirely understood, but it is hypothesized that the fibrosis of tissue causes local nerve compression, demyelination, and microvascular ischemia. The progression of RFS is gradual and can persist indefinitely.[9,10] Currently, there are no treatments to prevent or delay this fibrotic process.[10]

The neuromuscular and musculoskeletal effects of radiation therapy are of interest to physical therapists because of the functional implications of RFS. The effect of radiation on the musculoskeletal system is the progressive fibrosis of the tissues leading to a loss of elasticity, shortening of tissue fibers, and contracture causing a loss of range of motion and decreased function.[11-13] As previously mentioned, peripheral nerves may be damaged by compressive fibrosis or ischemia of surrounding tissue leading to pain, sensory loss, and weakness. The radiation field used in the treatment of head and neck cancer typically exposes multiple nerve roots to direct radiation injury. Because of this direct injury to upper cervical nerve roots, it is common for patients with head and neck cancer to present with greater weakness of the biceps, deltoids, and rotator cuff muscles compared to other upper extremity

musculature. Abnormal activity of the upper cervical plexus and spinal accessory nerve are other common findings in these patients.[12] The suprascapular nerve may develop a neuropathy as a result of radiation therapy that leads to low back pain and shoulder dysfunction.[11] Damage to the dorsal scapular nerve supplying the rhomboids also contributes to shoulder dysfunction.[13] This demonstrates the principle that muscles outside the radiation field can be affected if their innervating nerves pass through the radiation field.[9] This type of radiation injury may present clinically as visible atrophy of the trapezius and sternocleidomastoid (SCM), or protraction of the shoulder with weakness in the rhomboids and rotator cuff muscles depending on the nerve damaged.[10] Head and neck cancer patients also experience painful spasms, particularly of the trapezius, SCM, and scalenes. Clinically, the patient may describe the spasm as a feeling of stiffness or tightness in the muscle.[12] The continuous muscle contractions during spasm can also cause ischemia from compression of blood vessels in the area, which in turn sets off the inflammatory response and ultimately sensitizes nociceptive nerve fibers and contributes to the experience of muscle pain in these patients.[13] In severe cases, head and neck cancer patients develop a cervical dystonia, contracture, or torticollis along with wasting of the SCM and scalenes that can cause head drop from the muscle weakness. [11-13]

This case study details a clivus chordoma survivor who provided unique challenges to the treating physical therapists from the adverse effects of her cancer treatments detailed above. Therefore, the purpose of this case study is to highlight the successful treatment of neck pain despite limited treatment options from a complex past medical history.

Case Description

A 56-year-old female presented to an outpatient orthopedic physical therapy clinic with neck pain. At the time of initial evaluation, the patient was 15 months post-surgery for the removal of a clivus chordoma and concurrent cervical fusion of the occiput through the second cervical vertebrae. The recurrence of the cancer nine months following surgery resulted in radiation treatments to the cervical spine that concluded approximately three months prior to physical therapy treatment. This patient presented with chief complaints of bilateral neck pain and stiffness that developed over the course of one year following surgery and radiation treatments.

In addition to the presence of a cervical fusion, this patient was further limited by damage to cranial nerve 11 and the long thoracic nerve that occurred during surgery, leaving her with a thoracic nerve palsy and resultant deficits in strength and range of motion of the left upper extremity. Following surgery, she also suffered weakness of the right lower extremity and numbness in the left lower extremity that remained intermittent. The patient reported she had utilized a transcutaneous electrical nerve stimulation (TENS) unit previously with successful pain control for her neck and low back. Other personal factors of importance include the patient working full time as a registered nurse at an oncology clinic; a history of high activity levels including hobbies of running, weight lifting, yoga, and kayaking; her commitment to maintaining her level of physical activity; and the support of her husband and daughters. Patient goals for physical therapy included returning to prior recreational activities with less than 3/10 pain and becoming independent in symptom and pain management.

Given these factors and the complexity of this patient's medical history, it was necessary to modify the course of the physical therapy intervention in order to accommodate deficits and avoid exacerbation of pre-existing problems.

Clinical Impression #1

Based on prior medical history and subjective interviewing, this patient appeared to be a good candidate for soft tissue mobilization (STM) to structures of the cervical spine and shoulder, thoracic spine mobilization, therapeutic exercise for increased thoracic mobility and strengthening of postural stabilizers, TENS, and education on posture during activities of daily living (ADL).

Examination

Pain rating

At initial examination the patient rated her pain a 5/10, where at best it was a 2/10 and at worst a 7/10.

Functional limitations

At the time of initial evaluation, the patient reported moderate difficulty turning over in bed and severe difficulty with turning her head, looking up, using an elliptical, using a stair master, weight training, and running.

Range of motion testing

Range of motion (ROM) was assessed utilizing estimated percentages of expected full function. For example, the patient was observed to achieve only 25% of the full expected range of active right and left side bending of the cervical spine. See table 1.

Table 1. Active and Passive ROM

	Active	**Passive**
Cervical Spine	Flexion: 50% Extension: 25% Right Side bending: 25%* Left Side bending: 25%* Right Rotation: 25%* Left Rotation: 25%*	Flexion: 50% Extension: Not tested Right Side bending: 50% Left Side bending: 50% Right Rotation: 25% Left Rotation: 25%
Thoracic Spine - Lumbar Locked	<30˚ rotation bilaterally	

*****Indicates painful pulling sensation on opposite side during motion testing

Joint mobility testing

Joint mobility of the cervical spine was assessed in supine, while joint mobility of the thoracic spine was assessed in prone. Assessment techniques are listed in table 2.

Table 2.

	Force Direction	**End Feel**
Cervical Spine	Upper: Not tested due to fusion Lower: Posterior-Anterior Central	 Firm
Thoracic Spine	Posterior-Anterior Central	Firm

Neurological testing

Myotomes and dermatomes were assessed utilizing manual muscle testing and light touch sensation screening, respectively. See table 3.

Table 3.

	Level	**Grade**	
		Left	**Right**
Myotomes	C5- Deltoid C6- Bicep, wrist extensors C7-Tricep	4+/5 4/5 5/5	5/5 5/5 5/5
Dermatomes – light touch	Upper Extremity (all)	Intact	Intact

Palpation

Muscle guarding was noted in bilateral upper trapezius, scalenes, and cervical paraspinals.

Assessment

The assessment following initial examination concluded that this patient demonstrated a decrease in functional status and subjective and objective deficits that could be addressed by physical therapy intervention. At initial evaluation, the patient presented with primary deficits in cervical and thoracic mobility. It was the opinion of the treating physical therapist that these deficits could be successfully overcome by increasing thoracic range of motion and strength of upper extremities.

Intervention

This patient was treated for 13 visits over 8 weeks including the initial evaluation. Interventions are outlined chronologically below.

Initial evaluation-Week 1

Treatment provided during the initial evaluation began with education on postural awareness and the importance of improving thoracic mobility when active and passive cervical ROM is decreased. Education was also provided on the patient's current weight lifting routine with emphasis on avoiding upper trapezius substitution and improving core activation during stability exercises to limit the potential for developing cervical pain.

The patient was instructed in foam rolling for a home exercise program (HEP). This consisted of longitudinal foam rolling (roll along spine) with arms placed at shoulder abduction of 45˚ and 90˚, then shoulder abduction of 90˚ plus 90˚ of elbow flexion. The patient was guided through deep breathing with this activity to increase flexibility of anterior musculature.

The second exercise added to the home program at the initial visit was "book openings" or "open books". This is an exercise aimed at improving thoracic rotation. The patient is positioned in side lying with knee and hip flexion to lock out the lumbar spine. The bottom shoulder is protracted out from under the body with the arm outstretched, then the patient is asked to rotate through the thoracic spine by reaching back with the top shoulder and arm while opening the chest toward the ceiling as much as possible. The patient was guided through three repetitions of three deep breaths per side for this exercise.

Treatment Session-Week 1

The patient education and HEP provided in the initial evaluation was reviewed at this session. The patient was then instructed in diaphragmatic breathing with concurrent pelvic floor and transverse abdominus activation and educated on the importance of utilizing this technique during all core exercises completed independently. This sequence was added to the HEP. The session concluded with STM to the bilateral upper trapezius and pectorals.

Treatment Sessions-Week 2

The patient returned for two visits during the second week of therapy. The HEP was reviewed with the addition of overpressure by the physical therapist during book openings to avoid substitution through the low back and neck. It was noted that attention would be required to prevent excessive rotation during book openings as the patient was observed to utilize lumbar or cervical substitution during the exercise. At the end of the week, the patient reported the presence of low back pain that had not been present previously. The patient felt that this was exacerbated by and potentially caused by the book opening exercise from the HEP. At this time, the book opening exercise was temporarily discontinued from the plan of care. Transverse foam rolling of the thoracic spine was performed and added to the HEP. Sessions during week two were concluded with STM and stretching, to either bilateral upper trapezius or the right side lumbar paraspinals.

Treatment Session-Week 3

The patient attended one session during week three at which she reported an improvement in low back pain. The HEP was reviewed, including book openings which were completed pain free when the

patient was positioned in adequate hip flexion to lock out the lumbar spine. The patient was then instructed in proper technique for pelvic tilts and progressed to a pelvic tilt plus glute bridge. The session was concluded with STM to the bilateral upper trapezius.

Treatment Sessions-Week 4

At the beginning of week 4, the patient reported an exacerbation of low back pain after performing several hours of yard work over the weekend. See tables 5 and 6 below for results of a bilateral hip and low back screen performed in response. Over two sessions during the week, the HEP was reviewed, including book openings. Pain was present during this exercise if adequate stabilization was not provided close enough to the thoracolumbar junction. Due to the continued difficulties encountered during book openings, the patient was instructed in the thread the needle stretch as an alternative. This stretch is performed by placing the patient in quadruped position, then instructing the patient to reach one arm through the space between the opposite arm and leg. During the thread the needle stretch with this patient, the therapist provided stabilization for keeping hips level. When twisting to the left side, the patient was manually assisted with positioning of the left scapula due to the thoracic nerve palsy on this side. It was noted that this position elicited the tingling into the left leg that had been intermittent since the patient's surgery for chordoma resection.

The patient was also instructed in seated thoracic trunk rotation with a dowel held behind the back at waist level. The patient required moderate cueing to avoid substitution via movement through the hips and the lumbar spine. It was also noted that the patient had greater ROM when rotating to the left compared to the right side. Diagonal reaching patterns were also performed in sitting with repeated verbal cues for avoiding substitution patterns.

Week 4 sessions were concluded with STM to the bilateral upper trapezius and scalenes, and right side lumbar paraspinals. Grade IV thoracic central posterior to anterior vertebral joint mobilizations with breathing were intermixed during STM.

At the end of week 4, the patient reported minimal pain, but continued to experience tightness her lumbar paraspinals, upper trapezius, and scalenes that was worse on the right side than the left.

Bilateral hips and low back screen:

Range of motion testing: ROM of the lumbar spine was assessed in standing and ROM of the hips was assessed in supine. See table 5 for results.

Table 5. Active and Passive ROM

	Active	**Passive**
Lumbar Spine	Flexion: 100%, deviation to right side Extension: Not assessed Right Side bending: 75% Left Side bending: 75% Right Rotation: 25% Left Rotation: 25%	Not tested
Hips	All AROM found to be WNL	Flexion: 100% Extension: Not assessed Abduction/Adduction: WNL Internal Rotation: Right>Left

Within normal limits (WNL)

Palpation: Palpation of the innominates determined that the posterior superior iliac spines were level, but the left side was more prominent. Lumbar paraspinals were tender to palpation near the L4 and L5 vertebrae. At L5, soreness was noted to be more prominent on the left transverse process than right.

Special Testing: Special testing for hip pathology was completed in supine. See table 6 for results.

Table 6. Special Tests

	Left	**Right**
Straight Leg Raise	Negative	Negative
FABER	Positive	Positive > Left

Assessment: After completing the screen of the lumbar spine and hip, the physical therapist determined that low back pain was likely persisting due to a lack of integration of newly gained thoracic mobility into ADL and the patient's personal exercise regimen.

Treatment Sessions-Week 5

During the two treatment sessions of week 5, the HEP was reviewed in addition to the patient's typical exercise routine. Components of her routine that were potentially exacerbating low back pain were identified and addressed. The patient reported persistent low back tightness and sacroiliac joint irritation following episodes of increased pain over the previous weekend. This prompted the therapist to remove all rotational movements from the plan of care. Thus, book openings and the thread the needle stretch were replaced with standing stretches using a stretching cage to prevent irritation of the low back. Treatment sessions were concluded with STM to the lumbar paraspinals and piriformis.

Treatment Sessions-Week 6

There were no new treatments added during the two sessions of week 6. Previous exercises were reviewed and education on incorporating appropriate posture and avoidance of excessive lumbar lordosis during ADL was reinforced.

Treatment Session-Week 7

The patient reported no pain since her last therapy session and had not been taking any pain medication. At this time, she elected to decrease frequency of physical therapy sessions to once per week. Treatments from the previous week were repeated.

Treatment Session-Week 8

The patient reported maintenance of decreased pain levels and decreased areas of tightness. Soreness remained present at times, primarily in the right shoulder after prolonged activity. The patient reported she had not had any episodes of low back pain and felt that if she performed her HEP upon getting out of bed each day, she functioned without pain. This session focused on review of foam rolling from HEP and instruction in standing transverse rotation with yellow TheraBand for multifidus activation. STM to the bilateral upper trapezius, cervical paraspinals, and scalenes was performed as in previous sessions. At this time the patient felt she was managing symptoms effectively with her HEP and elected to keep her case open while she determined if more sessions were needed.

Outcomes

At the end of treatment, the patient subjectively reported full resolution of neck and low back pain. Tightness in these areas persisted at times, but the patient was independent in self-management strategies. The patient continued to perform exercise, work tasks, and ADL without pain and did not return to the clinic for another visit. The patient was discharged after approximately 4 weeks pain free without a therapy visit.

Upon initial evaluation, this patient scored a 47/100 on the Focus On Therapeutic Outcomes (FOTO) survey. The predicted FOTO score was a 62/100 to be achieved in 12 visits. After 9 visits, the patient completed a follow up FOTO survey and scored a 55/100. No additional FOTO surveys were collected from this patient.

Additional outcomes measures could have been collected for this patient in order to better quantify progress made in physical therapy. For a patient presenting with neck pain, the Neck Disability Index (NDI) is one such outcome measure that could be considered. The NDI has excellent test-retest reliability of 0.90-0.93 in patients who underwent cervical fusion for a degenerative disorder and test retest reliability of 0.86 in patients with chronic, nonspecific neck pain.[14,15]

Discussion

The purpose of this case study was to highlight the successful treatment of neck pain despite limited treatment options from a complex past medical history. This case study outlined the physical therapy episode of care for a patient with a history of clivus chordoma and subsequent surgical excision of the tumor, fusion of the first two cervical vertebrae, and a history of radiation therapy. This report details the intervention selections made for this patient and demonstrates the practice of altering plan of care based on patient presentation.

This patient initially presented with bilateral neck pain that had been developing over the course of one year during her treatment for a clivus chordoma. The patient was limited in treatment options due to the cervical fusion and neurological damage incurred during surgery and/or radiation therapy. Over the course of treatment, the patient presented with additional complaints of low back pain and shoulder pain in addition to the chief complaint of bilateral neck pain. The variability in presentation was used to guide treatment options and develop a self-management program for this patient. Because the damage to neuromuscular and musculoskeletal structures caused by radiation therapy is considered to be permanent and indefinitely progressive[8,9], the treating physical therapist had to consider the long term benefits of interventions such as manual therapy. Limited mobility of the cervical spine and left shoulder dictated careful consideration during intervention selection to avoid increasing pain through unwanted compensation. At the same time, education on appropriate modifications to movement patterns and beneficial substitutions was an integral part of the success of this course of intervention.

Several publications on the topic of clivus chordoma treatment, radiation therapy, and the overall neuromuscular complications experienced by cancer survivors acknowledge the role that physical therapy must play in the successful rehabilitation of cancer survivors[10,12,13,16]. Multiple reviews conclude that significant improvements in quality of life can be achieved with an exercise program for patients who experience adverse effects of cancer or radiation therapy.[9,10,16] Neuromuscular re-education is useful for improving kinesthetic sense and overall posture. An emphasis on home programs for self-management is considered best practice to maintain improvements in quality of life. Specific recommendations have been made for exercise to improve movement efficiency, endurance, muscle strength, and joint range of motion.[10,11,13]

After radiation therapy, muscles are fibrotic and firm to palpation and may have associated weakness.[16] The patient will often describe feelings of tightness, pulling, or cramping of the muscles in the affected area.[13] Despite these abnormalities, there is evidence to suggest that myofascial restrictions can respond favorably to myofascial release, joint manipulations, and other manual therapy techniques.[10] These intervention techniques aim to improve flexibility of the skin, connective tissue, and deep fascia to restore mobility between soft tissue layers. Furthermore, this type of intervention has been found to reduce pain and improve circulation in areas with RFS.[11,13] According to one 2012 publication, there are no data available on the treatment of shoulder dysfunction in cancer patients who received radiation therapy. However, the author does support the use of physical therapy to address core strength and posture, neck extensor weakness, pectoral girdle tightness, and rotator cuff weakness in attempt to restore appropriate alignment of the glenohumeral joint.[12]

Review of the related literature supports the interventions chosen for this patient. Manual techniques likely had a positive influence on tissue extensibility following radiation therapy.[10] Encouraging the patient to maintain her independent exercise routine and educating on the integration of appropriate movement modifications during activities may have positively affected this patient's symptoms.[9,10,16] Performance of thoracic vertebral joint mobilizations and establishing a HEP with goals to improve thoracic mobility is also supported by the limited research available for treatment of clivus

chordoma survivors.[10] Although the recurrence rate for clivus chordomas has been reported to be as high as 68%, five-year survival rates for the cancer are reported to be between 51% and 82%.[4] These statistics and the successful outcome of this case report provide an argument for continued study of therapy interventions for survivors of chordomas and other head and neck cancers.

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