Extensive lumbar chordoma and unique reconstructive approach

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Abstract Chordomas are rare, malignant, low grade, primary bone tumours arising from notochord remnants. They are rarely located in the lumbar spine and consequently, the literature is lacking about the condition. There are very few case reports on the technical aspects of resection of extensive lesions such as our case. Treatment can be potentially curative only if aggressive surgical resection is utilised. We describe a case of an extensive lumbar chordoma, and demonstrate the possibility of performing a staged anterior and posterior total tumour resection with planned transgression in conjunction with an expandable cage, vascularised rib, fibula and latissimus dorsi grafts for the treatment and reconstruction of extensive spinal disease with this condition.

Keywords Chordoma · En-bloc vertebrectomy · Free vascularised fibula graft · Free vascularised rib graft · Vascularised latissimus dorsi graft

Introduction

Chordomas are rare, malignant, low grade, primary bone tumours arising from notochord remnants. They are most commonly found in middle to older-aged adults with an incidence of <1/1,000,000 per year, accounting for <5% of primary spinal bone tumours [1, 2]. Although chordomas may appear anywhere along the spinal axis, most arise at the sacrum (50–60%) or skull-base (25–35%), consistent with the theory that they form from the notochord. Consequently, it is uncommon to find chordomas arising from the cervical (10%) or thoracolumbar spine (5%), making our case of an extensive chordoma of the lumbar spine, a rarity [1, 3, 4]. Our paper focuses on the technical aspects of this unique reconstructive surgery.

Case report

A 47-year old male smoker without relevant past medical history, presented with a 3-year history of progressive lower back pain that had rapidly deteriorated over 6 months. This was associated with a 3-month history of neurogenic claudication. Work injury exists from 5 years ago, however, radiological investigations at the time were reported as normal.

Examination findings include: straight leg raise 60° bilateral, absent ankle reflexes, reduced distal sensation and lumbar spine percussion tenderness. Abdominal examination revealed a tender palpable mass in the lower quadrants.

Standing X-ray illustrated a sclerotic L4 and destructive L5 vertebral bodies, with a grade 1 spondylolisthesis of L5/S1 (Fig. 1a, b). Magnetic resonance imaging (MRI) revealed an extensive heterogenous bone-based malignancy

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Fig. 1 47-year old male with chordoma. **a** AP radiograph of the lumbar spine illustrating sclerosis of L4 and a destructive lesion of L5. **b** Lateral radiograph of lumbar spine illustrating sclerotic L4 vertebral body and grade I spondylolisthesis of L5/S1. **c** T2-weighted sagittal MRI of lumbar spine showing bone-based malignancy from L3 to L5 causing severe canal stenosis. **d** T2-weighted axial MRI of lumbar spine highlighting borders of tumour (red arrows) infiltrating posteriorly into the paraspinal musculature. **e** T2-weighted coronal MRI of abdomen shows malignancy expanding into the abdomen and psoas muscles bilaterally

extending from L3 to L5 causing severe canal stenosis, with expansion of the posterior elements causing erosion into the paraspinal musculature posteriorly and expansion into the psoas and abdomen anteriorly (Fig. 1c, d, e).

Pathological diagnosis

Histopathological inspection of the resected mass revealed multiple varying sized nodules of large, pale, vacuolated cells showing mild nuclear pleomorphism (Fig. 2a, b), with some areas of metaplastic cartilage formation within the tumour. Immunohistochemistry was positive for keratin 19 (Fig. 2c), s100 (Fig. 2d), HBME1 and EMA staining (E-Cadherin negative). Histological features in conjunction with immunohistochemical profile were consistent with the diagnosis of chordoma.

Operative treatment

Due to the extensive size of the chordoma, a two-staged posterior and anterior total tumour resection with planned transgression was performed utilising, pedicle screw fixation, anterior and posterior vascularised bone grafting and an anterior expandable cage support. Due to the predicted massive loss of tissue, psoas muscle and paraspinal muscle, bilateral vascularised latissimus dorsi flaps were planned.

Part 1

Posterior approach involved dissection to reveal T10-sacrum, resection of the posterior elements with muscle margins of the tumour from L3 to L5 to the level of the pedicle/vertebral body junction, along with tumour dissection around the lateral elements into the paraspinal muscles and anteriorly onto the transverse processes of L2-Sacral alar (Fig. 3a). Pedicle screws (K2M, Inc. Leesburg, Virginia, USA) were inserted from T12 to L2 along with S1 and pelvic (iliac) screws (Fig. 3b). Bilateral vascularised 10th rib grafts raised on single superior pedicles were osteotomised segmentally to increase the reach of the graft (Fig. 4a, b). Bilateral latissimus dorsi turn-over flaps were utilised to fill the dead space formed from extensive paraspinal muscle resection. This procedure lasted 18 h, with 2.5 L of blood loss, with provision of 3 L of crystalloid, 6 units of packed red blood cells (PRBC) and 4 units of fresh frozen plasma. The patient mobilised on day 2 and was discharged on day 6.

Part 2

Anterior approach was performed 6 weeks following the posterior approach due to concerns with graft viability of the rib and latissimus dorsi grafts. Retroperitoneal exposure

Fig. 2 Histological and immunohistochemical examination of lumbar biopsy. **a** Chordoma tumour nodules and adjacent skeletal muscle (H&E, scanning magnification). **b** Characteristic chordoma histomorphology of large, pale, vacuolated physaliphorous tumour cells in amongst myxoid stroma (H&E, high magnification). **c** Pan-cytokeratin immunohistochemistry highlighting tumour cells (high magnification). **d** S100 immunohistochemistry highlighting tumour cells (high magnification)

Fig. 3 Posterior resection of chordoma. **a** Extensive resection of paraspinal muscle and posterior elements from L3 to L5 resulting in a very large dead-space. **b** Proximal pedicle screw fixation from T12 to L2 and distal fixation of S1 and pelvis

via a flank excision across the left costal margin in the left lateral position from L1-pelvis was performed. The peritoneal cavity was deflected anteriorly and the diaphragm was not taken down (Fig. 5a). Residual pedicles from posterior resection were removed. Slow tumour dissection and removal, from L2 end plate to S1 superior endplate occurred due to close proximity of major vessels. En-bloc resection was not technically possible from the anterior

approach, so an extensive intra-lesional approach was performed. Reconstruction between L2 and sacrum utilised maximum height OBLISK (Ulrich Medical, Ulm, Germany) expandable cage (Fig. 5b) and vascularised left free fibula flap raised in standard fusion connected end to end to small spinal branches of aorta and IVC (Fig. 6a). The procedure lasted 20 h, with 4 L of blood loss and provision of 8 units of PRBC. Neither procedure utilised

Fig. 4 CT reconstruction of lumbar spine with bilateral vascularised 10th rib grafts (white arrows) which have been osteotomised segmentally. **a** Posterior view, **b** lateral view

Fig. 5 Anterior resection of chordoma. **a** Anterolateral retroperitoneal exposure of tumour with peritoneal cavity and aorta deflected anteriorly and psoas muscle retracted posteriorly. **b** Orthotec OBLISK expandable cage inserted between the L2 end-plate and sacrum

manoeuvres to reduce blood loss, as cell-saver is contraindicated in cancer surgery. The patient mobilised on day 2 and was discharged on day 8.

Follow-up

Both operations were complication free, much to the surprise of the multiple surgical teams involved. The patient

appeared well 3-months post-operatively and only complained of residual pain from the excision made along the left costal margin. Upon 9-month follow-up, the patient was independent, mobile and his pain had improved significantly. The computer-tomography (CT) scan at this stage demonstrates early fusion across L3–S1 into the vertebrectomy cage (Fig. 7).

Adjuvant radiotherapy has been delayed to enable solid fusion. If any recurrent disease is noticed upon his 12-month review, radiotherapy will be necessary.

Discussion

Chordomas are primary bone tumours arising from the notochord remnant characterised by slow growth over years, often metastasising late in 40–60% to the lung, bone, soft tissue, liver, skin or lymph nodes [1, 2]. The 5-year survival rate (5YSR) is 50% while the 10-year survival rate (10YSR) is 28% [5]. The site and size of a chordoma impacts greatly on the outcome, with those arising from the vertebrae being more likely than other chordomas to be locally aggressive, metastasize (80 vs. 43%) and have low 5YSR. Lumbar chordomas like our patient have higher metastatic rates than in any other location [2–5], however, local aggressiveness rather than metastases is more likely to cause death and disability [5].

Signs and symptoms often appear to be a result of dysogenic or non-specific pathology, resulting in delayed diagnosis [3]. Symptoms most commonly arise from compression of the anterior column and roots. The most frequent complaint in those with lumbar chordomas is pain and paraesthesia. Motor changes can occur, however, paraplegia is rare [2]. Our patient complained of 3 years of back pain and 3 months of neurogenic claudication.

Pre-operative diagnosis of lumbar chordomas is also difficult as they are often confused for more common lumbar spine tumours, including a giant cell tumour, myeloma, metastasis, haemangioma and aneurismal bone cyst [2]. Currently, MRI is the preferred imaging modality for pre-operative diagnosis and assessment. MRIs can detect small chordomas before major symptoms develop, improve patient prognosis and increases the chance of the tumour being resectable [6]. Chordomas appear hypo- or iso-intense on T1-weighted images, whereas on T2-weighted images they appear hyperintense [2]. Radiographically, vertebrae appear irregular as chordomas are slow growing, allowing for bone remodelling. Chordomas are lobulated and can be soft or firm with focal areas of necrosis, cyst, ossification, haemorrhage and calcification. Characteristically, their histological appearance includes vacuolated cells with mucous producing physaliphorous [2, 5].

Fig. 6 Post-operative CT reconstruction. **a** AP view showing both vascularised fibula graft and expandable cage. **b** Lateral aspect demonstrating expandable cage. **c** Illustration of original extensive chordoma extending from L3 to L5

Fig. 7 Post-operative mid-sagittal CT at 9 months with bone graft extending into vertebrectomy cage (arrows)

Treatment

Surgical resection has become more aggressive over time, progressing from lesion debulking to total resection and aims for complete tumour removal with wide margins, decompression, reconstruction and stabilisation [2]. Aggressive resection is the only curative treatment because:

1. Clinical symptoms are characteristically late presenting meaning chordomas are often large upon diagnosis thus other oncological treatments are unsuitable [2, 5, 6]
2. Inadequate surgical margins are associated with poor survival and high relapse rates
3. Local tumour progression is a strong prognostic factor for poor survival rates [3, 4]
4. Chordomas are resistant against conventional radiotherapy (curative only at toxic doses >60 Gy) and chemotherapy [1].

In the majority of scenarios, however, margin-free excision is difficult due to surrounding neurovascular structures and extensive tumour size [3, 4].

Studies take divided stances on surgery's effectiveness in treating chordomas. While surgery allows for a 10YSR of 30–65%, some studies demonstrate high local recurrence rates of 66–85% and functional impairment [1]. Jawad and Scully's (2009) review of 962 patients with chordoma shows that surgery, in comparison to patients who do not

undergo surgical resection, significantly increases 5YSR (69 vs. 47%, respectively) and mean survival (96 vs. 54 months, respectively). The study also concluded that those aged <59 years with primary tumour size <8 cm have better survival rates and prognosis [1]. However, Ferraresi et al. (2010) study concluded that despite surgical advancements, long-term prognosis and control is still poor in these patients. As the disease is rare, most literature on surgery's effectiveness is restricted to case reports, single-centred case series and small population-based surveys with no definitive conclusions [1].

Radiotherapy still plays a role in preventing recurrence in patients with inadequate surgical resection and intralesional/close margins and as definitive therapy in inoperable patients. This role may increase with the emergence of stereotactic radiotherapy [3, 7]. Chordomas are generally chemo-resistant, however imatinib mesylate is showing promise in clinical trials [1, 7] with a prospective multi-centred phase II study of 55 patients revealing a 73% clinical benefit rate after 1 year [3]. While recent developments in radiotherapy and chemotherapy show promise, there is a lack of controlled, long-term follow-up studies to guide future practice [6].

Unfortunately, performing an en-bloc resection was not possible due to technical constraints. The extensive size of our patient's chordoma, resulted in an aggressive, staged, total surgical resection of the tumour with planned transgression, utilising an expandable cage and vascular rib, latissimus dorsi and fibula grafts for reconstruction.

En bloc resection

Studies show that en bloc resections can improve survival rate and decrease rates of local recurrence and thus are often utilised in chordoma treatment [6]. En bloc resection refers to complete tumour removal in one piece. Theoretically, total en bloc resection ensures there is no residual tumour, thus preventing tumour cells from contaminating adjacent tissue which increases disease-free survival [7].

Boriani et al's (2006) review of prospective studies clearly illustrates that margin-free en bloc resection is the only treatment which achieves a disease-free state. En bloc spondylectomies can increase the 5YSR without disease from 50 to 70% to up to 100% by reducing the risk of recurrence [7], however, slow-growing local relapse is inevitable following radiotherapy, intralesional surgery with/without radiotherapy or en bloc resection with an intralesional margin. Recurrence appears an average of 26 months post-treatment. Despite some cases of long survival (up to 10 years), the pain/disability caused by tumour regrowth causes poor quality of life [6].

In our case, Ferraresi et al. (2010) study and Jung and Shin's (2009) case, the patients underwent staged

combined anterior and posterior vertebrectomy. No studies compare the effectiveness between a posterior approach and a combined anterior and posterior approach. Theoretically, the posterior approach alone has poor direct visualisation of ventral structures increasing chances of major vessel injury. The combined approach can either be staged or simultaneous with the disadvantage of long operating times. One study comparing staged and simultaneous approaches in 117 patients revealed both methods had similar outcomes and complications with the simultaneous group having shorter operation times by ≈ 100 min and greater blood loss of ≈ 800 mL [4, 7]. Due to the extensive nature of our patient's chordoma, a staged approach was most appropriate.

The overall complication rate following total en bloc resection in most studies is 36.3%. Complications include major neurovascular injury, excessive bleeding, tumour cell contamination, spinal instability or abdominal herniation (following anterior approach) [6, 7].

Reconstruction

Spinal tumour resection results in a substantial loss of posterior elements and biomechanical instability making bone reconstruction and arthrodesis essential [4, 8]. In cases like ours where en bloc resection involves multiple levels, reconstruction is complex [8].

While there are many well-studied options for anterior reconstruction, options for successful long-term posterior reconstruction are in their early stages. Instrumented posterior reconstruction is often unsuccessful in the long term as biomechanical forces cause fatigue. Biological reconstruction with posterior arthrodesis and anterior support avoids this problem, but problems arise with adjuvant chemotherapy/radiotherapy as they interfere with healing, increasing the chance of arthrodesis failure [8]. For this reason, adjuvant radiotherapy has yet to be utilised in our case. Bone grafts depend upon the defect's size, location and instability. Whilst non-vascularised (rib/fibular/iliac crest) grafts can be used they are not ideal if soft tissue has been compromised by previous surgery, persistent infection, wide resections or radiotherapy [9, 10]. In cases involving multiple level resection like ours, vascularised bone grafts prove ideal [8, 10]. Theoretically, vascularised grafts should not be utilised with tumours as graft blood flow into the tumour bed could facilitate tumour cell dissemination and recurrent tumour growth, however, the risk of this occurring is low following en bloc resections [8, 11]. Both vascularised fibular and rib grafts have been popularised for spine reconstruction due to their length. Vascularised rib grafts, however, are limited to the thoracolumbar region as intercostal vessels limit their positioning. Free vascularised fibular grafts, however, can be

used anteriorly or posteriorly throughout the thoracolumbar region [9].

Over the last few decades, vascularised rib grafts were mainly utilised for treating infections and kyphosis. Since the 1980s, they have been used for a growing number of spinal pathologies with placement of vascularised rib grafts anteriorly due to their curvature, being the most promoted method described in the literature [11]. There is however, a lack of literature addressing vascularised rib graft use in the posterior spine [8, 11], however, Wilden et al. (2006) and Eastlack et al. (2007) both support their use. Indications for posterior use of vascularised pedicle rib grafts include:

1. >5 cm gap in the posterior spine
2. Anterior and posterior insufficiency
3. Compromised adjacent soft tissue or bone due to perioperative adjuvant treatment [8, 9].

Wilden et al.'s [11] study on 18 patients showed that pedicled vascularised rib grafts up to 30 cm in length could be used anteriorly or posteriorly to span up to ten levels and achieve union in an average of 6.8 months.

Conventional grafts are inconsistent in effectively achieving bony union in hostile environments caused by radiotherapy, oncologic resections and persistent infection [9]. However, recent studies have shown that patients with lumbosacral defects following spinal tumour resection and radiation, where traditional bone graft use is not possible, can have free vascularised fibular grafts to achieve successful bony union in an average of 4.5 months [9, 10]. Disadvantages of free vascularised fibular grafts include increased operating times, increased blood loss and technical difficulties achieving microvascular anastomoses. Whilst operation times are significantly longer, the amount of blood loss in fibular harvest generally does not require transfusion [9, 11]. Complications caused by fibular harvesting include transient peroneal nerve palsy, non-union, flap failure, infection and hematoma [10], however, these are uncommon [9].

Conclusion

Chordomas are rarely located in the lumbar spine and consequently, the literature is lacking about the condition.

Treatment can be potentially curative only if aggressive surgical resection is utilised. We have described a case of an extensive lumbar chordoma, and have demonstrated the possibility of utilising staged posterior and anterior total tumour resection with planned transgression, in conjunction with an expandable cage, vascularised rib, fibula and latissimus dorsi grafts for the treatment and reconstruction of a lumbar spine with this condition.

Conflict of interest None of the authors has any potential conflict of interest.

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