Acta Neurochir (Wien) (1996) 138: 1261–1266

Acta Neurochirurgica

Printed in Austria

Primary Spinal Column Sarcomas

E. Tasdemiroglu[1], E. Bagatur[2], I. Ayan[3], E. Darendeliler[4], and R. A. Patchell[5]

[1] Incirli Hospital, Neurosurgery Service, Istanbul, Turkey, [2] Incirli Hospital, Orthopaedic Surgery Service, Istanbul, Turkey, Departments of [3] Paediatric Oncology, and [4] Radiation Oncology, University of Istanbul, Institute of Oncology, Istanbul, Turkey, and [5] University of Kentucky Medical Center, Division of Neurosurgery, Lexington, KY, U.S.A.

Summary

Five cases of primary spinal column sarcomas are presented. Sarcomas primarily originating from paravertebral soft tissues were excluded. Patients' age ranged from 1 to 14 years (mean 8.4 years). The male : female ratio was 2 : 3. Two patients had Ewing's sarcoma (ES) originating from L5–S1 and L4–5 pedicles, respectively; two patients had mesenchymal chondrosarcoma (MCS) originating from L1–2 pedicles and L5 body, respectively; and one patient had osteogenic sarcoma (OS) of C4 body. All patients clinically presented with pain and progressive weakness of the extremities. The time that elapsed between the onset of symptoms and diagnoses ranged from one to five months. All cases were treated with chemotherapy, radiotherapy and subtotal tumour resection with spinal canal decompression. Two cases received posterior spinal fusion operations. Three patients were alive 10 to 98 months following diagnosis. Only the case with ES of L5–S1 pedicles was in complete remission and off therapy at the 98th postoperative month. The two MCS cases were in partial remission, and were receiving chemoterapy at the time of analysis. These tumours caused similar clinical findings and prognoses, and required combined treatment, which consisted of surgery, radiotherapy and chemotherapy; histologically three different types of malignant tumours are presented in the same category. We preferred surgical decompression and stabilization procedures especially for neurologically symptomatic patients, even if they had extensive tumours with high grades. By spinal canal decompression and stabilisation, we did not intend to cure the disease; however, we intended to provide neurological improvement, spinal stabilisation, improved quality of life, early mobilisation of the patient, and cytoreduction by means of surgical tumour ablation, which could render the chemotherapy more effective.

Keywords: Children; Ewing's sarcoma; mesenchymal chondrosarcoma; osteosarcoma; chemotheapy; spinal fusion.

Introduction

Nearly 70% of primary spinal bone tumours seen in children are benign[18]. Osteoid osteomas, and osteoblastomas, osteochondromas, and aneurysmal bone cysts account for over 40% of all primary spinal lesions seen in paediatric patients. Metastasis to the spine is a very common preterminal event seen in patients with relapsing disease. However, primary involvement of the vertebral column by osteosarcomas or chondrosarcomas is quite rare. The purpose of this study is to describe the clinical features, treatment options and outcomes of five such patients.

Patients and Methods

Five cases of osteosarcoma (OS), mesenchymal chondrosarcoma (MCS), and Ewing's sarcoma (ES) primarily originated from the spinal column were included in this study. Between January 1982 and June 1994 24 cases of OS, ES and MCS were admitted and treated at the University of Kentucky Medical Center. Four of these cases primarily originating from spinal column (the body, pedicle and/or lamina of the vertebra). Two cases of OS and MCS primarily originating from paravertebral soft tissues were excluded. A single case of MCS of the L5 body, was operated by the senior author in Turkey, and was included in the study.

The ages of patients ranged from 1–14 years (mean 8.4 years). The male : female ratio was 2 : 3. Two patients (cases 2 and 3) had Ewing's sarcoma (ES) originating from L5–S1 laminae and L4–L5 pedicles, respectively. Two patients (cases 1 and 5) had mesenchymal chondrosarcoma (MCS), originating from L1–2 pedicles and L5 body, respectively; and one patient had osteosarcoma (OS) (case 4) of the C4 body. Detailed clinical analyses of 5 cases are summarised in Table 1.

All patients presented with pain and progressive neurological deficits of the extremities. The time which elapsed between the onset of symptoms and diagnoses ranged in 1 to 5 months. Tuberculous spondylitis was misdiagnosed in one of the MCS cases (case 5). Pre-operative radiological diagnoses were obtained by plain x-rays, CT scans and/or MR imaging of the spine. All cases were treated with chemotherapy, radiotherapy and subtotal tumour resection, with spinal canal decompression. Two cases (cases 4 and 5) received spinal fusion.

Table 1. *Detailed Clinical Analyses of the Five Primary Vertebral Column Sarcoma Cases*

Case no.	Age/ Sex	Location	Histopathology	Clinical findings	Treatment	Outcome
1	1/F	L1–2 laminae	Poorly differentiated mesenchymal chondrosarcoma	One month history of progressive bilateral lower extremity weakness; bowel and bladder incontinence, decreased rectal tone	XRT to L1–5 levels (750 cGy); L1–5 total laminectomy and subtotal tumour resection; CT (VP-16, Ifosfamide, Carboplatinum, Doxorubicin)	Alive POM #20 Still under CT; mild left leg weakness able to ambulate
2	7/F	L5–S1 pedicles	Ewing's sarcoma	Five months history of right leg weakness, right thigh pain, urinary retention	XRT to L5–S2 levels (5580 cGy); L1–S2 total laminectomy and subtotal tumour removal; CT (VCN, Adriamycin, Cyotoxan)	Alive in remission POM #98
3	14/M	L4–5 pedicles	Ewing's sarcoma	Three months history of bilateral lower extremity weakness	L4–5 total laminectomy and subtotal tumour removal; CT (VCN, Cytoxan, 5-FU)	Died POM #8 Metastases to C6 body and the lungs
4	9/M	C4 body	Osteosarcoma (first diagnosis four months ago was osteoblastoma)	Four months history of neck stiffness, shoulder pain, right upper extremity weakness	L4 corpectomy, C3–4, C4–5 discectomies and fusion; C3–5 hemilaminectomy, C5–6 foreminotomy and posterior fusion; XRT cervical spine (1690 cGy) CT (Methotrexate/Leucovorine, Allopurinol, Adriamycin, Cisplatinum, Ifosfamide)	Died POM #24 Lung metastases
5	12/F	L5 body	Mesenchymal chondrosarcoma	Four months history of back pain, bilateral progressive lower extremity weakness and decreased pinprick sensation bilaterally below L4 level	L4–L5 total laminectomy, gross tumour resection, bilateral S1 and L4–L5 nerve roots decompression; L4–S1 posterior fusion with CD rods and transpedicular screws; CT (Cisplatinum Epiribucin, Ifosfomide); 5000 R boost XRT	Alive POM #10 Total neurological improvement

XRT radiotherapy, *CT* chemotherapy, *POM* postoperative month.

Results

Two cases died due to progression of the primary disease, during the postoperative follow-up. Case 3 had an ES of L4–5 pedicles and died during the 8th postoperative month, and case 4 had an osteosarcoma of C4 body, died at the 24th postoperative month. Both of these cases had progressive neurological deterioration during the terminal stage of their disease.

Case 3 had ES, originating from the posterior elements of L4–5 vertebrae. L3–5 total laminectomy and decompression of the cauda equine was accomplished. At the 8th postoperative month he returned with neck pain associated with vertebral body metastases to C6, and he died with progression of his disease.

Case 4 first presented with neck stiffness and shoulder pain. Plain x-ray of the cervical spine showed C4 compression fracture. The result of needle biopsy was haemangioma. He underwent C4 corpectomy, decompression of the C4 and C5 nerve roots and C3–5 anterior fusion. Pathology report showed an osteoblastoma. Four months after the first operation, he returned with right upper extremity weakness. He underwent C3–5 right hemilaminectomy, decompression of the C5 and C6 nerve roots, resection of the osteoblastic lesion. The posterior fusion was accomplished by threaded pins placed through the spinous processes of the C2 and C5, and autologous bone

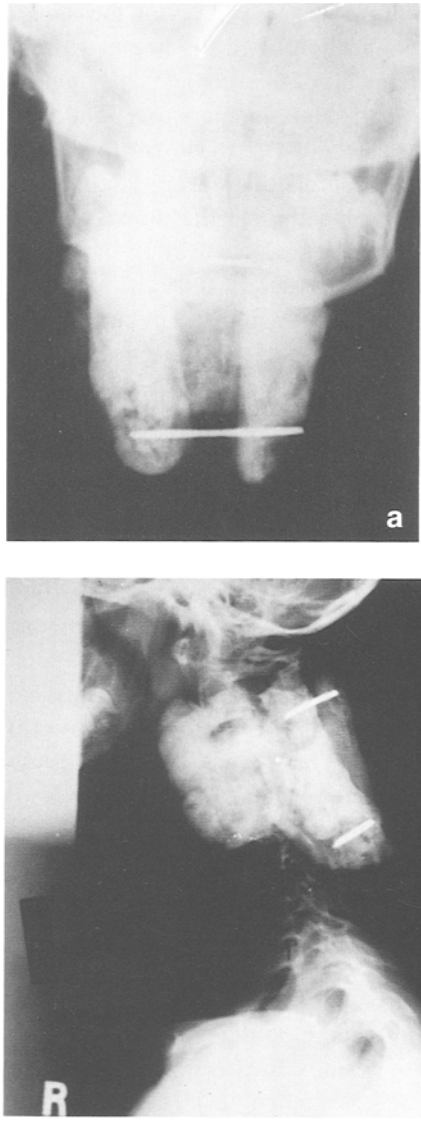

Fig. 1 a, b. The antero-posterior (a) and lateral (b) cervical spine x-rays showed an extensive paraspinal growth of a cervical osteoblastic lesion, and the threaded pins placed through the spinous processes of the C2 and C5 vertebrae, for posterior spinal fusion (case 4)

grafts were placed over the laminae between the C2 and C5 vertebrae bilaterally, and fixed to the threaded pins. However, the histopathological diagnosis of the osteoblastic lesion was osteosarcoma. During follow-up period plain cervical spinal x-rays showed an extensive paraspinal growth of the osteoblastic lesion (Fig. 1 a and b) and the MRI of the cervical spine showed severe spinal cord compression along with the tumour growth (Fig. 2). In spite of chemotherapy and radiotherapy, his neurological condition deteriorated and he died in 24th postoperative month.

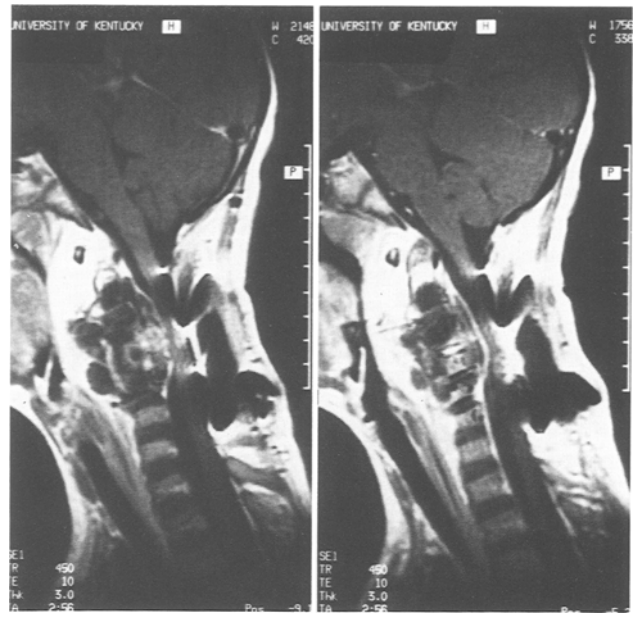

Fig. 2. The T1-weighted sagittal MR image of the cervical spine showed an enlarged mass of the cervical spine extending into the prevertebral tissue and spinal canal, and causing severe spinal cord compression (case 4)

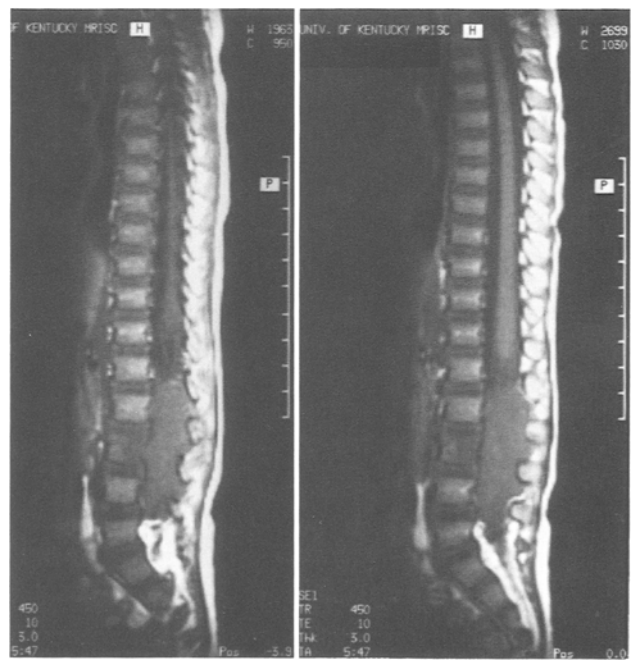

Fig. 3. The T1-weighted sagittal MR image of the spine showed a large extradural mass located below the conus, extending from L1 to L5, compressing the cauda equina and obliterating the sacrum (case 1)

Three cases were alive and neurologically improved 10 to 98 months after diagnosis. Cases 1 and 5 had mesenchymal chondrosarcoma, originating

from L1–2 pedicles (Fig. 3) and L5 body, respectively. Both cases received posterior spinal cord decompression with laminectomy and case 5 had L4–S1 posterior spinal fusion with transpedicular screws. Both cases improved neurologically. Case 2 had ES of the L5 pedicle. Following L5–S1 total laminectomy and subtotal tumour resection, she received radiotherapy and chemotherapy.

Only the case with ES of L5–S1 pedicles (case 2) was in complete remission, neurologically intact, and off therapy at the 98th postoperative month. The other two MCS cases (cases 1 and 5) were in partial remission and were receiving chemotherapy at the time of analysis.

Discussion

We present three different malignant tumours in a single category, because these tumours shared some similar properties in the paediatric age group. Although these tumours originated from bones or cartilages of the spine, the clinical findings, treatment modalities and prognoses resembled each other. Although ES, OS or MCS commonly metastasise to the spine; primary spinal origin of these tumours are quite rare.

Ewing's sarcoma is an uncommon primary malignancy of bone arising from primitive marrow elements and affecting children, adolescents and young adults [4, 13]. Primary spinal column ES rate with the majority originating in the sacrum is ranged from 3.2% to 4.9% [4, 12–14]. Pilepich *et al.* [12] reported 22 cases of primary vertebral ES cases and noted that 14 of these patients had neurological deficits at the time of diagnosis. In the series of Grubb *et al.* [5], they encountered 36 primary ES cases of the spine. Although they have not described the age groups in their study, more than half of their patients had neurological symptoms at the time of diagnosis. The five year survival rate was 33%. The mean survival time was 2.9 years. They could not find any significant correlation among the location of the tumour in the spine, the length of disease free survival, or incidence of the metastases. They stated that the patients enrolled in the Intergroup Ewing's Sarcoma Study had significantly better rates of disease free survival and overall survival. They suggested that surgical treatment consistent with gross resection, decompression and/or fusion was still controversial and patients undergoing laminectomy must be followed closely for the development of progressive kyphosis.

We present two primary spinal column ES cases. One of these cases is off therapy and symptom free for 98 months. This patient received subtotal tumour resection, chemotherapy and radiotherapy. Although the same treatment protocol (aggressive surgical tumour removal, chemotherapy and radiotherapy) was applied to the second case, he died with multiple metastases during the 8th postoperative month.

OS is the most common primary malignant bone tumour (excluding multiple myeloma). But primary spinal column OS is quite rare. Less than 2% of all primary OSs originate from the spine [4, 14, 18]. Because of its rapid dissemination with frequent bony metastases, multiple metastatic lesions to the vertebral column are more common than primary involvement. Treatment of lesions in the spinal column is usually difficult, and the outcome in these tumours has traditionally been very poor. In a review of 27 cases of OS of the spine median survival was 10

Table 2. *Summary of the Primary Paediatric Spinal MCS Cases in the Literature*

Author	Age/Sex	Vertebral level	Attachment	Treatment	Postop course	Condition Survival
Rubinstein, Schneider	15/F	T9–10	dura	R	–	alive 2 years
(5 cases)	18/F	T9	dura	GTR	recurrence 6 years	alive 9 years
	7/F	T10	dura	R	–	alive 3 years
	5/M	L2–4	dura	R	–	alive 2 years
	17/M	T6	dura	R	metastases 3 months	died 3 months
Chan HSL *et al.*	10.5/F	T1–4	dura	GTR	–	alive 1.5 years
Tasdemiroglu *et al.*	1/F	L1–2	dura	CT, XRT, R	–	alive 20 months
(2 cases)	12/F	L5 body	dura	CT, XRT, R	–	alive 10 months

CT chemotherapy, *XRT* radiotherapy, *R* resection, *GTR* gross total resection.

months from the time of diagnosis. Only 7 patients survived more than a year, and one patient survived over 5 years [15]. Vertebral OS arises from the anterior elements in more than 95% of cases. Malignant degeneration of osteochondroma or transformation of an osteoblastoma (OB) is also possible [4]. Although, subtotal excision of the benign OB is usually curative, recurrence of the tumour should inspire a careful review of the histology for the possibility of OS [8]. Primary OS of the cervical spine is quite rare. Only 11 cases have been reported in the English language literature since 1925 [1, 3, 7, 10, 11, 15, 17, 19]. In almost half of the these cases the preliminary diagnoses were benign OB; but the tumours recurred and during the second operation the histopathological diagnosis proved to be OS. OS cases of the spine could initially be misdiagnosed as osteoblastoma. The exact histopathological diagnosis of the lesion could be OS and the histopathological differentiation between OS and OB may be difficult, either due to absence of obvious anaplasia or the lack of the presence of certain atypical cells on microscopy [4, 10, 15]. By combined treatment (surgery, chemotherapy, radiotherapy), we obtained 24 months survival for our case.

MCSs are rare tumours of the bone and soft tissue, and were first described by Lichtenstein and Bernstein in 1959 [9]. Characteristic histopathological appearances differ in this tumour from classical chondrosarcoma. The CNS is the most common reported site of their extra-osseous origin [6]. Although their histological appearance is quite distinct, their rarity, frequent location in the meninges, and highly vascular nature may lead to the erroneous diagnosis of angioblastic meningioma or haemangiopericytoma [6]. A clinical course of frequent local recurrences at long intervals after the initial resection is also highly characteristic of these tumours. Fewer than 100 cases of MCS have been reported in the adult literature and the condition is even rarer in the paediatric age group.

Six cases of primary paediatric (<18 years of age) spinal column MCSs have been reported [2, 6] (Table 2). Although those cases were thought to primarily originate from the spinal cord dura mater, massive destruction of the vertebral elements was also seen. In our cases, we could not detect the primary origin whether from the dura mater or the vertebral elements of the spine, although both tumours were firmly attached to the dura mater. The treatment protocol for our MCS cases were aggressive surgical removal, chemotherapy and radiotherapy.

Before the CT era, since the early radiological diagnosis of primary spinal column sarcomas was difficult, treatment of these malignant tumours was mostly palliative, to obtain temporary relief of pain and neurological symptoms and signs. Besides, multi-drug chemotherapy regimens were not available, as the primary tool for cancer treatment. Traditional treatment consisted of limited tumour excision and radiotherapy. The outcomes associated with this approach were poor. Grubb *et al.* [5], did not recommend spinal canal decompression and/or tumour resection, if the tumour size was extensive and the grade was high. However, an aggressive surgical approach has been advocated for primary spinal column malignancies [16, 18]. Sundaresan [16] reported 7 cases undergoing wide resection or vertebral body resection combined with radiotherapy. Although 3 of those patients had died of their disease at the mean of 11 months, following their operation, four others were still alive, and three had no evidence of disease at mean survival of 52 months.

We preferred surgical decompression and fusion procedures especially for neurologically symptomatic patients, even when they had extensive tumours with high grades. By surgical decompression and stabilisation procedures we did not intend to cure disease; however, we intended to provide neurological improvement, spinal stabilisation, improvement of the quality of life, early mobilisation of the patient, and cytoreduction by surgical tumour ablation.

Conclusion

Combined treatment consisting of chemotherapy, radiotherapy and surgery should be the treatment of choice for primary spinal column sarcomas. The effect of chemotherapy is temporary and local recurrences along with distant metastases could not be prevented, at least, without cytoreductive surgery. Aggressive surgical intervention such as total or subtotal removal of the affected vertebra(e) with fusion should be the choice of surgical approach for this type of tumours. Pre-operative chemotherapy may render delayed surgery more successful for local recurrences, and finally postoperative adjuvant chemotherapy with or without radiotherapy should be the last step of the combined treatment for primary malignant vertebral column tumours [6, 7, 17]. More aggressive management of these tumours results in longer survival, but longer follow-up is needed before assumptions can be made about our ability to cure these tumours.

References

1. Ben Miled K, Siala M, Hamza KR, Rais H, Ben Rhouma T, Kammoun M, Hamza R (1988) Primary osteosarcoma of the cervical spine; one case. J Neuroradiol 15: 294–300
2. Chan HSL, Turner-Gomes SO, Chuang SH, Fitz CR, Daneman A, Martin OJ, Becker LE (1984) A rare cause of spinal cord compression in childhood from intraspinal mesenchymal chondrosarcoma. A report of two cases and review of the literature. Neuroradiology 26: 323–327
3. Fielding JW, Fietti VG Jr, Hughes JEO, Gabriellan J-CZ (1976) Primary osteogenic sarcoma of the cervical spine (a case report). J Bone Joint Surg 58A: 892–894
4. Freidlaender GE, Southwick WO (1982) Tumors of the spine. In: Rothman RH *et al* (eds) The spine, 2nd Ed, Vol 2. Saunders, Philadelphia, pp 1022–1040
5. Grubb MR, Currier BL, Pritchard DJ, Ebersold MJ (1994) Primary Ewing's sarcoma of the spine. Spine 19: 309–313
6. Harsh GR IV, Wilson GB (1984) CNS mesenchymal chondrosarcoma (case report). J Neurosurg 61: 375–381
7. Kebudi R, Ayan I, Darendeliler E, Agaoglu L, Bayindir C, Turantan I, Kinay M, Bilge N (1994) Primary osteosarcoma of the cervical spine. A pediatric case report and review of the literature. Med Ped Oncol 23: 162–165
8. Klein DM (1989) Extramedullary spinal tumors. In: Mc Laurin RL *et al* (eds) Pediatric neurosurgery, 2nd Ed. Saunders, Philadelphia, pp 443–452
9. Lichtenstein L, Bernstein D (1959) Unusual benign and malignant chondroid tumors of bone: a survey of some mesenchymal cartilage tumors and malignant chondroblastic tumors, including a few multicentric ones, as well as many atypical benign chondroblastomas and chondromyxoid fibromas. Cancer 12: 1142–1157
10. Marsh HO, Choi C-B (1970) Primary osteogenic sarcoma of the cervical spine originally mistaken for benign osteoblastoma. A case report. J Bone Joint Surg 52A: 1467–1471
11. Mnaymneh W, Brown M, Tejeda F, Morrison G (1979) Primary osteogenic sarcoma of the second vertebra. J Bone Joint Surg 61A: 460–462
12. Pilepich MV, Vietti TJ, Nesbit ME, Tefft M, Kissane J, Burgert O, Pritchard D, Gehan EA (1981) Ewing's sarcoma of the vertebral column. Int J Radiat Oncol Biol Phys 7: 27–31
13. Pizzo PA, Poplack DG, Horowitz ME, Hays DM, Kun LE (1993) Solid tumors of childhood. In: DeVita VT Jr *et al* (eds) Cancer principles and practice of oncology, 4th Ed, Vol 2. Lippincott, Philadelphia, pp 1738–1791
14. Savini R, Giunti A, Briani S (1985) Benign and malignant tumors. In: Bradford DS *et al* (eds) The pediatric spine. Thieme, Stuttgart, pp 131–154
15. Shives TC, Dahlin DC, Sim FH (1986) Osteosarcoma of the spine. J Bone Joint Surg 68A: 660–668
16. Sundaresan N, Rosen G, Huvos AG, Krol G (1988) Combined treatment of osteosarcoma of the spine. Neurosurgery 23: 714–719
17. Tigani D, Pignatti G, Picci P, Savini R, Campanacci M (1988) Vertebral osteosarcoma. Ital J Orthop Traumatol 14: 5–13
18. Weinstein JN (1991) Differential diagnosis and surgical treatment of primary benign and malignant neoplasms. In: Frymoyer JW *et al* (eds) The adult spine. Principles and practice, Vol 1. Raven, New York, pp 829–860
19. Yoshino MT (1991) Osteosarcoma of the cervical spine. Am J Roentgenol 157: 1357

Comment

The authors have identified five most unusual cases of sarcoma, three different types, and presented clearly their clinical material. Their review of the literature is pertinent, and the discussion reflects equally well a knowledge of paediatric bone tumours of the vertebral column and their palliative treatment. The tabular material is pertinent, and succinctly presented.

A. Raimondi

Correspondence: Erol Tasdemiroglu, M.D., Incirli Caddesi, Deniz Apt. 74/7, Bakirkoy, Istanbul, TR-34740, Turkey.