

Primary bone tumours of the spine: Presentation, surgical treatment and outcome

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Abstract

Background: Primary bone tumours of the spine are a group of rare tumours that include both benign and malignant lesions. Resection is associated with a high morbidity rate.

Methods: We retrospectively reviewed all the primary bone tumours of the spine that were surgically treated at our unit between 2005 and 2012 (haematological malignancies were excluded).

Results: Fifteen cases were included that presented at a median age of 36 years (range 8 to 65 years). Pain was the most common presenting symptom. Three patients had significant neurological deficits at time of presentation and in two cases there was an improvement after surgery. The median delay in diagnosis was 7 months (range 1 to 36 months). A variety of surgical strategies was employed with the use of adjuvant radio- or chemotherapy in six cases. Twelve benign and three malignant tumours were resected. Complications (sepsis, failure of fixation and recurrence) were encountered in five cases (33%).

Conclusions: Primary bone tumours of the spine are often associated with a significant delay in diagnosis. Surgical strategy should be individualised for each case. Acceptable results can be achieved with this approach.

Key words: primary bone tumour, bone tumour, primary spine tumour, spine tumour, resection

Introduction

Primary bone tumours of the spine are a heterogeneous group of conditions that includes both benign and malignant tumours, with only 6% of these tumours considered malignant.¹ The spine is a frequent site of metastasis, but primary bone tumours of the spine are rare and make up only 4.2% of spinal tumours.² Eleven per cent (11%) of primary musculoskeletal tumours occur in the spine² with certain tumours having an affinity for the spine. Although rare, these tumours offer a considerable diagnostic dilemma and therapeutic challenge. Timely diagnosis is crucial, as immediate attention could affect overall prognosis.³

Primary bone tumours are more common in the thoracic and lumbosacral regions than in the cervical spine.⁴ These tumours occur according to a typical anatomical distribution within the vertebra. In general, malignant tumours occur more frequently in the anterior elements and benign tumours in the posterior elements.³⁻⁵

Primary bone tumours of the spine are rare and offer a considerable diagnostic dilemma and therapeutic challenge

The aims of treatment of these axial neoplasms include complete resection if feasible, preservation of neurological function and stabilisation of the spine.⁴ In the paediatric age group the growing spine is an added consideration. The quality of evidence that guides treatment is low due to the rarity of these conditions, mostly limited to case series.⁶⁻⁸ Resection of tumours in the spine often leaves large bony defects and is associated with a high morbidity, and complications in up to 35% of patients have been described.⁹ Both sepsis and mechanical failure are prevalent. Previous papers report a deep sepsis rate of greater than 5%, mechanical failure in 7% and mortality related to the surgery in 2% of cases of en bloc resections in the spine.¹⁰ Combined anterior and posterior approaches and number of levels resected are predictors of increased complications.¹⁰

The aim of our study was to review the primary bone tumours of the spine treated at our unit regarding the presentation, the surgical strategy employed and the outcome.

Materials and methods

Following Institutional Ethics approval (reference HREC 029/2013) a retrospective review was performed of all the primary bone tumours of the spine that received surgical treatment at our unit from 2005 to 2012. We excluded all haematological malignancies. We treated 15 cases during this period that were included according to these criteria.

A case note and imaging review was done where patient demographics, details of presentation, clinical information and the management strategy were analysed. All complications and mortalities were recorded.

Results

The median age at presentation was 36 years (range 8 to 65 years) with eight male and seven female patients affected. The median follow-up was 18 months (range 3 to 60 months).

Table I: Histological diagnosis

Benign tumours		
Active	Haemangioma	3
	Osteoid osteoma	1
	Eosinophilic granuloma	1
Aggressive	Osteoblastoma	1
	Giant cell tumour	2
	Aneurysmal bone cyst	4
Malignant tumours		
Stage IIb	Osteosarcoma	2
	Leiomyosarcoma of bone	1
TOTAL		15

The median delay in diagnosis was 7 months (range 1 to 36 months) from the time a medical opinion was first sought to a diagnosis being made. Pain was the most common presenting symptom (15 from 15 cases) and nine cases presented with pain alone. Pain was mostly axial pain in the affected region (11 of 15 cases), although three patients presented with chest pain and one with flank pain. Other presenting symptoms included neurological symptoms and odynophagia. Two patients presented with a recurrence of a benign aggressive tumour following incomplete resection at another unit.

Ten patients had no neurological deficit at time of presentation. Two patients presented with radicular symptoms and another three had significant neurological deficits at time of presentation. Of these three patients, one was graded as a Frankel A, one a Frankel C and one a Frankel D. All three cases with significant neurological deficit were in patients with benign aggressive lesions. Two of these cases had significant improvement after surgery. We had no cases of neurological deterioration following surgery.

We treated 12 benign and three malignant tumours. The histological diagnoses are summarised in *Table I*.

In all cases the surgery was performed by the senior author. A variety of surgical strategies was utilised. An intralesional resection or debulking of the tumour was performed in six cases and a marginal resection was performed in nine cases (see *Table II*) to correlate with diagnosis. This was achieved through a posterior only approach in six cases and a combined approach in nine cases. The combined approaches were performed at a single sitting in five and staged in four cases. In 14 patients a fusion was performed. A posterior only fusion was performed in four cases (one uninstrumented and three instrumented) and a combined posterior and anterior fusion was performed in ten cases. In ten cases there were significant bone defects after resecting the tumours. These were addressed with cages with bone graft in five cases and strut allografts in five cases.

Adjuvant therapy was used where indicated. Adjuvant radiotherapy alone was used in four cases, adjuvant chemotherapy in one case and a combination in another one case (*Table II*).

There were complications in five cases (33%). This included one case of deep sepsis with failure of fixation in the same patient. This required staged revision of instrumentation with the patient symptom free at time of last follow-up. Three cases had spread or recurrence of the tumour. Two of the recurrent cases were in malignant tumours and one in benign aggressive tumour. In the one recurrent malignant tumour a repeat attempted marginal resection was performed. The other two recurrent cases were not re-operated as resection was not possible with acceptable morbidity. We had three deaths in the series.

A variety of surgical strategies was utilised

Table II: Summary of patients

	Diagnosis	Age (yr) & sex	Neurological status	Resection	Surgical approach	Adjuvant therapy	Complications
1	T9 osteoid osteoma	38, male	No deficit	Intralesional	Posterior	None	None
2	T3 eosinophilic granuloma	36, male	Frankel C	Intralesional	Staged posterior & anterior	None	None
3	T12 osteosarcoma	62, female	No deficit	Marginal	Staged posterior & anterior	Chemotherapy	Recurrence, died
4	C3 epithelioid haemangio-endothelioma	44, male	No deficit	Intralesional	Anterior & posterior	Radiotherapy	None
5	T4 aneurysmal bone cyst	18, male	Frankel A	Marginal	Posterior	None	None
6	L5 giant cell tumour	10, male	No deficit	Marginal	Staged posterior & anterior	Radiotherapy	None
7	T1 osteoblastoma	12, male	Frankel D	Marginal	Posterior & anterior	Radiotherapy	None
8	T11 leiomyosarcoma of bone	65, male	No deficit	Marginal	Posterior & anterior	Radiotherapy	Recurrence, died
9	T9 osteosarcoma	45, female	No deficit	Marginal	Posterior	Chemotherapy & radiotherapy	Died
10	C2 aneurysmal bone cyst	8, female	No deficit	Marginal	Posterior	None	None
11	L5 haemangioma	51, female	L4 radiculopathy right	Intralesional	Posterior & anterior	Embolisation	Incisional hernia
12	T8 giant cell tumour	31, female	No deficit	Intralesional	Staged anterior & posterior	None	Recurrence
13	T7 haemangioma	39, female	No deficit	Intralesional	Posterior	None	None
14	T7 aneurysmal bone cyst	26, male	No deficit	Marginal	Posterior & anterior	None	Sepsis, failure of fixation
15	L2 & 3 aneurysmal bone cyst	13, female	L2 radiculopathy bilateral	Marginal	Posterior	None	None

All three patients with malignant tumours died within the follow-up period at a median of 18 months after surgery (range 3 to 59 months). Death was due to recurrence or spread of the tumours in two cases and unrelated in one case.

The patients' treatment and complications are summarised in Table II. Figures 1 to 3 show examples of cases from the series.

The aims of treatment of these axial neoplasms include complete resection if feasible, preservation of neurological function and stabilisation of the spine

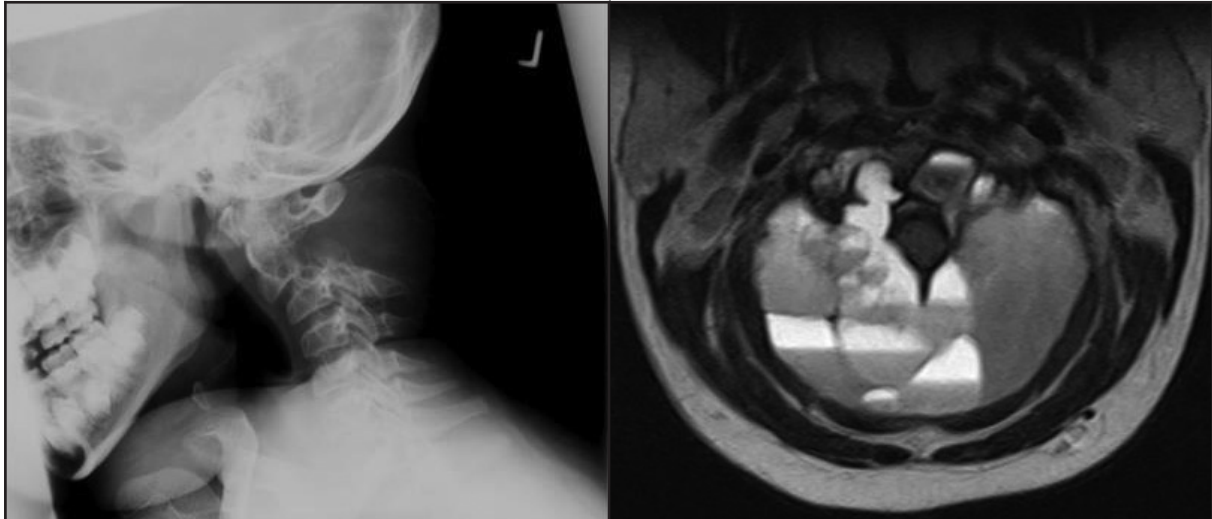


Figure 1A

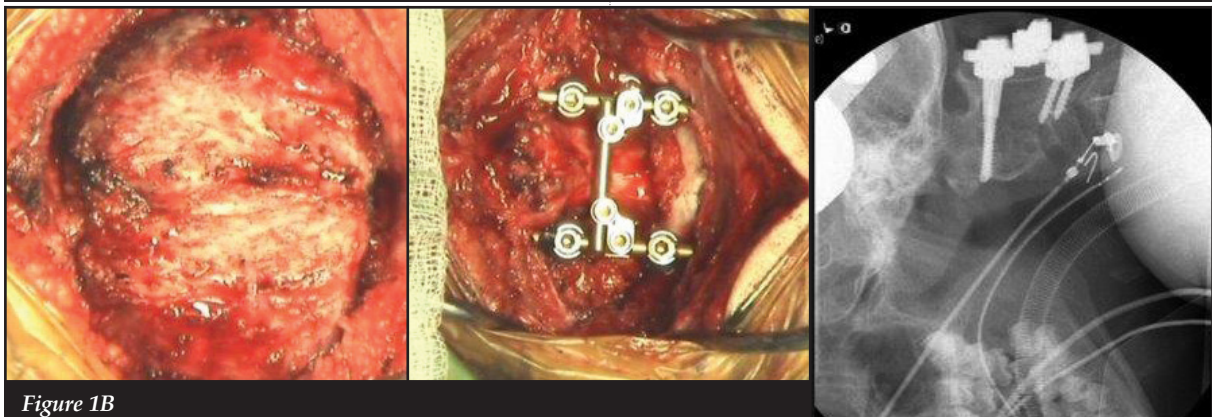


Figure 1B

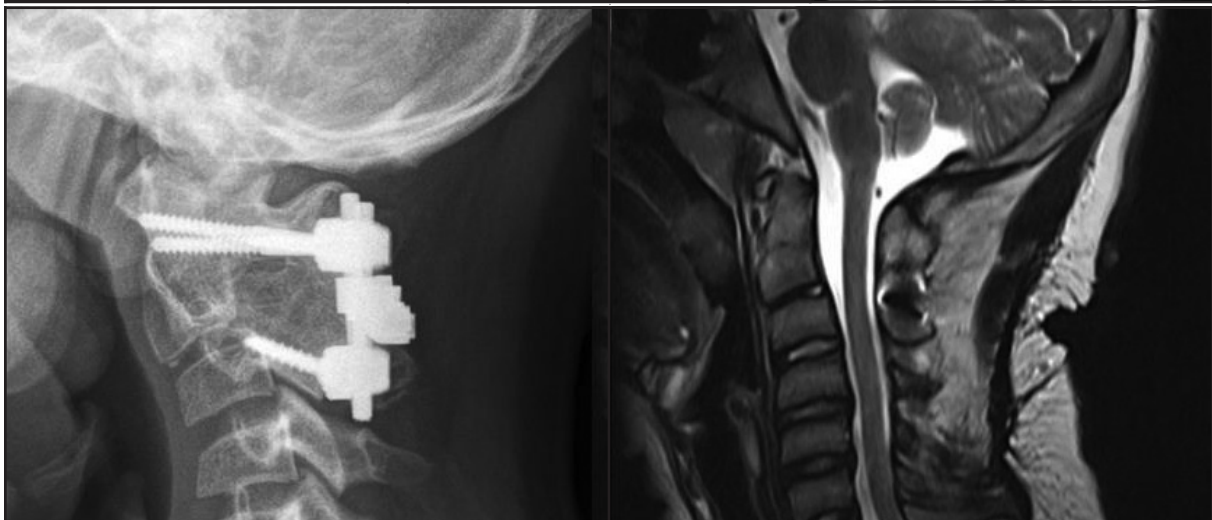


Figure 1C

Figure 1. An 8-year-old female patient presented with neck pain and odynophagia. A- Plain lateral radiograph of the cervical spine revealed an expansile lesion of the posterior elements of C2, and an axial T1-weighted MRI sequence showed typical features of an aneurysmal bone cyst. B- Posterior resection with instrumented fusion was performed in a single sitting. C- Follow-up radiograph and a sagittal T1-weighted MRI sequence showed successful fusion and no signs of recurrence at two years' follow-up.



Figure 2A

Figure 2B



Figure 2D



Figure 2E

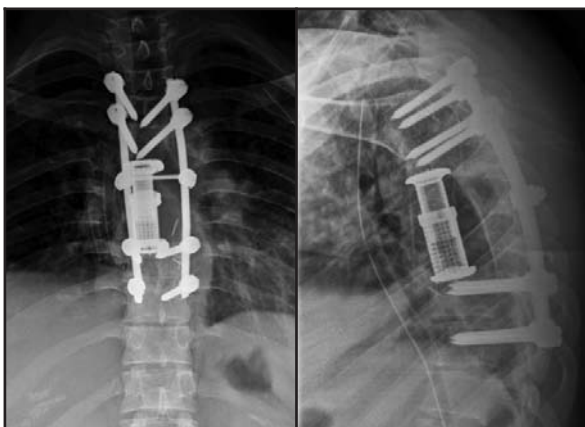


Figure 2C

Figure 2. A 26-year-old male patient presented with persistent pain two years after having surgery elsewhere for an aneurysmal bone cyst in T7. A- Plain radiographs of the index procedure and B- sagittal T2-weighted MRI sequence showed residual or recurrent tumour. A revision marginal resection with posterior instrumented fusion and anterior corpectomy and cage was performed. C- Post-operative radiographs showed revision metalware in situ. D- One year post-revision surgery patient presented with failure of fixation and sepsis. Radiograph shows pull-out of the posterior instrumentation. The posterior instrumented fusion was revised in a staged manner to a longer construct. Fourteen months post-revision surgery the patient was symptom-free. E- Follow-up radiographs at that stage revealed no signs of loosening of metalware or recurrence of tumour.

Discussion

The most common presenting complaint of patients with primary tumours is pain, with roughly 60% complaining of axial pain and 25% of radicular symptoms.³ This is confirmed in our series. The presence of a neurological deficit carries a worse prognosis.¹¹ Pain at rest or night pain is red flag symptoms that warrant further investigation. Spinal deformity is obvious when present, but occurs in less than 10% of patients. The most common cause of a painful scoliosis in adolescents is an osteoid osteoma.⁵

A delay in diagnosis is common and our finding is confirmed in other series.⁴ This is due to the presenting symptoms often being vague, pain often being the only symptom, and the initial radiographs often only showing subtle or no abnormalities leading to the symptoms not being properly investigated. Most patients report a slow, gradual onset of pain in the involved area. In benign tumours of the spine an average of 26 weeks of symptoms before presentation are reported.⁴ This was confirmed in our series where the average delay in presentation was 7 months.

A variety of classifications systems is used for primary bone tumours of the spine. The Enneking staging system¹² was initially described for primary bone tumours of the appendicular skeleton, but is applied to tumours of the spine as well. It prognosticates as well as guides surgical resection margins and has been validated in the literature.² The Weinstein-Boriani-Biagini surgical staging system¹³ is an anatomical classification based on the Enneking system that describes the number of vertebra as well as the number of sectors within a vertebra involved. Recommendations are then made for the most appropriate approach to resection of the lesion. We used the Enneking staging system. According to this system we treated 12 benign and three malignant tumours. Of the benign group five could be classified as active and seven as aggressive.

Appropriate treatment may be observational (Enneking grade 0 latent lesions) or surgical (most other lesions), depending on the level of pain, instability, neurologic compromise and the natural history of the lesion.⁵ The aims of surgery are complete resection of the lesion where feasible with preservation of neurological function.

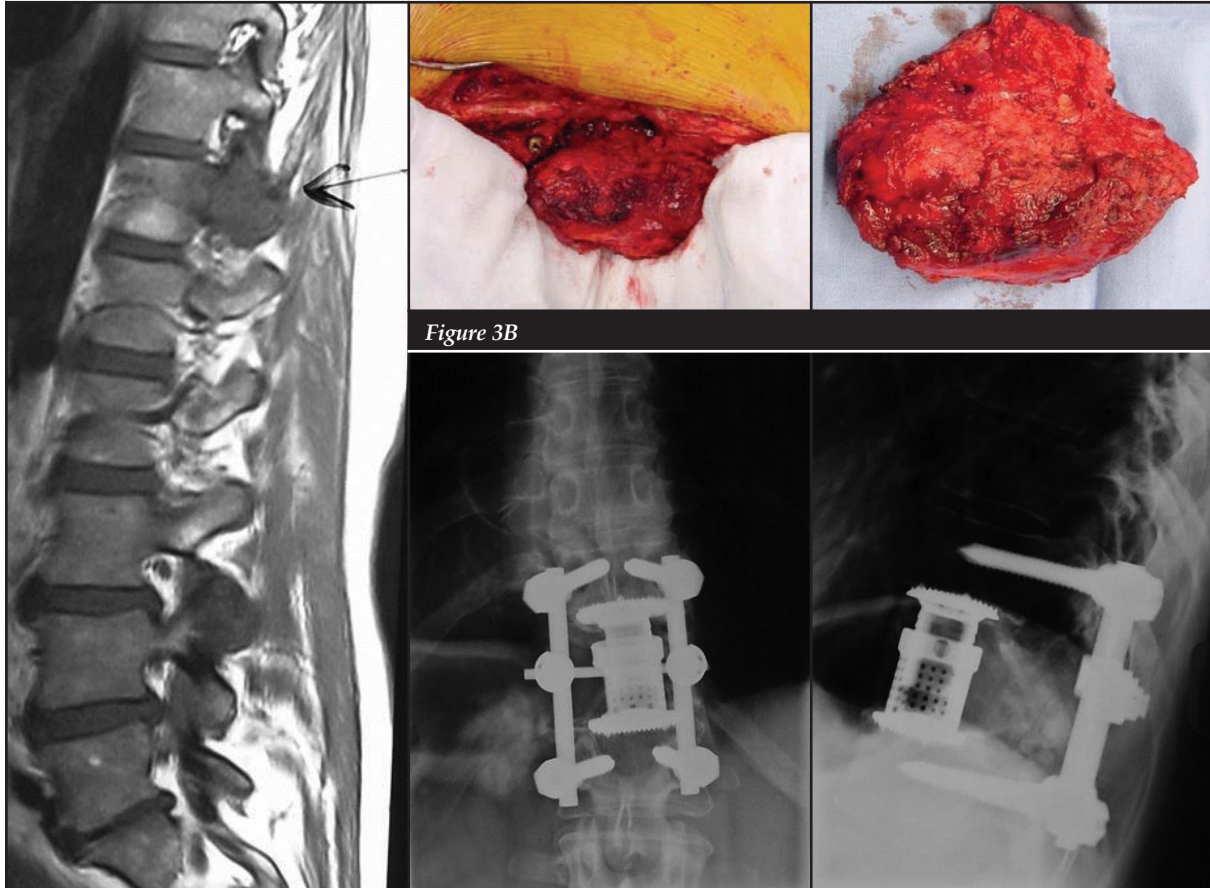


Figure 3A

Figure 3C

Figure 3. A 62-year-old female presented with flank pain and loss of weight. A- T2-weighted MRI sequence showed a mass in T12. Biopsy confirmed an osteosarcoma. B and C- Following neo-adjuvant chemotherapy a staged posterior resection with posterior instrumented fusion was performed followed by an anterior corpectomy and cage with bone graft. Unfortunately the patient had a recurrence of the tumour at four years and despite repeat surgery died within the follow-up period.

In paediatric cases the growing spine is an added consideration.⁴ Patients should have individualised approaches and treatment plans because of the many variations of natural history, extent and level affected, local and distant spread, neurological deficit, instability, bone defect after resection and whether resection is feasible with acceptable disability.³ We used a variety of surgical strategies customised to the patient and the tumour. The specific surgical strategies are summarised in *Table II*.

In peripheral malignant sarcomas the most important factor affecting survival is complete resection of the tumour with a wide margin.^{14,15} There are however no reports prescribing surgical margins in spinal tumours. Traditional intralesional resection in a piecemeal fashion is thought to increase the likelihood of local recurrence. True wide resection would cause unacceptable morbidity as it would involve resecting the segment of spinal cord.

Total en bloc spondylectomy in a single stage from an all posterior approach was described by Tomita.¹⁴ Here wide margins are achieved except at the pedicles and occasionally the spinal canal. This approach decreases local recurrence rates.¹⁶⁻¹⁸ In benign aggressive lesions surgical eradication also provides the best long-term cure.^{6,18} We had three cases of local recurrence in our series. Two were in malignant tumours where a marginal resection was performed. The other recurrent case was a thoracic giant cell tumour encapsulating the aorta and forcing the surgeon to perform an intralesional resection.

There are no reports prescribing surgical margins in spinal tumours. Traditional intralesional resection in a piecemeal fashion is thought to increase the likelihood of local recurrence

Recent advances in chemotherapy have led to improved survival in malignant primary bone tumours, including in the spine.¹⁹ Adjuvant radiotherapy is indicated in some malignant and benign aggressive lesions.²⁰ The use of local radiotherapy does increase the risk of major local complications, including sepsis.¹⁰ We used radio- and chemotherapy in a number of cases (see *Table II*) as guided by our local oncology unit.

The main limitations of this study are the retrospective character and the relatively small numbers.

We conclude that primary bone tumours of the spine are often associated with a significant delay in diagnosis. Surgical strategy and adjuvant therapy should be individualised for each case. Acceptable results with minimal complications can be achieved with this approach.

The content of the article is the sole work of the authors. No benefits of any form have been or are to be received from a commercial party related directly or indirectly to the subject of the article.

The research has been approved by the local departmental research council and ethics committee (reference HREC 029/2013).

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