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PRIMARY EWING'S SARCOMA OF THE SPINE IN A TWO-YEAR-CHILD

Neurosurgery

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ABSTRACT

Ewing's sarcoma is a common malignancy of the bone and soft tissues in pediatric patients. It mostly affects the long bones and pelvis, and less commonly the flat bones and vertebrae. Primary Ewing's sarcoma affecting the spine is very rare(Yan et al., 2011). It accounts for only 3.5 to 14.9 percent of all primary bone sarcomas. The age of presentation ranges from 12 to 24 years (median 21 years) (Ferguson, 1999; Sharafuddin et al., 1992; Klimo Jr. et al., 2009). We report an unusual case of primary ES of the spine in a two-year-old boy, who presented to us with paraparesis and features of cauda equina syndrome.

KEYWORDS

1. Introduction

Ewing's Sarcoma (ES) is a highly malignant bone tumour. It involve any part of the skeleton but the most frequent parts are the ilium and diaphysis of femur and tibia [1, 2]. Primary ES of the spine is extremely rare [3]. It accounts for only 3.5 to 14.9 percent of all primary bone sarcomas. The age of presentation ranges from 12 to 24 years (median 21 years) [4–6].

2. Case History

A two-year-old male child was brought with a 2 month history of progressive weakness of both the lower extremities, difficulty in standing and walking, and progressive loss of bowel and bladder function. He had no history of trauma, back pain, and failure to thrive. He had no significant past, personal, or family history. All developmental milestones were achieved for his age.

On clinical examination there was paraspinal fullness and complete loss of power below the level of the knee joint in both lower extremities. (Hip flexion and knee extension was grade 5; ankle dorsiflexion, great toe extension, and ankle planter flexion were grade zero.) Bulk was normal and tone was reduced. There were decreased sensations below the level of L3 dermatome in both the lower limbs. Perianal sensations were reduced. Ankle reflex and planter (Babinskis) response were absent bilaterally.

His laboratory parameters were normal except for a raised erythrocyte sedimentation rate (70 mm/hour). X-rays of the whole spine, chest, and abdomen were normal. Ultrasonography of the abdomen and pelvis showed abnormal distension of the bladder suggestive of the possibility of neurogenic bladder.

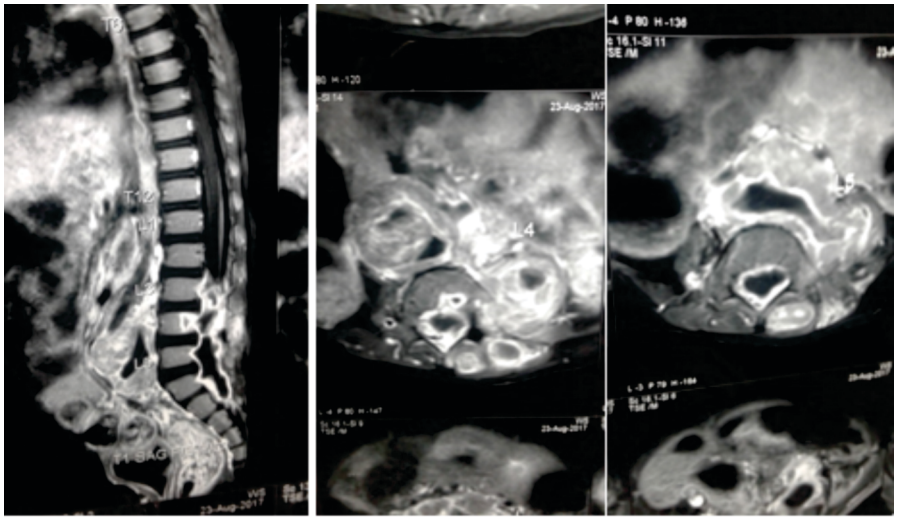

Figure 1: Sagittal projection of postcontrast T1 images tumour mass invading the spinal canal and posterior elements at L3, L4,L5 and S1 vertebral levels. Axial projection of postcontrast T1 MRI image demonstrating tumour mass.

MRI of the lumbosacral spine (Figures 1(a) and 1(b &c)) revealed a soft tissue mass in the spinal canal, posterior elements, and Right Psoas muscle.

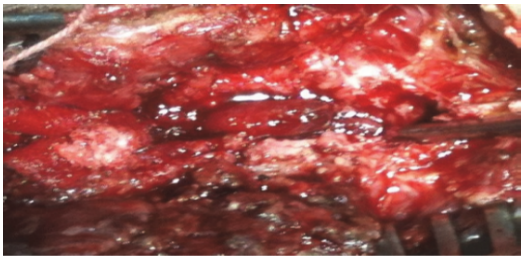

Figure 2:- Laminectomy with decompression showing tumour mass in spinal canal

The child underwent a laminectomy with decompression (Figure 2). The diagnosis of Ewing's Sarcoma was confirmed on histopathology. Histopathology showed small round cells packed in nests(Figure 3). Following surgery the patient had a good recovery. Postoperatively the patient was referred to a specialized oncotherapy centre for needful.

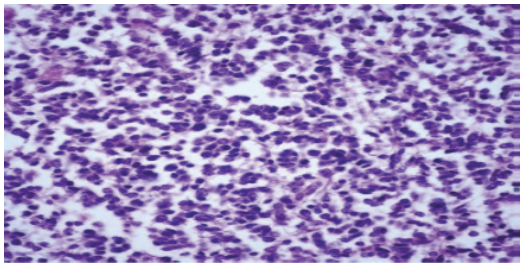

Figure 3: Showing small round cells uniformly packed in nests.

3. Discussion

Ewing's Sarcoma (ES) is a small round cell tumour and accounts for one quarter of all primary bone tumours during childhood. Its peak incidence is during the second decade of life and it is very rare after 30 years of life [7]. ES usually presents with pain and swelling of the affected bone and vertebral involvement occurs in less than 5 percent of cases [8]. It has a poor prognosis but multimodality chemotherapy has increased life expectancy by 40 percent. Primary ES of the spine is a very rare condition [9]. Our case report is an extremely rare case of primary ES of the spine in a two year old boy. The initial interpretation of the MRI scan by the radiologist suggestive of infective process like tuberculosis.

In a retrospective study of Widhe et al., at the first visit, a bone tumour was suspected in only 19 percent of the cases of primary ES of the spine [10, 11]. A high index of suspicion and careful physical examination is required for the diagnosis of this condition. Signs of spinal cord compression may be the only initial indicators for primary ES of the spine [11–13].

Histopathology is the mainstay of diagnosis of small round cell tumours. The differential diagnoses of small round cell tumours include neuroblastoma, primitive neuroectodermal tumours of bone (PNET), malignant lymphoma, rhabdomyosarcoma, and ES.

MRI scan is more sensitive than CT in the early detection on ES [14]. These tumours have variable sensitivity to radiation and chemotherapy due to biological heterogenecity. The classical chemotherapy regimen followed in ES consists of VACA [15].

4. Conclusion

The purpose of this study was to report the incidence of such a rare tumour in a very young child. To the best of our knowledge, report of primary ES of the spine at the age of two years is extremely rare.

References

[1] M. A. Alfeeli, S. Y. Naddaf, and G. M. S. Syed, "Ewing sarcoma of the rib with normal blood flow and blood pool imagings on a 3-phase bone scan," Clinical Nuclear Medicine, vol. 30, no. 9, pp. 610–611, 2005.

[2] Q. Zhu, J. Zhang, and J. Xiao, "Primary dumbbell-shaped Ewing's sarcoma of the cervical vertebra in adults: four case reports and literature review," Oncology Letters, vol. 3, no. 3,pp.721–725, 2012.

[3] Y. Yan, T. Xu, J. Chen, G. Hu, and Y. Lu, "Intraspinal Ewing's sarcoma/primitive neuroectodermal tumors," Journal of Clinical Neuroscience, vol. 18, no. 5, pp. 601–606, 2011.

[4] W. S. Ferguson, "Chronic leg pain in an adolescent male," Medicine and health, Rhode Island, vol. 82, no. 11, pp. 407–409, 1999.

[5] M. J. A. Sharafuddin, F. S. Haddad, P.W. Hitchon, S. F. Haddad, and G. Y. El-Khoury, "Treatment options in primary Ewing's sarcoma of the spine: report of seven cases and review of the literature," Neurosurgery, vol. 30, no. 4, pp. 610–619, 1992.

[6] P. Klimo Jr., P. J. Codd, H. Grier, and L. C. Goumnerova, "Primary paediatric intraspinal sarcomas. Report of 3 cases," Journal of Neurosurgery: Pediatrics, vol. 4, no. 3, pp. 222 229, 2009.

[7] L. I.Dini, R.Mendonc¸a, and P. Gallo, "Primary Ewings sarcoma of the spine: case report," Arquivos de Neuro-Psiquiatria, vol. 64, no. 3, pp. 654–659, 2006.

[8] Y. Mori, H. Tsuchiya, T. Tsuchida, N. Asada, T. Nojima, and K. Tomita, "Disappearance of Ewing's sarcoma following bacterial infection: a case report," Anticancer Research, vol. 17, no. 2, pp. 1391–1397, 1997.

[9] M. R. Grubb, B. L. Currier, D. J. Pritchard, andM. J. Ebersold, "Primary Ewing's sarcoma of the spine," Spine, vol. 19, no. 3, pp. 309–313, 1994.

[10] B.Widhe and T.Widhe, "Initial symptoms and clinical features in osteosarcoma and Ewing sarcoma," Journal of Bone and Joint Surgery—Series A, vol. 82, no. 5, pp. 667–674, 2000.

[11] F. A. Paul, "Ewings sarcoma as an etiology for persistent back pain in a 17-year-old girl after trauma to the back," Journal of the American Osteopathic Association, vol. 95, no. 1, pp. 58–61, 1995.

[12] M. Kogawa, T. Asazuma, K. Iso et al., "Primary cervical spinal epidural Extra-osseous Ewing's sarcoma," Acta Neurochirurgica, vol. 146, no. 9, pp. 1051–1053, 2004.

[13] P. Mukhopadhyay, M. Gairola, M. C. Sharma, S. Thulkar, P. K. Julka, and G. K. Rath, "Primary spinal epidural extraosseous Ewing's sarcoma: report of five cases and literature review," Australasian Radiology, vol. 45, no. 3, pp. 372–379, 2001.

[14] A. S. Goktepe, R. Alaca, H.Mohur, and U. Coskun, "Paraplegia: an unusual presentation of Ewing's sarcoma," Spinal Cord, vol. 40, no. 7, pp. 367–369, 2002.

[15] L. I.Dini, R.Mendonc¸a, and P. Gallo, "Primary Ewings sarcoma of the spine: case report," Arquivos de Neuro-Psiquiatria, vol. 64, no. 3, pp. 654–659, 2006